The Hospice and Palliative Medicine Approach to Caring for Pediatric Patients

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UNIPAC EIGHT

HOSPICE AND PALLIATIVE CARE TRAINING FOR PHYSICIANS • A SELF-STUDY PROGRAM

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Third Edition

UNIPAC 8

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Continuing Medical Education

Purpose

A UNIPAC is a packet of information formatted as an independent-study program. It includes learning objectives, a pretest, reading material, clinical situations for demonstrating knowledge application, references, and a posttest. This independent-study program is intended for physicians and physicians-in-training. It is designed to increase competence in palliative medical interventions for improving a patient's quality of life, particularly as death approaches. Specific, practical information is presented to help physicians assess and manage selected problems. After reading the UNIPAC, physicians are encouraged to seek additional training in hospice and palliative medicine.

Learning Objectives

Upon completion of this continuing medical education (CME) program, a physician should be better able to

- Identify similarities and differences between adult and pediatric palliative care.
- Identify barriers to providing comprehensive palliative care for pediatric patients from the time of diagnosis of a life-threatening condition.
- Use effective communication techniques when discussing palliative care, terminal illness, and death with children and their families.
- Recognize normal and complicated grief and initiate effective interventions for helping families through their bereavement.
- Initiate effective management of psychosocial issues, including social issues, related to children with life-limiting conditions.
- Identify and explain ethical and legal issues related to palliative care for pediatric patients, including the management of refractory symptoms.
- Assess pain in pediatric patients.
- Manage pain and opioid-related side effects in pediatric patients.
- Assess and manage other nonpain symptoms in pediatric patients.
- Assess and manage refractory symptoms in pediatric patients.
- Manage a child's death.

Recommended Procedure

To receive maximum benefit from this UNIPAC, the following procedure is recommended:

- Review the learning objectives.
- Complete the pretest before reading the UNIPAC.
- Study each section and the clinical situations.
- Review the correct responses to the pretest.
- Complete the posttest and evaluation by following the instructions located on page 110 of this book.

Some accrediting organizations may not accept the CME from the third edition of the UNIPAC series if you have already obtained CME for the previous edition of the same UNIPAC. For clarification, please contact your accrediting organization.

Accreditation Statement

The American Academy of Hospice and Palliative Medicine (AAHPM) is accredited by the Accreditation Council for Continuing Medical Education (ACCME) to provide continuing medical education for physicians. AAHPM designates this continuing medical education activity for a maximum of six (6) *AMA PRA Category 1 Credits™*.

Physicians are eligible to receive credit by completing both the online evaluation form and posttest. The Academy will keep a record of *AMA PRA Category 1 Credits™*, and the record will be provided on request; however, physicians are responsible for reporting their own credits when applying for the AMA/PRA or for other certificates or credentials.

Disclosure

In accordance with the Accreditation Council for Continuing Medical Education's Standards for Commercial Support, all CME providers are required to disclose to the activity audience the relevant financial relationships of the planners, teachers, and authors involved in the development of CME content. An individual has a relevant financial relationship if he or she has a financial relationship in any amount occurring in the last 12 months with a commercial interest whose products or services are discussed in the CME activity content over which the individual has control. AAHPM requires that all relevant financial relationships be resolved prior to planning or participating in the activity.

Nancy Hutton, MD FAAP, has disclosed no relevant financial relationships. **Marcia Levetown, MD FAAP,** has disclosed no relevant financial relationships. **Gerri Frager, MD RN FRCPC,** has disclosed no relevant financial relationships. **Kimberly A. Bower, MD,** has disclosed no relevant financial relationships. **Jeanne Lewandowski, MD FAAHPM,** has disclosed no relevant financial relationships. **C. Porter Storey, Jr., MD FACP FAAHPM,** has disclosed no relevant financial relationships. **Stacie Levine, MD,** has disclosed no relevant financial relationships. **Joseph W. Shega, MD,** has disclosed no relevant financial relationships.

Review and Revision

This book was reviewed by AAHPM's UNIPAC Review Task Force and reapproved by the American Academy of Hospice and Palliative Medicine's board of directors.

Term of Offering

The release date for the third edition of this UNIPAC is May 31, 2008, and the expiration date is December 31, 2011. Final date to request credit is December 31, 2011.

Posttest Pass Rate

The posttest pass rate is 75%.

Additional Information

Additional information is available from the American Academy of Hospice and Palliative Medicine, where staff can direct you to physicians specializing in end-of-life care.

Pretest

Before proceeding, please complete the following multiple-choice questions. The answers are listed on page 107 of this book.

1. A 5-year-old child is undergoing a bone marrow aspiration. Despite the use of local anesthetics, she cries and states that it hurts. The most appropriate response is to
   A. believe the patient and give additional pain medication
   B. ask the nurse if the child needs more pain medication
   C. ask the parents if they think the child is accurately reporting the pain
   D. tell the child you have already given enough pain medication.

2. An infant is born with Trisomy 18. Because of his chromosomal anomaly, he is predicted to have no relational potential. In the United States, how do federal regulations affect the choices available to this patient and his surrogates?
   A. Under no circumstances can life support be withdrawn.
   B. An ethics committee must be consulted before discontinuing life support for any child under the age of 1 year.
   C. Only children with diagnosed irreversible coma or brain death are allowed to forgo life support.
   D. When life-prolonging medical interventions will not reverse an ultimately lethal condition, they may be forgone or withheld regardless of age.

3. When using the FACES pain rating scale to assess a child's pain, the correct procedure involves which of the following?
   A. Ask the parent to match the child's expression to one of the drawings.
   B. Ask the child to point to the face that best shows how much pain she has now.
   C. Hold the scale next to the child's face and determine which drawing looks most like the child's expression.
   D. Ask the child which face "on average" represents the way she feels.

4. Which of the following is the best indicator that a child is not experiencing pain?
   A. The child is asleep.
   B. The child has a normal blood pressure.
   C. The child has a normal respiratory rate.
   D. The child says he does not have pain.

5. A young child with congenital heart disease experiences significant intraoperative cerebral ischemia. He is receiving care in a pediatric intensive care unit. Which of the following must be present before integrating palliative care principles into this child's care?
   A. An order not to escalate life-sustaining medical interventions
   B. A prognosis of 6 months or less, if the disease runs its normal course
   C. An understanding that palliative care can include both intensive care and life-prolonging therapies
   D. Transfer of the child out of the ICU

6. Effective management of procedural pain includes which of the following?
   A. Provide analgesia and distraction maneuvers, such as reading a book with the child.
   B. Tell the child to be strong because the procedure will help make him better.
   C. Reassure the child that the pain is not that bad and it will be over soon.
   D. Keep the parents out of the room because they may heighten the child's anxiety.

7. A 14-year-old boy is recently diagnosed with osteogenic sarcoma. There is no evidence of metastasis, and the primary tumor is small. There is a good chance he can be cured of his disease with surgery and chemotherapy; nevertheless, he exclaims, "Why is God doing this to me? I don't want to die!" The best response to his question is to listen to his concerns, reassure him that he is very likely to be cured, and
   A. explore his concerns and call for more spiritual support as needed
   B. share your views about religion and God's intentions
   C. suggest that his anxiety may interfere with his ability to get better
   D. suggest that his spiritual distress is inappropriate, given his prognosis.

8. A 3-year-old child underwent surgery for a Wilms tumor 2 days ago. He continues to report severe pain, despite receiving regular dosages of acetaminophen. When addressing staff concerns about pain treatment, which of the following is a correct statement?
   A. Children of all ages are at higher risk of respiratory depression from opioids compared with adults.
   B. Children are at higher risk of addiction because of their neurologic plasticity.
   C. Children do not experience severe pain because they have not yet developed the nerves necessary to transmit pain.
   D. Children should receive pain medication, including opioids, consistent with their level of pain.

9. Danny is a 10-year-old boy with end-stage leukemia. When planning home care for Danny, which of the following interventions is most likely to be helpful to his family?
   A. Wait until Danny gets home to determine which durable medical equipment is needed.
   B. Discuss Danny's current and anticipated symptoms and their treatments with Danny and his family.
   C. Reinforce to the family that Danny is expected to die within 6 months.
   D. Avoid school-related issues, because he is unlikely to return to school.

10. A child with cystic fibrosis reports severe joint-related pain. Maximal dosages of several NSAIDs were tried, but his pain remains at a score of 7 out of 10. He has been hospitalized for respiratory decompensation only rarely, and his weight is stable. Which of the following is the best intervention for managing his pain?
    A. Psychological interventions alone should be added to his NSAIDs.
    B. Opioids and physical and psychological interventions should be added to his NSAIDs.
    C. Physical therapy alone should be instituted.
    D. Only psychological and physical therapy should be instituted; opioids should not be considered until death is imminent.

11. An adolescent with end-stage cancer who previously had a stable opioid regimen now requires escalating dosages of opioids for pain relief. Which of the following is the most likely cause of the child's request for increased dosages?
    A. Development of tolerance
    B. Development of addiction
    C. Progression of the cancer
    D. Depression; the request may be a veiled suicide attempt

12. One week ago, 4-year-old Brian's sister, Julie, died at home after a long illness. Even though he was told that Julie is dead, Brian asks his mother when Julie will come back. His reaction is indicative of
    A. a state of denial
    B. memory loss due to posttraumatic stress disorder
    C. a developmentally appropriate concept of death
    D. severe psychopathology.

13. Five-year-old George's mother died 2 weeks ago. Since then, if anyone mentions her name, George runs from the room screaming, "Mommy, where are you? Why won't you come home?" Otherwise, he refuses to talk about her. Which of the following strategies is most likely to be effective for a member of the healthcare team to help George explore his grief?
    A. Talk with him about his feelings.
    B. Use creative means of expression, such as play therapy.
    C. Encourage him to write down his feelings.
    D. Tell him that his mother has gone away on a long trip.

14. Which of the following statements about procedural pain is *incorrect*?
    A. Propofol is an effective analgesic for procedure-related pain.
    B. Inadequate management of recurrent procedure-related pain may result in severe, long-term psychological morbidity.
    C. In addition to analgesia, sedation may be indicated for the management of procedural pain.
    D. The management of a child's first experience with a painful procedure affects subsequent responses to the same procedure.

15. A 5-year-old with a newly diagnosed brain tumor has a very poor prognosis and has undergone numerous tests. She is feeling weak and has not seen her friends or gone to school in 2 weeks, but has not undergone any disease-specific therapy. She asks what is going to happen to her. The best response is to
    A. provide only good news and positive statements so she does not lose hope
    B. provide a very detailed description of her condition so she will feel informed
    C. clarify the information desired, then provide clear, honest answers to her questions
    D. suggest that she ask her parents, because they know her best.

16. A 12-year-old child with AIDS-related neuropathy reports severe pain. In this situation, the classes of agents most likely to help in the management of the child's neuropathic pain are
    A. opioids and tricyclic antidepressants
    B. tricyclic antidepressants and phenothiazines
    C. opioids and nonsteroidal antiinflammatory agents
    D. steroidal agents and tricyclic antidepressants.

17. When asked by a concerned parent if her 8-year-old child should go to the funeral of a friend, which of the following is the best response?
    A. Children should not attend funerals until they are older, can understand death, and have developed mechanisms to handle loss.
    B. Regardless of their wishes, children should attend funerals to help them understand the irreversibility of death and to encourage them to get on with their lives.
    C. Children should be offered the opportunity to attend a funeral; if they wish to go, a supportive parent or trusted adult should accompany them.
    D. A child's attendance at a funeral should be based solely on parental preference, regardless of the child's age.

18. Following the death of a 16-year-old boy he had cared for, a medical student enters the room of the grieving family and cries with them. After emerging from the room, the best response of the attending physician is to
    A. gently let him know that it is unprofessional to cry, particularly in the presence of a patient's family
    B. suggest that decreased emotional involvement will help him survive in this profession
    C. acknowledge the emotional impact of a patient's death and offer to spend time talking about his feelings
    D. ask him if he has a personal history of depression and suggest a consultation with an employee assistance program.

19. A 12-year-old girl with end-stage AIDS is experiencing escalating pain when she is dying. To manage escalating pain, all the following are reasonable routes of opioid administration *except*
    A. intravenous
    B. transdermal
    C. subcutaneous
    D. oral.

20. A 3-year-old child undergoes a near-drowning episode. Six months later he is persistently comatose and has had several episodes of aspiration pneumonia related to feedings administered through a gastrostomy tube. The medical team recommends medications and a fundoplication (surgery to prevent gastroesophageal reflux). The family wants to know what their alternatives are. To provide adequate informed consent, the physician must include which of the following statements in the discussion?
    A. Feeding is a comfort measure and cannot be withdrawn.
    B. You may forgo nutrition, hydration, antibiotics, and any other medical therapy that does not promote agreed-upon goals of care.
    C. You may ask for escalating dosages of medication to ensure your child will die in the near future.
    D. You will be reported to child protective services for child neglect if you do not agree to surgery to prolong your child's life.

21. A child with a chronic illness has just died. Hospital policy requires asking for an autopsy on all decedents. When considering the physician's duty to the family, which of the following statements is accurate?

 A. Optimally, discussion about autopsy should be delayed until shortly after the child dies.

 B. Very few families find solace or learn valuable information when an autopsy is performed.

 C. The autopsy will disfigure the body and eliminate the option of an open-casket ceremony.

 D. Limited autopsies can be performed and organs can generally be replaced in the body, if desired by the family.

22. A parent whose 9-year-old son died 2 years ago comes to your office. She relates that she stayed in bed and could not stop crying for 2 months after her son died, that she still cries often (weekly), but has returned to work and is able to care for her home and family. She confides that sometimes, especially in places or situations that remind her of her son, she hears his voice. She wants to know if she is "crazy." Which of the following is the best response?

 A. She is having a normal grief response; a support group for bereaved parents or counseling may help.

 B. She is describing symptoms of psychosis and requires a mental health consultation.

 C. She is having a pathologic grief reaction and should be referred for professional help.

 D. Verify that she is not functioning at home or at work before making a recommendation for hospitalization.

23. Peter, a 6-year-old boy who received a bone marrow transplant, is expected to die in the next 72 hours. His 8-year-old sister was the marrow donor. She and her 4-year-old brother haven't seen Peter in several weeks and are asking to visit him in the ICU. His parents ask for advice about letting them visit. They are fearful that the children will be overwhelmed by the experience. The healthcare team can assist the parents in supporting their children by

 A. reassuring the parents that the children can visit Peter, but help the parents first explain to the children what they will see, and then offer to accompany them

 B. explaining that there is no good reason to subject young children to such a terrible situation

 C. advising against this, as the sister may feel especially guilty, having been his donor

 D. telling the parents that the visit is advisable as long as the parents do not cry in front of their children.

24. A 17-year-old boy is killed by a drunk driver. A policeman arrives at the family's home to deliver the tragic news. The boy's 5-year-old sister is home and overhears the policeman's conversation with her parents. The parents begin sobbing, but the girl asks to go outside to play. The child is exhibiting which of the following?

 A. Callous disregard for her brother

 B. A developmentally appropriate coping response

 C. Signs of significant mental illness

 D. Defiance of her parents

25. Julio, a 15-year-old boy with cystic fibrosis, is in the terminal phase of his illness. Nevertheless, he wants to go to school. However, he cannot tolerate school for longer than 1 hour. His parents ask for advice about the advisability of letting him attend. Which of the following is the best response?

 A. Julio would benefit more by conserving his energy and staying home.

 B. Age-appropriate activities are important to a child's quality of life; Julio can attend school as long as he wants to.

 C. Though it may be beneficial to Julio, it would be too burdensome on family members and schoolmates.

 D. If Julio wants to go to school, he should be able to stay for a minimum of a half-day.

26. Alisha, an 8-year-old child with an uncorrectable complex congenital heart lesion, pulmonary hypertension, and progressive dyspnea and cyanosis, asks if she is going to die. Which of the following statements is the most appropriate response to her question?

    A. Everybody dies sometime.
    B. Your sickness is bad, but you won't die of it.
    C. Can you tell me why you are asking about that today?
    D. Unfortunately, you will die of this sickness.

27. Leonardo, an 18-year-old with end-stage cystic fibrosis, is being treated with antibiotics and his usual regimen of inhaled bronchodilators and enzymes. He has been receiving morphine for relief of breathlessness. For the past 2 days he has needed daily rescue doses totaling 90 mg of immediate-release morphine in addition to his usual dose of sustained-release morphine, 15 mg twice a day. Which of the following would be an appropriate change in his opioids?

    A. 60 mg of sustained-release morphine twice a day, with rescue doses of 15-mg immediate-release morphine
    B. 30 mg of sustained-release morphine twice a day, with rescue doses of 15-mg immediate-release morphine
    C. No change; continue his 15-mg dose twice a day to avoid the possibility of tolerance.
    D. Use a different opioid because he is approaching his maximally tolerated dosage.

28. Which of the following is appropriate monitoring of a change in the medication regimen for Leonardo, from question 27?

    A. Continuous pulse-oximetry at home
    B. Admission from home to the hospital for monitoring
    C. Respiratory-rate monitoring by family 24 hours a day
    D. Patient self-report of relief of dyspnea and side effects

29. A nonverbal child has a history of chronic pain from pancreatitis. Which of the following is usually the least helpful indicator for assessing chronic pain in nonverbal patients?

    A. Difficulty being consoled
    B. Self-limited movement
    C. Change in blood pressure
    D. Decreased interaction with her environment

30. Candice is a 3-year-old girl with severe nausea and occasional vomiting despite regular doses of ondansetron following chemotherapy for a large intraabdominal neuroblastoma. On examination, she has bowel sounds and her abdomen is not distended. Which of the following options is the most appropriate to add to her current antiemetic treatment?

    A. Metoclopramide
    B. Intermittent suction via a nasogastric tube
    C. Dimenhydrinate
    D. Sea bands to both wrists

31. Tony is an 11-year-old boy with spastic cerebral palsy whose pain from spasticity was previously controlled with NSAIDs but now requires around-the-clock morphine through his gastronomy tube. Because opioids are known to be constipating, which of the following is an appropriate bowel regimen?

    A. Monitor Tony's bowel habits for 1 to 2 days; if he doesn't have a bowel movement, start docusate.

    B. Start lactulose now; if he does not have a bowel movement within 1 or 2 days, add senna.

    C. Start both docusate and senna now.

    D. Avoid chronic use of laxatives by increasing his daily intake of fiber and fluids.

32. Sammy is a 13-year-old with long-standing joint pain. He has had chest pain for the past 2 days, which has been poorly relieved with his usual every-4-hour dose of codeine (1 mg/kg/dose PO) and acetaminophen (15 mg/kg/dose PO). He currently rates his pain as 6 out of 10. When evaluating him for the cause of the increased pain, which of the following is the best intervention?

    A. Increase only his codeine dosage by 50% to 200%.

    B. Increase the frequency of his regular dosages of codeine and acetaminophen to every 2 hours.

    C. Discontinue both the codeine and acetaminophen and start morphine, 1.0 mg/kg/dose PO every 4 hours.

    D. Discontinue the codeine only and start morphine, 0.3 mg/kg/dose every 4 hours.

33. A 4-month-old infant has epidermolysis bullosa. She has progressive skin involvement and increased pain despite regularly administered acetaminophen. Which of the following measures can be safely added to the pain regimen for children under 6 months of age?

    A. Begin with the usual 0.1 mg/kg starting dose of SC or IV morphine for children, then titrate to relief of pain.

    B. Increase the acetaminophen only, because infants are at high risk for respiratory depression when treated with opioids.

    C. Begin with SC or IV morphine at 0.025 to 0.033 mg/kg/dose, then titrate to pain relief.

    D. Infants have a rapid metabolic rate; begin SC or IV morphine at 0.15 to 0.2 mg/kg/dose, then titrate to relief of pain.

34. Nine-year-old Lisa has a glycogen storage disease and is profoundly developmentally delayed. She suffers from apparent pain of unclear etiology. Her pain was well managed until recently, when she required a 50% increase in her morphine to achieve adequate pain relief. Her parents' main concern is the onset of jerking movements. Seizures have been ruled out, and the physician has diagnosed opioid-related myoclonus. Which of the following interventions is *least* likely to be helpful?

    A. Discontinue the morphine and begin a different opioid recommended for moderate-to-severe pain.

    B. Add an adjuvant medication in an attempt to decrease opioid requirements.

    C. Decrease her morphine dosage by 25%.

    D. Add clonazepam to treat the myoclonus.

35. A 10-year-old boy has generalized bone pain that he rates as 8 out of 10 at the time of diagnosis of leukemia. Which of the following medications would be the most appropriate therapy for managing his pain?

    A. Pentazocine

    B. Codiene

    C. Acetaminophen

    D. Morphine

36. Which of the following medications should be *avoided* as an analgesic choice for a child with disease-related cancer pain and thrombocytopenia?
    A. Codiene
    B. Ibuprofen
    C. Acetaminophen
    D. Hydromorphone

37. Angela is an 8-year-old child with a variant of spinal muscular atrophy and difficulty breathing. The best method of measuring her dyspnea is to
    A. count her respiratory rate
    B. ask her to rate her breathlessness on a 0 to 10 scale
    C. observe the degree of accessory muscle effort
    D. measure her oxygen saturation by pulse oximetry.

38. Leon is a 12-year-old boy with end-stage AIDS. Despite all attempts to relieve his pain, including multiple opioid trials and adjuvant medications, Leon continues to experience severe neuropathic pain in his extremities. Sedation at the end of life may be the only intervention that will provide him with relief. Which of the following is *not* necessary before proceding with sedation?
    A. Obtain written permission from Leon's primary caregiver allowing sedation at the end of life.
    B. Discuss the rationale and guidelines for sedation at the end of life with all members of the healthcare team and with Leon's family members.
    C. If you are a clinician without sufficient palliative care experience, seek consultation to ensure the symptom is truly refractory.
    D. Talk with Leon about treatment options, including sedation.

39. Which of the following interventions is most appropriate for inducing sedation, if required, at the end of Leon's life?
    A. Increase his opioid by 50%.
    B. Change to an alternative opioid and titrate to sedation.
    C. Add a sedating neuroleptic.
    D. Add chloral hydrate.

An Overview of Pediatric Palliative Care

Demographics Related to Childhood Death

Although childhood death rates generally have declined over the last several decades, in 2005 a total of 53,501 children from birth through age 19 years died in the United States. There were 28,440 deaths of children younger than 1 year, 11,358 deaths of children between 1 and 14 years, and 13,703 deaths of children between 15 and 19 years.[1]

Infant mortality, defined as deaths of children younger than 1 year old, comprises over half of all childhood deaths. Of infant deaths, more than half occur during the first week of life. The most common causes of infant death are disorders related to short gestation and low birth weight or other complications of pregnancy, congenital malformations, deformations, chromosomal abnormalities, and sudden infant death syndrome.

Causes of death of children 1 to 14 years of age are different than those for infants (**Figure 1**). In this group, unintentional injuries and malignant neoplasms are most common. Unintentional injuries are the leading cause of death for those 15 to 24 years old, followed by homicide, suicide, and malignant neoplasms. An array of uncommon genetic and congenital conditions may cause death in later childhood or adolescence.

Similarities and Differences Between Palliative Care for Children and Adults

Palliative care for children differs from palliative care for adults in many ways, including

- the relative rarity of childhood deaths in North America and Europe, where palliative care is most available
- the epidemiology of childhood death and the broad array of rare syndromes, defects, and abnormalities that contribute to uncertainty when determining prognosis
- interpersonal dynamics, both familial and professional
- developmental issues affecting communication and the experience of illness and dying
- legal and ethical issues related to never-autonomous individuals
- less well-developed measurement tools for evaluating pain, other symptoms, and quality of life in the pediatric population
- the paucity of evidence about the effectiveness, or

Figure 1. Leading Causes of Death Among Children Ages 1-14: 2002

Ages 1–4

Unintentional Injury	
Congenital Malformations	3.4
Malignant Neoplasms	2.6
Homicide	2.7
Diseases of the Heart	1.1
Pneumonia and Influenza	0.7
Septicemia	0.5

Ages 5–14

Unintentional Injury	6.6
Malignant Neoplasms	2.6
Congenital Malformations	1.0
Homicide	0.9
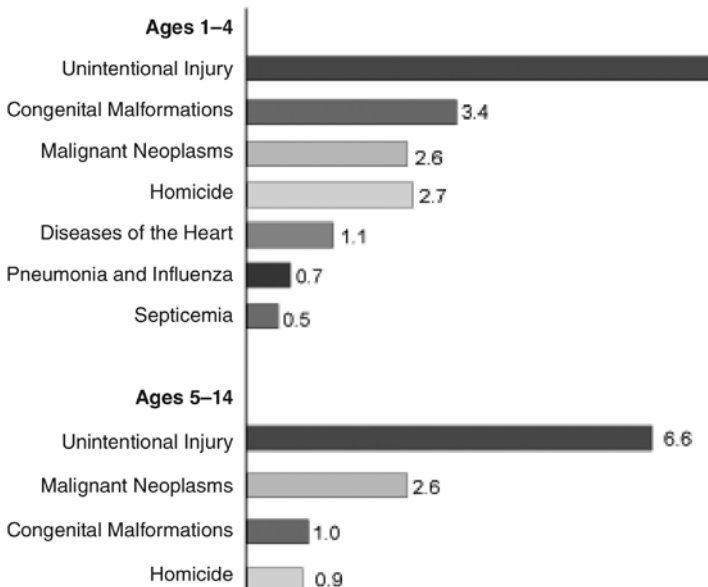
From US Department of Health and Human Services, Health Resources and Services Administration, Maternal and Child Health Bureau. Child Health USA 2004. *Rockville, MD: US Department of Health and Human Services, 2004.[2]*

lack thereof, of symptom-treatment modalities

- school and community issues, including societal expectations
- malignancies of childhood (eg, leukemias and lymphomas)—once they become refractory to primary treatments—that tend to progress fairly rapidly to death (In the adult population, by contrast, long-term metastatic disease is common among fatal adult malignancies like breast and prostate cancer. A few malignancies of childhood, notably brain tumors, may be associated with a prolonged course. Children who die of cancer receive aggressive treatment at the end of life and many have substantial suffering and poorly controlled symtoms.)[3]
- the nature and duration of bereavement for survivors of pediatric death; children's deaths are so challenging for us as a society that we do not have words analogous to "widow" or "orphan" to designate family members who survive a child's death.

For many children living with a variety of congenital syndromes or chronic disorders such as metabolic disorders, cystic fibrosis, muscular dystrophies, and other uncommon disorders, premature death is the expected outcome from the time of diagnosis. When the diagnosis is made at or near birth, families miss the opportunity to experience health before the transition to illness.

Palliative interventions should be initiated at the time of diagnosis and continue throughout the short-ened lives of such children, whether the immediate goal of medical treatment is to prolong life or maximize functioning and comfort.[4-6]

The rarity of many terminal conditions of childhood and the uncertainty about prognosis and timing of death (eg, in most cases, a neurodevelopmentally disabled child will die "unexpectedly" from pneumonia) underscore the need to provide palliative care to children from the time of diagnosis to avoid lost opportunities caused by "unexpected" deaths.

Pediatric Palliative Care

Palliative care is about optimal living despite the presence of a life-limiting condition—it is not about dying. Interventions directed toward prolonging life can be presented to parents as compatible with interventions primarily directed toward palliation; these two types of intervention are not mutually exclusive. Frequently, the same interventions both prolong life and palliate symptoms (eg, treatments such as antibiotics, mucolitics, and chest physical therapy used for children with cystic fibrosis). This dual approach enables children and their families to benefit from family-centered palliative care in a timely manner. The traditional model of palliative care as represented in **Figure 2** fails to meet the needs of the vast majority of children who are ill from birth, who have life-limiting illness, or who die of sudden causes, such as trauma.

Figure 2. Traditional Palliative Care Services

Treatment directed primarily toward cure	Treatment primarily supportive of physical, emotional, and spiritual needs	Bereavement

Diagnosis   Move to Palliative Care   Death
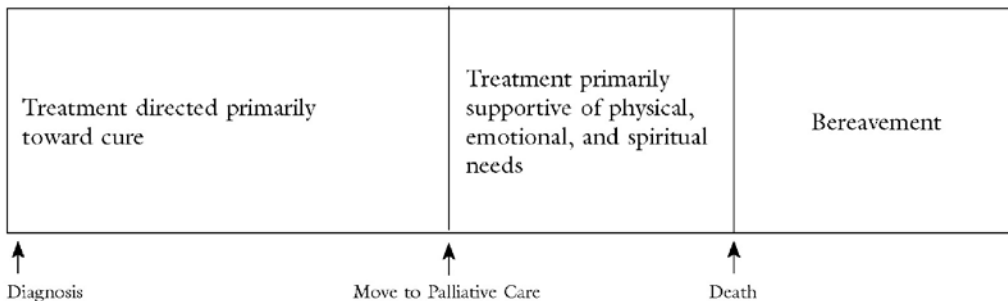
From Palliative Care Services Guidelines: Health Canada. © 1989 by the Minister of Public Works and Government Services, Canada. *Adapted with permission.[7]*

See **Figure 3** for a proposed continuum of pediatric palliative care. Although the figure refers to palliative care for infants, children, and adolescents with AIDS, the continuum is applicable regardless of the child's illness or condition.

The basic components of interdisciplinary palliative care—effective communication, psychosocial care, spiritual care, comprehensive pain and symptom control, and grief and bereavement support—are appropriate from the time of diagnosis, whether or not a child is narrowly defined as being at the end of life.[8,9] Palliative interventions can ease the experience of illness for children and their families regardless of diagnosis or the current goal of care. If and when the goal of prolonging life becomes unduly burdensome, the treatment plan can be modified to further emphasize palliative interventions.

Ideally, the child is cared for from diagnosis to death by the same interdisciplinary care team, which should be competent to provide the essentials of palliative care. If expertise in pediatric palliative care is unavailable locally, needed consultation may occur by telephone or online.

Improving Pediatric Palliative Care

The single most important way to improve pediatric palliative care is to create a widespread understanding that palliative care goals and life-prolonging goals can be pursued simultaneously. To effect important changes in the field, this understanding must be shared by healthcare practitioners, families, and payors.

Education and training in pediatric palliative care are not yet widely available. Although palliative care providers are competent in end-of-life care principles and philosophy and pediatric practitioners are skilled in treating pediatric diseases and are familiar with routes of medication administration, both types of clinicians will likely benefit from additional training in effective communication with children, their parents, and their siblings about death, dying, and bereavement. Additional training in pharmacologic and nonpharmacologic symptom management is also needed. The involvement of knowledgeable healthcare professionals willing to provide palliative interventions from the time of diagnosis can not only profoundly improve the quality of a child's remaining life but also can support the family's bereavement and adjustment to the loss.

Figure 3. Proposed Palliative Care Services for Children

The Continuum of Palliative Care in HIV-Infected Infants, Children and Adolescents

From Oleske JM, Czarniecki L. Continuum of palliative care: lessons from caring for children infected with HIV-1. Lancet. 1999;354:1287-1291. © 1999 Elsevier Science. Reprinted with permission.[6]
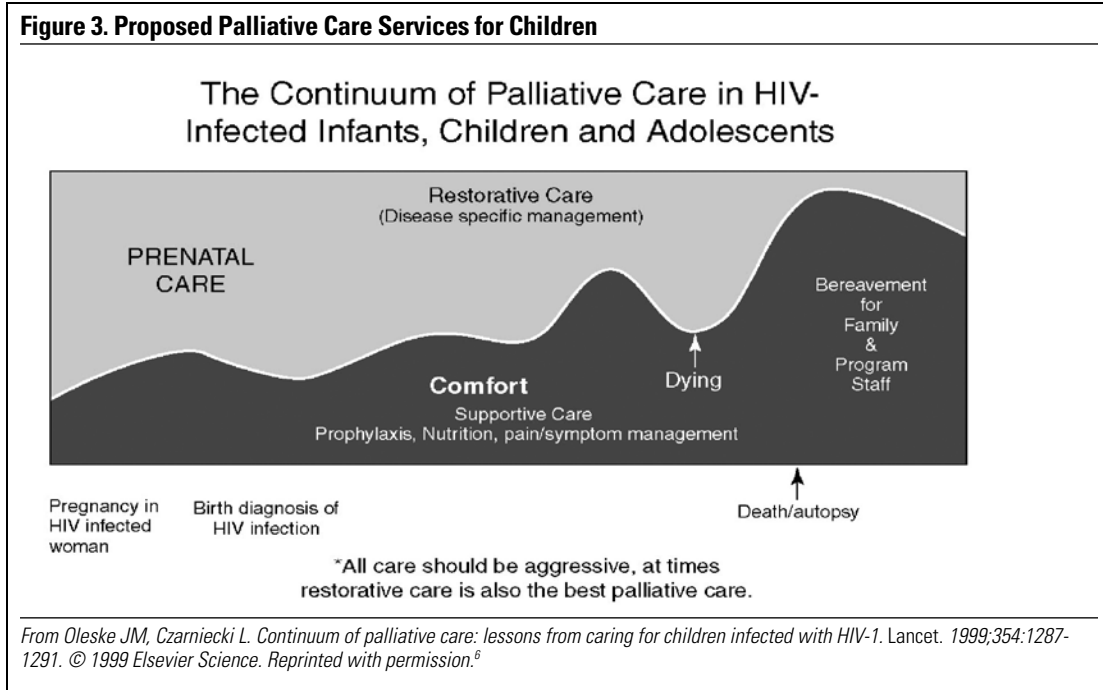
Burnout Prevention in Pediatric Palliative Care

Although the death of a child is always tragic, professional satisfaction can be derived from knowing that the child's comfort and function were maximized throughout his or her lifetime because of palliative interventions, that the family's grief and distress were lessened, and that a peaceful death was achieved. Acknowledging the distress of healthcare professionals and providing them with emotional support can positively influence the therapeutic choices they are willing to offer to the child and family and increase the professional longevity of pediatric palliative care providers.[5] Despite common reservations about providing pediatric palliative care, the majority of hospice and palliative care staff, as well as pediatric health professionals, report a deep sense of satisfaction after improving the quality of life of a child who subsequently dies. On the other hand, poorly managed deaths lead to self-doubt, anxiety, and ultimately avoidance of patients with life-limiting illnesses.

Research in Pediatric Palliative Care

Because the field of pediatric palliative medicine is relatively new, little research has been completed, particularly in the management of symptoms other than cancer-related pain. Evidence-based decision making would be advanced by institutional commitment to funding important areas of inquiry, including pain and symptom management, effective grief and bereavement interventions, and interventions for alleviating emotional and spiritual distress in children and their families.[10]

Historically, children and pregnant women have been excluded from therapeutic drug trials, resulting in a paucity of information about correct dosages and toxicities in these populations. The National Institutes of Health and the Food and Drug Administration have instituted new policies requiring justification for excluding children from proposed protocols. However, previously approved drugs, including standard analgesics, are exempted from the new regulations. Healthcare personnel must continue using some medications without authoritative data on safety, toxicity, and efficacy.

This book is designed to acquaint healthcare practitioners with a base of knowledge and a defined set of skills for caring for children with life-limiting conditions. Some of the recommendations are based on the results of research, and others are based on long-standing experience. The recommendations are provided in the hope that practitioners will integrate the principles of pediatric palliative care from the time of a child's diagnosis, throughout the child's illness, until death.

Reimbursement Issues

In the United States, limitations of reimbursement mechanisms often result in pediatric palliative care either not being provided or being provided without compensation. Valuable time spent on empathetic counseling, recurrent explanations, and identifying developmentally appropriate interventions for the child patient and siblings is very poorly reimbursed. Historically, cognitive or counseling interventions have not been valued by the medical reimbursement system. The time needed to communicate effectively with the child, siblings, and parents (who serve as surrogates) must be appropriately acknowledged and compensated, or pediatric palliative care is likely to exist only in textbooks and manuals.

The Medicare and Medicaid hospice reimbursement models were designed with adult cancer patients in mind, a template that fits poorly with most lethal conditions of childhood. Nongovernmental insurers usually mimic the Medicare and Medicaid requirements. For children to be admitted to most hospice programs, a certification of terminal illness (a prognosis of 6 months or less, if the disease runs its usual course) must be completed by the referring physician. This certification is both a psychological and practical barrier to hospice and palliative care for children. A large number of children living with life-limiting conditions do not meet this hospice eligibility criterion. Even when children are likely to die within 6 months, physicians and parents often fail to recognize the grave prognosis or worry that they are giving up hope if a hospice referral is made. Palliative care services are misunderstood by some payers who consider them duplicative when provided concurrently with home

health care. Moreover, specialized services for children, such as child-life therapy to enhance child communication and coping, are often considered "medically unnecessary," precluding payment for services. Medicare hospice benefit regulations may prevent children whose conditions require hours of nursing care or frequent respite care from simultaneously receiving palliative care services. The insurance needs and financial stresses caused by a child's illness (eg, a child with severe neurodevelopmental impairment requiring suctioning, a gastrostomy tube for feeding, and a seizure disorder requiring medication) may require both parents to work outside the home, a requirement that may be inconsistent with adequate care for the child because of the lack of affordable, skilled pediatric caregivers available when parents work. Creative community alternatives, such as parent exchanges with guaranteed immunity from liability, may serve to fill these gaps. In some areas of Canada, the government funds ongoing respite care and training for laypeople to care for children with life-limiting conditions.

For additional information about reimbursement, see UNIPAC 1.

Communicating with Children and Their Families About Palliative Care, Terminal Illness, and Death

Open, honest communication with children and their families is an essential component of optimal care. Communication barriers at multiple levels exacerbate the challenges associated with caring for children with life-limiting conditions. The barriers may exist between the family and healthcare professionals, the child and family, and among the various health professionals involved in the child's care.

At the time of diagnosis, families are often understandably shaken by the bad news. They may have difficulty hearing and understanding information. Receiving a tape recording of the meeting during which the diagnosis was disclosed can improve comprehension, decrease anxiety, and ensure accurate dissemination of information to absent family members.[11]

Parental reactions commonly include disbelief, shock, and anger. Families often assume a fighting stance of "If we try hard enough, we can beat this problem." Although at the beginning this may be very appropriate, as time progresses and treatments provide less benefit it is often difficult for families to abandon this mindset. They often request continued attempts to prolong the child's life longer than may seem reasonable to healthcare providers and other outsiders. Clear, honest, compassionate communication throughout the course of the child's care helps to prevent this situation.

Parents are also likely to be experiencing such emotional pain themselves that they can hardly bear the thought of subjecting their child to bad news. Parents may have difficulty supporting the disclosure of information to their child. Promoting a safe environment for families to discuss difficult topics with their child helps ensure positive effects of sharing information honestly and compassionately for both the child and the parents. For more information on communication and the process of communicating bad news, see UNIPAC 5.

Developmental Issues

Communication with children is affected by many factors, including psychosocial issues and developmental stages. Children's thought processes differ substantially from those of adults; moreover, a child's usual level of understanding is affected by personal experiences, emotions, and the context of the situation.[12]

Euphemisms

Young children, generally those younger than 8 years, require concrete explanations about illness and death. The use of euphemisms (inexact terms to "soften" the message) can be confusing and frightening; for example, saying that "Grandma has gone away" instead of "Grandma has died" may lead to childhood fears that anyone who "goes away" will never return. Children need to understand that people who have died do not eat, their heart and lungs do not work, they do not sleep or wake up, and they do not feel pain, hunger, cold, or sadness.

Finalism and Attribution

Children's perspectives are also governed by finalism, the belief that bad things happen to people who are bad and that being good protects people from bad things. Such beliefs can distort a child's (and often an adult's) understanding of the causation of illness and may produce unacknowledged or unsuspected feelings of guilt. For some children, admitting to having pain may be the same as admitting that they have been bad. Skilled communication is necessary when asking children about their pain, their thoughts, and their feelings.

Attribution refers to the reason for an event. When caring for sick children it is important to ask them, for example, "What do you think is happening to you now?" This question should be followed by "Why do you think this is happening?" These questions may elicit otherwise undetected fears and misconceptions that need to be addressed. For example, a child with

newly diagnosed leukemia may think that she acquired the disease because she disobeyed her parents and ate too much chocolate. When not addressed, such fears and feelings of guilt cause significant unnecessary suffering.

Correcting misinformation and providing developmentally appropriate explanations are important therapeutic interventions when caring for children. Clarifying the reason for a child's question, determining what and how much information is desired, and pacing the discussion in response to the child's cues are essential principles in pediatric palliative care. For example, an appropriate response to a child who asks "Am I dying?" might be, "I wonder why you are asking me that today?" Responses could range from "My leg is hurting more and more, and my Grammie died when she hurt like this" to "Mommy is crying a lot. I'm worried about her." The child's response may indicate a need for improved pain management or additional psychosocial support for the parent. A child who asks, "What happens after you die?" might want to know where the body is kept before it goes to the grave. Failing to ask the child for clarification may lead healthcare professionals to assume such questions automatically indicate spiritual distress rather than, in some cases, simple curiosity.

Egocentrism

Childrens' egocentrism may cause them to believe that everyone knows what they are thinking. They may not realize they have to communicate their thoughts verbally. Communication can be encouraged by soliciting the child's opinion and clearly indicating that his or her ideas are important in part by acting on articulated concerns whenever possible.

Expressive Therapies

Effective communication with younger or disabled children may require creative interventions to interpret the child's experience, for example, using expressive therapies such as art, play, or music therapy. Specialized training may be needed to use such therapies. In *Care of the Dying Child,* Richard Lansdown describes a 5-year-old boy who was unwilling to discuss his illness.[13]

When Lansdown and the boy built a LEGO room together, the boy described the room as a "laboratory where people's blood was tested." When asked what happens if treatment doesn't work, the child replied that the people would die. When asked if he was going to die, the boy said he thought so but didn't want to. This led to further exploration of his fears and amelioration of his anxiety. Even simple observations of behavioral changes, such as increased irritability with medical treatments, can provide insight into a child's perception of the illness and wishes regarding treatment.

The Child's Participation in Treatment Decisions

Poor communication is not uncommon between parents and a sick child about the child's disease and prognosis. Parents frequently avoid discussing difficult issues such as prognosis with their child because they are concerned the child will not be able to cope with the information. Children are often much more aware of what is happening with their health than they allow their parents to know. The parents' attempt to protect the child and the child's desire not to upset the parents often interfere with communication.

Because some children fear abandonment more than pain or death, they often suffer in silence to avoid alienating significant others. Parents may need guidance on communicating effectively with their child about pain and suffering, as well as recognizing when the goal of prolonging life is no longer reasonable. In one study conducted in Sweden, 429 parents who had lost children to cancer answered survey questions about whether or not they had talked about death with their child. One hundred forty-seven parents reported talking with their child about death. None of these parents regretted having this conversation. In contrast, 69 of the 258 parents (27%) who did not talk with their child about death regretted their decision to forgo this conversation. Parents who had sensed that their child was aware of his or her imminent death were more likely to regret having given up the opportunity to talk with their child about death.[14]

The primary team's psychosocial staff or the hospice and palliative care staff can help the child's family and attending physician clarify options, make decisions, and resolve conflicts. As developmentally appropriate, children should be included in treatment decisions by being asked how they think the treatments are going and if they have any wishes that their parents or care staff can help them achieve. In such situations, some children have responded by saying they don't believe the treatment is working, they wish they could end it, and they want to spend more time away from the hospital. However, they may choose to continue such treatments because of their parents' wishes.[15,16] Some parents elect to continue treatment despite the expressed wishes of the child; however, compromise can usually be reached by developing a mutually agreed-upon time frame and parameters; for example, "Let's try this for 2 weeks, then see how you are responding to the treatment and how you feel about continuing."

Like adults, children want to control their surroundings. Decision-making skills improve with practice, age, and maturation. Children with normal intelligence (IQ above 70) have opinions about their care, and they know better than anyone the burdens of therapies and tests. For example, a 2-year-old child can choose whether a blood test is drawn from the right or left arm. For chronically ill children, opportunities will likely arise to gradually increase a child's involvement in decision-making. Chronically ill children also know best what their bodies are telling them. Myra Bluebond-Langner, in her classic work *Private Worlds of Dying Children,*[17] documents significant changes in the behaviors of even very young children (3 years old) when they begin to feel through internal signals that a therapy is not working and they will soon die. They become irritable and uncooperative in a manner inconsistent with previous behavior patterns.

Clinicians and families should honor a child's priorities, goals, and preferences. When physicians communicate needed information clearly and compassionately in a developmentally appropriate manner, then present potential options and solicit the childrens' opinions, they provide their patients with opportunities to participate in the development of a care plan that is consistent with personal experience and philosophy.

Parents' and siblings' bereavement often is eased by knowing that they respected and honored the child's wishes and showed their love by assisting with the achievement of final goals. **Table 1** offers some suggestions for supportive communication with children.

Table 1. Suggestions for Supporting an Ill Child[18]

Listen to what the child needs and wants.

Children are capable of being in control of their dying process; support them in whatever ways they need.

Grief work can be facilitated by encouraging the child to participate in the funeral planning, burial arrangements, and other rituals.

Children are curious about their illness, its prognosis, and about dying; provide opportunities for the child to talk.

Be comfortable with your own beliefs about death and dying so you can approach a child with openness and honesty.

Let the child control the conversation and set the pace for questioning.

Look for cues to what the child really wants to talk about.

Do not just lecture or give information.

Be truthful.

Use simple, concrete language.

Use pictures and stories for both gaining and sharing information.

Be aware that simply giving children the "facts" can be inadequate because of their different sense of reality; always verify a child's understanding of your answers and explanations.

Be prepared to accept fears and anxieties experienced by children that may seem irrational from an adult perspective.

Children often have fears related to separation and pain, such as separation from their mother, painful or traumatic procedures, and the deaths of other children. Anticipate these fears and provide opportunities for the child to voice such fears.

A Child's Acquisition of Information About Illness

A child's understanding of an illness both in terms of its severity and its implications or meaning about outcome ("I'm so sick from this, I could die") is related to events that occur during the course of the illness. See **Table 2** for a description of a sick child's acquisition of information about an illness and corresponding perceptions. Although the original table was based on a cancer model with relapses and remissions, it is applicable to children living with other life-limiting conditions. The child's understanding of illness is a dynamic process without a rigid sequence. Children frequently vacillate between phases.

Children's Concepts of Death

Children are not born with concepts or fears of death. Their concepts progress from infantile fears of separation from the warmth and smell of caretakers to eventual awareness of more existential constructs. Without ever being told, even very young children know when they are seriously or terminally ill.[19,20] Parents' denial or pretense, or denial or pretense from other significant adults, only cause children to lose trust in their caregivers and feel more frightened and isolated.[17]

Table 2. Phases in a Sick Child's Acquisition of Information and the Critical Events That Influence Understanding of Illness

Phase	Child's Information	Trigger Event	Child's Self-Concept
1	It's a serious illness (not all children know the name of the disease)	Parent being informed of diagnosis (parents often are a primary source of information for children)	"I used to be well, but now I am seriously ill." For children with chronic disease: "I was in my previous state of health, but now I am seriously ill."
2	The names of the drugs used in treatment, how they are given, and their side effects	Parents being informed that the child is in remission or the condition is worsening; child speaking to other children with similar illnesses	"I am seriously ill and will get better."
3	Purposes of special procedures and additional treatments consequent to the side effects of therapy and the relationships between particular symptoms and procedures	The first relapse or exacerbation	"I am always ill and will get better."
4	A larger perspective of the disease as an endless series of relapses or exacerbations with periods of improvement	Several further relapses or exacerbations and periods of improvement	"I am always ill and will never get better."
5	A perspective of the disease as a series of relapses or exacerbations and periods of improvement, ending in death	Child learns of the death of an ill peer	"I am dying."

Note. *A child's perception may be influenced by peers with serious illness and by the presence of a chronic disease, such as cystic fibrosis or muscular dystrophy. From the time of diagnosis, the child should be kept informed in a developmentally appropriate manner.*

From Bluebond-Langer M. The Private World of Dying Children. *Princeton, NJ: Princeton University Press, 1978. © 1978, Princeton University Press. Reprinted with permission.*[17]

Lisa

Lisa, a 10-year-old child with advanced Ewing's sarcoma, understood that her chemotherapy had been changed because "it was no longer working and [her] doctors were not sure if the new one would stop the cancer either." She had not discussed her impending death with her parents or her brother Sam out of fear that they couldn't handle the stress. They, in turn, didn't inquire about her worries because they "didn't want to upset her." However, Lisa confided in members of her care team, who were able to listen to her distress. She said she was having nightmares and asked questions about the likelihood of getting better. She clearly indicated an understanding of the seriousness of her illness.

Question

At this point the team should
A. call the oncologist and ask her to clarify the likelihood of cure
B. protect Lisa's family by telling Lisa not to worry
C. gently offer to sit down with Lisa, Sam, and their parents to talk
D. refuse to get involved since Lisa may not be "terminal."

Correct Response and Analysis

The correct answerer is C, even though it is challenging to participate in such difficult conversations.

The Case Continues

The team suggested to Lisa's parents that she was ready to talk about her hopes and fears. They began the discussion by saying, "We really hope this new treatment will get rid of your cancer." Lisa responded, "Me too, but what if it doesn't work?" The staff responded, "Maybe, while hoping for cure, we should also make some plans in case that doesn't happen. What is most important to you now?" Lisa responded, "I'd like to perform in my school play next month and have friends over afterward for a party at the house." At this point, she was comfortable enough to express a concern that had been bothering her. She said, "If I do die, I worry that you will miss me and will be very sad." Her parents reassured her. "We would be very sad and would miss you terribly. We will always love you and remember you. It would be very different without you if you die, but we promise we will help each other and Sam through this."

Children's wishes about their care at the end of life are shaped by their stage of development and their illness experience. Infants and preschoolers need presence, love, and affection from their families. School-age children often want anything that will allow them to return to school, to be with friends, and to learn. Because older teenagers realize their opportunities to experience the benefits and responsibilities of adulthood are slipping away, they may wish to get married, graduate early from high school, try new activities, stay with friends, or live alone. The child's goals and the family's goals may not be the same. The primary care or hospice and palliative care team can help ensure that important goals are attained by providing excellent symptom management and facilitating communication between the child and parents.

See **Table 3** for information on a child's concepts of death and related supportive interventions. Clinicians should consider the developmental stage when communicating with seriously ill or bereaved children about death and dying. However, a child's understanding of death is often affected by intellectual capacity, the illness experience, and other life events. The recommendations in Table 3 must be used only as a guide. The critical points to remember are
- quality-of-life issues are affected by a child's understanding of illness and death
- a child's view of his or her own quality of life should be honored
- a child's wishes about his or her care should be considered and honored whenever possible.

Table 3. Well Child's Concepts of Death, Implications, and Recommended Supportive Interventions[17,18,21-23]

Life Period	Some Major Characteristics	Predominant Concepts of Death	Supportive Interventions
Infancy: 0-2 years	• No conscious thinking • Limited language abilities • Reality that is based on physical needs being met • Experience of the world through sensory information • Awareness of tension, the unfamiliar, and separation	• Separation	• Provide maximum physical relief and comfort • Provide comfort through sensory input (eg, touch, rocking, sucking) • Provide comfort with familiar people and transitional objects (eg, toys)
Early Verbal Childhood: 2-6 years	• Magical thinking • May play with stuffed animal by repetitively laying it down "dead," then standing it up "alive"	• May see death as reversible • May not believe death could happen to them • May equate death with sleep • May believe they can cause death by their thoughts, such as wishing someone would go away (magical thinking)	• Minimize the child's separation from usual caregivers (eg, parents), or provide reliable and consistent substitutes • Dispel misconceptions about death as a punishment for bad thoughts or actions and the child's guilty feelings • Provide concrete information about the state of being dead (eg, "a dead person no longer breathes or eats")
Middle Childhood: 7-12 years	• The child may request graphic details about death, including burial, decomposition, etc. • In the early stage, understanding that death can result from external causes, such as accidents • In the later stage, understanding that death can result from internal causes, such as illness	• Generally aware that death is final and irreversible but view it as unpredictable • Aware that death is personal; it can happen to them	• Evaluate for fears of abandonment, destruction, or body mutilation • May benefit from specifics about the illness and treatments and reassurance that treatments are not punishments • Maintain the child's access to peers • Foster the child's sense of mastery and sense of control
Adolescence and adulthood: older than 12 years	• May want to speak about unrealized plans (eg, school, marriage)	• Understands the finality and universality of death, but may feel distanced from it • Has the ability to develop natural, physiological, and theological explanations of death	• Allow the child's expressions of anger • Provide privacy for the child • Support reasonable measures for the child to achieve independence • Maintain the child's access to peers • Consider peer support groups for the child

Modified from Stevens M. Psychological adaptation of the dying child. In: Doyle D, Hanks GWC, Cherny N, Calman K, eds. Oxford Textbook of Palliative Medicine. *3rd ed. London: Oxford University Press; 2005:799[24]; Rando TA.* Grief, Dying and Death: Clinical Implications for Caregivers. *Champaign, IL: Research Press; 1984: 385-391. © 2005 Oxford University Press. © 1984 Research Press. Adapted with permission.*

Presenting the Concept of Pediatric Palliative Care to Families and Professionals

Because it is difficult for both family members and physicians to acknowledge a child's life-limiting condition, they may reject the concept of hospice care, in particular the label "terminally ill." Although unwillingness to acknowledge a fatal illness or condition also applies to adult patients, it is even more problematic when used for children, whose deaths are not considered part of the natural course of events in our culture. Parents may be very reluctant to withhold resuscitation or forego therapies that might become available in the future. In addition, professional uncertainty about life expectancy often precludes children from receiving needed palliative care services when programs require a prognosis of 6 months or less.

Vocabulary

Choice of vocabulary is particularly important when explaining palliative services to families. Families may reject the word *hospice,* equating it with death and limited medical interventions. The terms *supportive care* or *palliative care* may be more acceptable because they more accurately express the concept that intensive palliative interventions will always be provided to ensure comfort regardless of other interventions.

Initiation of Pediatric Palliative Care

The optimal time to initiate comprehensive, family-oriented, child-centered palliative care is at the time of diagnosis of a life-limiting condition, whether or not prolongation of life continues as a goal.[5,9,25-27] In some instances, palliative care may be the sole focus of care, for example, when a child is born with an inoperable cardiac lesion. In other cases, life-prolonging and palliative therapies may be provided concurrently, as with cystic fibrosis. Regardless, integrating palliative care principles at the time of diagnosis helps relieve the child's and family's physical, emotional, and spiritual suffering.

Including the child in decision making is an integral component of palliative care planning and decision making. As developmentally appropriate, clinicians may focus discussions on answering the child's questions, soliciting the child's wishes about care setting and caregivers, clarifying the child's expectations, and addressing his or her concerns about the management of nausea, pain, and loneliness. Communication is often effectively facilitated by child-life therapists or psychologists.[9,16]

Emotional Support in Care Planning

Members of the interdisciplinary care team and hospice and palliative care practitioners can provide a forum in which parents can express their concerns about decisions to continue or forgo further life-sustaining treatment and receive reassurance that both are loving choices. Emotional support is critical to ease the family's distress as the goals of care transition from primarily life prolonging to primarily palliative.

The ability of the primary care team to continue caring for the child at home is often a determining factor when parents consider care settings. Care in the home is most likely to result in improved patient comfort and good family bereavement outcomes because of increased contact and participation in care. However, some families prefer the child's death to occur in a hospital.

Discussions of interventions for anticipated symptoms and pain exacerbations must be included when developing treatment plans and preparing the family and child for care in the home. Gentle exploration of the family's main concerns (and the child's, as developmentally appropriate) about the last hours of life and the death event itself is an important part of preparing the family. A team member should gently describe how the child may appear in the final hours of life, including changes in breathing patterns and levels of consciousness. Many families have subsequently expressed appreciation for this information, saying that they felt more prepared and less afraid when changes occurred.[28]

Juanita

Juanita was a 3-year-old child with profound cognitive impairment and seizures from a congenital brain malformation. She was hospitalized numerous times, each admission requiring blood tests and repeated IV access for antibiotics to treat pneumonia. On the tenth admission, her family, growing increasingly concerned about Juanita's comfort, began wondering if they were helping or merely hurting Juanita.

Question

How should such a difficult situation be approached?
A. Ask the chaplain to come pray with them.
B. Report them to Child Protective Services
C. Offer palliative sedation
D. Schedule a time to meet with the family in a private conference area.

Correct Response and Analysis

As difficult as it may be, there is no substitute for a caring discussion of the issues, so the correct response is D. During a discussion of care options, the physician acknowledged that loving families worry about such issues. Immediate and long-term goals for Juanita were explored using the following questions about medical interventions (hospitalization, blood tests, and IV antibiotics) to guide the discussion[9]:

- How likely is it that continuing the current medical interventions will reverse the problem with her brain?
- How likely are the current interventions to prevent further deterioration of her condition and associated medical problems, including seizures and recurrent pneumonia?
- Will the current medical intervention improve the way Juanita feels?
- Could the current medical intervention make Juanita feel worse? If so, for how long?
- What is it like for Juanita to have the current medical treatments?
- How do Juanita's current medical treatments impact the family?
- What is likely to happen with and without the current medical treatments?
- Are there any alternatives to the current medical treatments? (If there are alternatives, address each one, using the above questions.)

The Case Continues: Goal of Care and Revised Treatment Plan

During the discussion, it became clear that the family's main goal was to enhance Juanita's comfort. They no longer believed that the burdens of invasive treatments were justified. Together, they developed a treatment plan that would ensure Juanita's comfort and control her fever, cough, and seizures while forgoing painful treatments for her recurring pneumonia. The plan also addressed increases in nursing and psychosocial support while the family cared for Juanita at home. In addition, needed medical equipment and respite care were discussed. The plan assured more comfort for Juanita and increased emotional and practical support for Juanita's parents and siblings, interventions likely to improve their confidence when caring for her at home.

Care Setting

The interdisciplinary team should discuss and honor the child's and family's wishes for care setting (eg, home, hospital, hospice inpatient facility) whenever possible. With support from an interdisciplinary care team, families may decide to care for their child at home until death occurs, to diminish the child's sense of isolation. When home care is chosen, the primary care team and hospice and palliative care staff should be available as resources, and a healthcare professional should be available 24 hours a day. It is important to remember that even with optimal interdisciplinary team care, the medical needs of some children may overwhelm a family's capacity to provide this care, necessitating in-home nursing care or admission to a hospice or hospital facility.

Regardless of care setting, the child and family should receive emotional and spiritual support, and the siblings and parents should be encouraged to participate in the child's care as much as they desire.[29] Participating in care is important for healthy sibling bereavement, often alleviating a sense of being alone, frightened, unvalued, unhelpful, and uninformed.[30,31]

Family Support

Families with seriously ill children need education, emotional support, and practical assistance.[32] Because parents often want to perform as much of the caregiving as possible, they need to feel confident about managing symptoms at home and about allowing their child to die peacefully in familiar surroundings.[33] Caregivers often need respite from the responsibilities of caregiving, whether for a few hours or a few days, during prolonged chronic illness. Caregivers usually appreciate help with everyday chores and with the care and amusement of the patient's siblings.

Families with seriously ill children are frequently young and often have limited financial and emotional resources. They may feel abandoned by friends and family who, not knowing what to say or do, avoid the family altogether. Siblings commonly lack their parents' full attention and may exhibit anger, jealousy, guilt, and doubts about self-worth.

Some families will benefit from support groups, which may decrease their sense of isolation and help dispel misconceptions about certain topics, such as withholding information from children. Other options should be made available, including one-on-one counseling or family therapy with experienced, trained professionals such as psychologists, psychiatrists, child-life therapists, or social workers. Along with other professionals, including members of the primary care team, such healthcare professionals can talk with siblings about their concerns and help parents interpret the siblings' responses, thus averting unnecessary misunderstandings and conflicts.[34]

In addition, social workers can assist with common problems, such as loss of income resulting from one parent remaining at home to provide care. They can also help with making funeral arrangements, talking with siblings about death, and informing the school and community about the child's condition and ultimate death. Chaplains and child-life therapists can provide valuable help in these situations, as well.

Autopsy

Hospital policy may require that an autopsy be requested from families for all decedents. Ideally, discussions of autopsy should occur before the child dies. When an autopsy is performed, the family often learns important information that may provide them with some sense of solace and more complete information for reproductive planning. Parents should know that an autopsy will not disfigure their child's body, even for an open-casket funeral. If the family desires, a limited autopsy can be performed; organs are usually replaced afterward.

Physician's Role

Physicians with palliative care expertise can improve symptom management, support patients and families in establishing goals of care, assist with grief work, facilitate conflict resolution, maximize the child's quality of life during the time that remains, and support the child and family as death approaches. An effective approach to caring for dying children and their family members includes the following behaviors:

- acknowledging the seriousness of the illness
- clarifying the child's and family's treatment goals and reevaluating them as needed when changes occur in the child's condition
- providing a series of options to families regarding treatments and supportive therapies
- establishing a family care plan, including preferred location of care and interventions at the time of death
- providing continuity of care with the same team of caregivers
- recognizing the multidimensional nature of the child's and family's suffering.

Grief and Bereavement: Family Members and Medical Caregivers

The death of a child is often described as causing the deepest pain imaginable for a family. A child's death frequently results in significant feelings of guilt for parents and siblings, contributing to distress throughout the entire family system. To ameliorate these adverse effects, the interdisciplinary team should encourage a patient- and family-centered dying process whenever possible. Even when a child's acute, fatal conditions are managed in pediatric and neonatal intensive care units, palliative interventions and family involvement are important.

Grief Support Through Chronic Illness

When a chronic illness is likely to result in the death of a child, medical caregivers should provide emotional support for the entire family throughout the child's life. Because the idea of a child dying is so aversive in our society, hospice and palliative care programs may need to serve as family advocates to ensure the child's physical and emotional suffering are acknowledged and carefully considered when making decisions about treatment. Such support can result in less suffering for the child and, in many instances, in spiritual and emotional growth resulting in a the child's parents, grandparents, and siblings achieving a healthier bereavement experience.[31] In one Swedish study, 449 parents who had a child who died of cancer 4 to 9 years before the study answered survey questions about whether and to what extent they had worked through their grief. Parents who had shared their problems with others during the child's illness and who had access to psychological support during the last month of their child's life were more likely to have worked through their grief.[14]

Ongoing grief support for the loss of an expected normal, healthy life is vital for both the affected children and family members. Parental grief and guilt about a child's life-limiting condition can seriously impair the family's quality of life and its ability to make medical decisions. Siblings, often forgotten in the wake of a crisis, are affected by the tragedy not only during the lifetime of the affected child but also throughout their own lifetimes. Grandparents, who experience twofold grief over losing a grandchild and their inability to remedy the situation for their own child, may also be overlooked. They may receive fragmented and inaccurate second-hand information.

Assisting Siblings

Grieving children frequently alternate between expressing their grief and seeming to ignore the situation. It is not unusual for young children to go and play immediately after a death, even if the death was of a sibling. Parents need to be warned ahead of time that their surviving children may use play or visiting friends as coping mechanisms; such activities bear no reflection on the depth of their feelings about the deceased person. Some children are uncomfortable witnessing their parents' distress and want to leave, and others want to stay to provide comfort for their parents. Children must be encouraged to cope in ways that are most effective to them as individuals. Keeping to routines enhances a feeling of safety and comfort. Unacknowledged grief can result in acting-out behaviors. Setting limits and enforcing the usual discipline help grieving siblings understand they are still important.

Because parents may be too distressed to provide adequate emotional support for their children, other adults who are emotionally close to the children can help by providing necessary support and assistance. A sibling's ability to perform schoolwork may deteriorate. School counselors may be of help, particularly if services are provided free of charge. On the other hand, school counselors may feel unprepared to help a student with grief and may welcome additional training from local hospice staff or other trained personnel.

The Moment of Death

Even in the context of prolonged illness, death, no matter how long it has been anticipated, is always a surprise and a shock, even for people who have been coping very well. Often, the moment of death is more

emotionally distressing than the family had imagined it would be. The irreversibility of death is acutely felt when the last breath is taken. Usual responses include statements such as "But he was just here!" "I'll never hear his laugh again!" and "It didn't seem like he would go so soon."

Bereavement

Grief begins at the moment of diagnosis, fluctuates with remissions and exacerbations of the disease, escalates at the time of death (though it may be mixed with feelings of relief and guilt), and continues at varying levels for years afterward. Each individual grieves in his or her own way. It is important to encourage the family and their friends and communities to respect and support the grieving styles of each person as long as they are not self-destructive.[35] Grief manifests in many ways, for example, hearing the child's voice, "seeing" the child everywhere, crying, becoming silent, needing exercise, needing to be alone, or needing to talk. Parents commonly grieve very differently, with fathers more often preferring silence and solitude and mothers often crying and needing to talk. The silent partner can help the more expressive partner go to counseling or support groups. The demonstrative partner should avoid accusatory remarks such as "You don't even care!" and respect the more silent partner's need to get away (eg, going on a fishing trip).

Bereaved parents often experience renewed and intense grief on occasions that would have been significant benchmarks in the life of the deceased child.[36] Events such as the child's birthday, high school graduation, wedding, or the birth of a grandchild may rekindle intense grief. A helpful intervention is to warn bereaved parents and siblings that exacerbations of grief can occur but do not reflect mental illness or emotional instability. Research has shown that grief profoundly effects surviving siblings and parents for at least as long as 9 years after the death of a child.[14] Complicated grief is not measured by duration alone but by the level and duration of the bereaved person's personal neglect, dysfunction, or self-destructive behavior.

Adapting to a New Reality

Over time the pain of grief lessens, but it never disappears entirely. The lessening of pain and a renewed ability to once again enjoy life do not constitute abandonment of the deceased child. Families need permission to adapt to their new reality and to understand they are not abandoning their child when they no longer dwell daily on memories of the deceased. Some families benefit from modifying their traditions, such as holiday-related rituals, which can help them feel more comfortable celebrating despite the loss of a loved one.

The Grief of Medical Caregivers

Medical caregivers also grieve the loss of patients. Opportunities for professional caregivers to grieve their losses help prevent burnout, reduces employee turnover, and prolong the willingness of healthcare professionals to care for dying children. The following strategies can be healing for healthcare professionals:

- sharing deep emotions with fellow staff, friends, and spouses at the time of death
- attending the funeral
- participating in formal or informal memorial services during which bereaved family members and members of the healthcare team share memories of the deceased child.

Interdisciplinary interventions have proven to be helpful in an urban academic medical center.[37] Writing condolence cards, journaling, writing poetry, and walking are other means of self-care.

For more information on grief and bereavement, see the articles referenced above as well as UNIPAC 2.

School and Community Issues

Because terminal illness and death in school-age children is relatively rare, witnessing the sudden death or decline of a child with a life-limiting condition can be shocking for the entire community, particularly for other children. Children are often taught by their parents' examples and society to fear death. Young children may assume that all illness is transmissible. To encourage their acceptance of and interaction with a child living with a life-limiting condition, healthcare professionals should provide schools with general information about illness and death. With the child's and parents' permission, it can be helpful to provide information about the child's specific illness or condition. Children need to know why an illness or condition occurs and that it is not contagious. Issues of guilt and self-blame should be addressed because of the frequency of magical thinking. For example, a child might think, "I used to hate Jessie and now he is dead. I must have made him sick because I wanted him to go away."

Preparing for a Child's Return to School

Before a child's return to school, explanations of his or her appearance, abilities, and limitations will increase the likelihood of a good school experience and may maximize the school's willingness to be flexible regarding schedules and other administrative matters. The child and family are usually willing to share this information after explanations about its relevance. Nevertheless, school personnel must emphasize and respect the confidential nature of some information.

In addition to these general issues, the child's parents or a member of the interdisciplinary care team should give specific information to school personnel most likely to interact with the child about the medications that the child will need to take while at school, how to treat breakthrough pain or other symptoms, and, in case of emergency, any restrictions on normal procedures such as an out-of-hospital do not attempt resuscitation (DNAR) order. School personnel may have difficulty understanding and absorbing the concept of an out-of-hospital DNAR order, and they may require extended discussion about this topic, particularly because it implies the child may experience a life-threatening event or may even die at the school. Ideally, clinicians should provide all information in writing, documenting what interventions to use when symptom management is required, including specific medications and dosages and nonpharmacologic interventions. Such written instructions should also include the therapies that should be withheld, such as CPR. Finally, information about whom to call in the event of an emergency is extremely important.

Ideally, trained school personnel or a member of the interdisciplinary care team (such as a child-life worker, grief counselor, social worker, or nurse) should be available to support the child's schoolmates throughout the child's illness and after his or her death.

Funerals

Physicians may be asked about the advisability of allowing children to attend funerals. The best recommendation is to encourage children to attend if they want to go with the caveat that a designated, supportive adult first provide information about what a funeral is and what the child may see; that adult should be available to accompany the child to the funeral to provide support and to help the child leave, if needed. Adults can also describe other ways that children can say goodbye to the deceased if they choose not to attend the funeral. Regardless of the child's decision about attending the funeral, caregivers should answer his or her questions and provide emotional support.

After the death occurs, schools can help organize a memorial service to help schoolmates and members of the larger community achieve closure via a recognized venue for grieving and leave-taking, particularly for those not able to attend the funeral. Memorial services that include the child's family, classmates, families of classmates, teammates, friends, and others have the added value of providing support within the context of shared grieving. This approach facilitates continued discussion and support after the service concludes.

Ethical and Legal Issues

Article 12 from the United Nations Convention on the Rights of the Child states, "Parties shall assure to the child who is capable of forming his or her own views the right to express those views freely in all matters affecting the child, the views of the child being given due weight in accordance with the age and maturity of the child."[38]

Legally, parents are the final arbiters of what information will be shared with their child regardless of the child's capacity to make decisions. Practically, children vary in their capacity to express their wishes about the goals of medical care and usually need to rely on parents, caregivers, or other responsible adults to interpret their needs and wishes.[39] This responsibility can feel overwhelming for parents. However, good communication; realistic, factual information; and emotional support allow families to consider the goals that are possible, choose goals that are valued, and develop a plan of care that averts suffering to the maximum extent possible.

Baby Doe Regulations

The inherently difficult process of making treatment decisions for infants with poor prognoses is further hampered in the United States by widespread misunderstanding of the Baby Doe regulations.

The current Baby Doe regulations were enacted in 1984 and went into effect in 1985. They are funding requirements for states to receive child abuse funds that are regulated by the states and technically optional.

No criminal or professional sanctions are associated with the Baby Doe regulations. They are untested by the courts. The regulations require provision of life-sustaining treatment for all infants under the age of 1 year, regardless of projected quality of life, unless the infant is imminently dying or permanently comatose. In addition, when a life-limiting condition is irreversible, treatment is not required—an exception that many healthcare professionals fail to understand. The key portion states

[The withholding of medically indicated treatment is] the failure to respond to the infant's life-threatening conditions by providing treatment (including appropriate nutrition, hydration, and medication) which, in the treating physician's (or physicians') reasonable medical judgment, will be most likely to be effective in ameliorating or correcting all such conditions, except that the term does not include the failure to provide treatment (other than appropriate nutrition, hydration, or medication) to an infant when, in the treating physician's (or physicians') reasonable medical judgment any of the following circumstances apply: (i) The infant is chronically and irreversibly comatose; (ii) The provision of such treatment would merely prolong dying, not be effective in ameliorating or correcting all of the infant's life-threatening conditions, or otherwise be futile in terms of the survival of the infant; or (iii) The provision of such treatment would be virtually futile in terms of the survival of the infant and the treatment itself under such circumstances would be inhumane.[40]

Hospice and palliative care are appropriate alternatives to ineffective and burdensome life-extending treatments. Many prominent bioethicists and organizations specializing in pediatrics or medical and bioethics, such as the Hastings Center, believe the regulations are unethical.[41,42]

Decision-Making Capacity

Members of the interdisciplinary team can evaluate the child's decision-making capacity by providing developmentally appropriate explanations, options, and consequences and then asking the child to restate the information in his or her own words. The capacity to make healthcare decisions is assumed when individuals can restate such information. However, capacity is not an all-or-nothing phenomenon. A child may have capacity to make determinative (final) decisions about some issues but not others. Chronically ill children are widely acknowledged to have decision-making capacity regarding their illness and its treatment well beyond that expected of healthy peers. It is not unusual for an exceptionally mature understanding of illness to coexist with regressive behavior in other areas.

Capacity changes with time. A child's developmental understanding of personal death is an important factor to consider when involving the child in decisions about changing the goals of care from life prolongation to maximizing quality of life. The child's opinion will carry much greater weight if he or she understands that death is permanent.

Including Children in Decision Making

Caregivers must include a child in the decision-making process to the extent the child desires and is able to participate.[15,43,44] No other person can adequately present the child's view. It is incumbent on medical care providers to encourage the goal of honestly and compassionately sharing developmentally appropriate information with the child and his or her siblings, particularly in response to their questions. Caregivers should be aware of the known positive benefits of shared information and the significant problems associated with deception and coercion when disclosure is blocked.[17]

However, children rarely volunteer their feelings or conceptions of suffering. Moreover, children commonly defend their parents' denial even at the cost of increased suffering for themselves. They must be given permission to express their true feelings. Children suffer significantly when they feel abandoned and unable to trust the adults caring for them.

Regardless of age, children should be allowed to exercise discretion and autonomy consistent with their maturity and degree of understanding. Even toddlers can exert control over which hand will have the IV or which loved one will remain in the room during procedures. Older children can determine the timing of and need for conscious sedation and make other procedure-related decisions.

Emancipated Minors

Most states have passed statutes creating an emancipated-minor status based on financial independence or marital or military status; however, the right of emancipated minors to make medical decisions varies state by state.[45] The Mature Minor Doctrine[46] provides legal authority for children with decision-making capacity to make decisions about their medical care, but professional caregivers must be aware of the doctrine and be willing to implement it.

Research and Assent

The National Institutes of Health (NIH) and other research and institutional review boards (IRBs) require that children 7 years and older provide assent (identical to consent but without legally binding authority) to participate in experimental studies. Nevertheless, a child's wishes are often unsolicited or ignored despite the fact that many chronically ill children have remarkably mature views of illness and death as well as opinions about what medical care is valuable.[47,48] Discussions about treatment-related issues can be improved with age-appropriate communication and continued, open discussions with the child and family. As always, communication is an ongoing process, not a one-time event.

Presenting Benefits and Burdens

The availability of new technologies, such as transplants, extracorporeal membrane oxygenation (ECMO), and liquid ventilation can further confuse the decision-making process.[49,50] Parents and children cannot assess the relative benefits and burdens of such treatments because they lack experience with such interventions. Physicians too often emphasize the positive aspects of technical interventions, making it even more difficult for parents and children to make informed decisions. Physicians must present balanced information when discussing the likelihood and nature of hoped-for benefits and the known burdens of a treatment, including procedure-related pain, time spent in a hospital or physician's office, probable side effects of the treatment, and the treatment's likely impact on the child's length of life and the child's and family's quality of life. This information must be communicated in lay language with the use of diagrams and pie charts as needed to help parents and older children understand statistical information.

When patients and families are struggling with the decision-making process, the physician should recommend a course of action and explain why it is likely to

be the best option for their particular situation, keeping in mind the child's medical condition, the child's and family's known values, and the overall goals of treatment. Physicians can reasonably explain to families why their treatment options may be limited and refrain from offering interventions that will not benefit the child. The vocabulary physicians use when conveying options has a profound impact on the ultimate decision made and must be chosen carefully. Children should be included in discussions of treatment options whenever possible and appropriate, depending on their wishes and developmental stage.

Palliative care must be presented as an active, effective, equally valid treatment option and not as "doing nothing" or abandoning hope. Palliative care can be provided concurrently with curative or life-extending treatments, or it may be the sole focus of care.

Pediatric Pain Assessment

General Principles

Pain at the end of life is a distressing symptom commonly experienced by children dying of cancer, HIV/AIDS, and other complex conditions. However, pain can be very effectively treated with fairly simple measures for the majority of children, as evidenced by various pediatric studies.[51,52] Treating procedural pain and conditions known to be painful is a moral and ethical imperative regardless of the patient's age, communication or cognitive abilities, or position on the illness trajectory.[53,54] Despite the availability of effective interventions for preventing and treating pain and other symptoms, the provision of excellent pain and symptom management continues to be impeded by misconceptions about assessing symptoms in children.[55] Factual information and evidence can counteract these widespread myths.

- Assessment of pain in children must be tailored to the child's developmental stage.
- The gold standard of pain measurement is patient self-report, whether the patient is an adult or child. Standard measurement tools must be chosen according to the child's developmental capacity. As in the geriatric population, significant illness can cause a regression of skills and understanding. In contrast, some chronically ill children mature beyond their expected stage of cognitive development.
- To obtain honest reports of pain from children, healthcare providers must earn the trust of child, who may fear that painful or distressing interventions, such as blood tests or hospital admission, will be the result of acknowledging pain. Trust is achieved by urgent and effective response to reports of pain and other symptoms.
- Preventing and treating side effects of medical interventions is critical.
- It is important to use the child's own words for pain, such as "hurt," "boo-boo," or "owie."
- Physicians must ask the child about pain regularly and document the child's reports. It is critical to reassess the child's pain or other symptoms following an intervention (eg, giving an analgesic).
- Physiologic indicators, such as changes in pulse or blood pressure, may not be reliable indicators of pain in a chronically or critically ill child.
- Behavioral indicators, such as facial grimacing or crying, are unreliable and may be absent in the presence of chronic pain or critical illness.
- Children with pain may use play or sleep as coping mechanisms. These behaviors do not mean the child is free of pain.

FACT 1. Children, including neonates, feel pain and experience increased morbidity and mortality when inadequate analgesia is provided.

Children's perception of pain, including premature neonates, has been documented by studies that unequivocally demonstrate increasing stress responses, as measured by biochemical parameters, as well as increased morbidity and mortality in infants receiving inadequate analgesia perioperatively compared with those who receive adequate analgesia.[56-58] There is evidence that ineffective pain management early in life can have long-term consequences on subsequent pain responses. For example, neonates circumcised without analgesia experience more pain during routine immunizations, as measured by behavioral responses, when compared with children who receive appropriate analgesic interventions.[59] The concept that an infant's immature nervous system attenuates pain has not only been refuted, but evidence indicates that neonates actually experience increased sensitivity to pain because inhibitory pain tracts are the last to develop.[56]

FACT 2. Pain and other symptoms can be assessed effectively using developmentally appropriate tools.

Pain Assessment Tools

Although the experience of pain is critically influenced by contextual and emotional issues, this section focuses

on the physical assessment of symptoms. To achieve the greatest possible amelioration of distress, the multifactorial components of all symptoms must be addressed. Practitioners must assess the physical, psychological, and spiritual or existential (ie, what meaning the pain has for the child and family) components of pain and other symptoms.

Some clinical situations are so routinely associated with pain that its presence may be assumed in the noncommunicative patient, even in the absence of other signs. In these situations it is appropriate to provide empiric analgesia and then reassess changes in the child's behavior.

Physiologic parameters, along with other factors, can help indicate the presence of acute pain, for example, in the immediate postoperative period. However, physiologic parameters, including increased heart rate, diaphoresis, and facial grimacing, are not useful when assessing chronic pain in children because such signs attenuate markedly with time. These parameters also are unreliable when assessing pain in ill children, who may be experiencing tachycardia or increased respiratory rate for other reasons. Other criteria must be used.

Tools for Assessing Postoperative Pain in Neonates

The CRIES scale was designed to measure postoperative pain in neonates, but it can provide a useful guide for assessing pain from other causes in the neonatal population.[60] The scale consists of five physiologic and behavioral indicators, each of which can be rated as 0, 1, or 2. A maximum score of 10 is possible; analgesic administration is indicated for a score of 6 or greater. The name of the scale, CRIES, reminds clinicians to assess the following indicators:

- Crying
- Requires oxygen supplementation for normal oxygen saturation
- Increased vital signs (heart rate and blood pressure)
- Expression
- Sleeplessness

Behavioral Observation Tools for Assessing Pain in Very Young Children

Very young children, or those who are cognitively impaired or have regressed because of illness, frequently experience pain but may be unable to provide verbal self-reports of pain.[61] In such instances behavioral observations, preferably by someone who knows the child well, are used to assess pain. Examples of behavioral assessment tools include the CHEOPS (ie, Children's Hospital of Eastern Ontario Pain Scale) and the FLACC (ie, Faces, Legs, Activity, Cry, and Consolability) (**Table 4**). The FLACC was validated in a sample of 147 children younger than 3 years who were hospitalized with a variety of disease processes.[63] Each of the five behaviors is scored from 0 to 2; a higher score indicates more pain.

Table 5 describes the DEGR® scale developed by Gauvain-Piquard and colleagues to measure chronic pain in very young and nonverbal children.[64] The scale consists of 15 items in three categories, or dimensions, of behaviors: 6 items specific to pain, 5 related to psychomotor inertia, and 4 related to anxiety. Each item is scored from 0 to 4, with a total possible score of 60. The scale was validated in a sample of 152 child cancer patients between the ages of 2 and 6 years for measuring pain of long duration rather than acute postoperative or procedural pain. The cutoff score was fixed at 12 to maximize the sensitivity of diagnosing pain in this population.

Self-Report Tools for Children at the Developmental Age of 3 to 6 Years

Pain measurement scales are helpful for providing the child, family, and serial caregivers with a common language to report pain and the efficacy of interventions. Some children may refuse to use scales because they dislike them, are tired of being asked about their pain, or do not understand the concept of pain assessment. In such cases, staff should use alternative methods to assess and reassess pain and other symptoms in ways that are acceptable to the child.

Hester's Poker Chips[65] are generally used at a developmental age of 3 years or older. Four poker chips are presented to the child, then the assessor says, "One chip is for a small or little amount of pain. Four chips are for the most pain (or "owie" or "boo-boo", etc.) that you

Table 4. FLACC: Pain Assessment Tool

	Scoring		
Categories	**0**	**1**	**2**
Face	No particular expression or smile	Occasional grimace or frown, withdrawn, disinterested	Frequent to constant frown, quivering chin, clenched jaw
Legs	Normal position or relaxed	Uneasy, restless, tense	Kicking or legs drawn up
Activity	Lying quietly, normal position, moves easily	Squirming, shifting back and forth, tense	Arched, rigid, or jerking
Cry	No cry (awake or asleep)	Moans or whimpers; occasional complaint	Crying steadily, screams or sobs, frequent complaints
Consolability	Content, relaxed	Reassured by occasional touching, hugging, or being talked to; distractible	Difficult to console or comfort

From Pediatric Nursing, 23*(3)*, Merkel, Voepel-Lewis, Shayevitz, & Malviya, The FLACC: A behavioral scale for scoring postoperative pain in young children, © 1997 Jannetti Publications, Inc. Reprinted with permission.[62]

have or could think of having." The child is then asked to place the number of chips in the assessor's hand that corresponds to the amount of pain that he is having now. Any four small objects of uniform color, shape, and size can be used as alternatives to poker chips.

Tools appropriate for use with children younger than 7 years feature limited numbers of choices (generally fewer than six) and concrete descriptors such as faces. Numerical descriptors alone are too abstract. The pictures can be photographs or drawings of faces showing expressions of increasing distress. The Oucher Scale uses photographs and is available in multiethnic representations.[66] Some children may have experience with and prefer to continue using a particular measurement tool. Whatever the tool chosen, it should be used consistently with that particular child.

See **Figure 4** for the Bieri Faces Scale, a validated and well-tested faces scale.[67] When using the Bieri scale or another faces scale, it is helpful to anchor the endpoints of the scale by saying to the child, "This face shows someone who has no pain and this face shows someone with the worst pain that you have had or can imagine." The child is then told, "Point to the face that shows how much pain (or "owie" or "boo-boo", etc.) you are having."

Self-Report by Children at the Developmental Age of 7 Years and Older

At about the developmental age of 7 years and older, children can use a standard 0 to 10 numeric scale. However, if the child is accustomed to using a simpler scale, there is no reason to abandon it. Some practitioners prefer to use a 0 to 5 scale. It is critically important to document the maximum number on the scale being used; a 5/5 symptom distress score demands more urgent attention than a 5/10 score.

Table 5. DEGR® Scale*

This scale consists of 15 items in three dimensions: 6 behaviors specific to pain, 5 psychomotor inertia items, and 4 anxiety items. The item numbering refers to their order in the scale. Each item is scored from 0 (none) to 4 (severe), with a total possible score of 60. A score higher than 12 indicates pain.

Behaviors Specific to Pain

Item 2. Unnatural postures: The child avoids certain painful positions or adopts a particular position to relieve a painful area. This item should be studied when the child is sitting or lying down with no physical activity. It should not be confused with the antalgic position during movement.

Item 4. Protection of painful areas: The child seems to continuously avoid all contact with painful areas.

Item 6. Expressing pain: This item concerns the way in which the child says that he is in pain, either spontaneously or when asked, during the period of observation.

Item 8. Indicating painful areas: The child locates his pain, either spontaneously or when asked.

Item 10. Pain avoidance when moving: The child spontaneously avoids all movement or tries not to move part of his body. To be scored during sequences of movements (eg, walking), possibly induced. Motor slowing should not be noted here.

Item 14. Reactions to examination of the painful area: When a painful area is examined, the child resists, pulls away, or reacts emotionally. Only the child's reactions to the examination should be noted and not any previous reactions.

Psychomotor Inertia

Item 3. Resignation: The child is resigned to everything that happens to him. He does not try to protest or resist. Should be scored during an unpleasant situation, such as venipuncture.

Item 5. Withdrawal: The child sometimes "withdraws into his shell."

Item 7. Lack of expression: Concerns the ability of the child to register and express feelings by his tone of voice, eyes, and facial expression. Should be scored when the child is active (eg, during games, meals, and chatter)

Item 11. Lack of interest in surroundings: Concerns the child's available energy for interaction with his environment.

Item 13. Slowness and paucity of movement: The child's movements are slow, restricted, and rather stiff, even some distance away from painful areas. The trunk and large joints are particularly motionless. Should be scored in relation to a normal child's movements.

Anxiety Behaviors

Item 1. Tenseness: Concerns the degree of nervous tension in the child's body.

Item 9. Hostility:† Measures the child's hostility toward those around him.

Item 12. Wariness at being moved: When the child is moved for a meal, bath, or the like, he is wary, says or shows how he wants to be moved, resists or holds onto the adult's hand.

Item 15. Crying easily: Concerns the extent to which the child cries to express himself.

Comments

***This scale can also be used to assess pain in adult patients with dementia or delirium.*

† Irritability may be a more appropriate descriptive word for this behavior.

From European Journal of Pain, 3*(2), Gauvain-Piquard A, Rodary C, Rezvani A, Serbouti S. The development of the DEGR®: a scale to assess pain in young children, 165-176.[64] © 1999 Elsevier. Reprinted with permission.*

Figure 4. Faces Pain Scale—Revised (FPS-R)
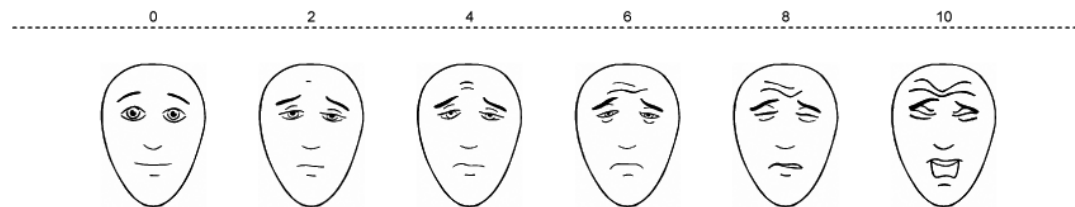
FPS Instructions: In the following instructions, say "hurt" or "pain," whichever seems right for a particular child. "These faces show how much something can hurt. This face [point to left-most face] shows no pain. The faces show more and more pain [point to each from left to right] up to this one [point to right-most face—it shows very much pain. Point to the face that shows how much you hurt [right now]." Score the chosen face 0, 2, 4, 6, 8, or 10, counting left to right, so "0" = "no pain" and "10" = "very much pain." Do not use words like "happy" and "sad". This scale is intended to measure how children feel inside, not how their face looks.

From Hicks CL, von Baeyer CL, Spafford P, van Korlaar I, Goodenough B. The Faces Pain Scale—Revised: toward a common metric in pediatric pain measurement. Pain. 2001;93:173-183. Bieri D, Reeve R, Champion GD, Addicoat L, Ziegler J. The Faces Pain Scale for the self-assessment of the severity of pain experienced by children: development, initial validation and preliminary investigation for ratio scale properties. Pain. 1990;41:139-150. © 2001 International Association for the Study of Pain (IASP). Reprinted with permission.

CLINICAL SITUATION

Tony

Tony is a nonverbal, developmentally delayed 18-month-old infant with HIV encephalopathy and markedly increased tone and irritability. The nurses report that Tony is inconsolable and seems worse with attempts at comforting with physical measures, such as swaddling, rocking, gentle stroking, and being given a pacifier. He was fed 1 hour ago and has had normal bowel movements.

Question

Which of the following would you use to assess whether Tony is experiencing pain? (Choose all that apply.)

A. Ask one of his caregivers to describe Tony's usual indicators of pain, as distinguished from indicators of hunger or frustration.

B. Assess Tony's behavior before and after an analgesic trial.

C. Observe Tony's posture during maneuvers, such as bathing and diaper changes.

D. Try to match Tony's expression with those on a faces scale.

Correct Response and Analysis

The correct responses are A, B, and C. Use of the DEGR® scale would be appropriate in this situation. The faces scales are tools for self-assessment of pain. Correct use of a faces scale involves a verbal child choosing the face that best represents the intensity of his pain, not a caregiver's attempt to match the child's expression to a face.

Gina: Pain and Symptom Management in a 5-Year-Old Child

Gina, a developmentally normal 5 year old, has a relapse of her leukemia. She refuses to walk and is irritable, despite being given 15 mg/kg of acetaminophen PO every 4 hours.

Question One

How would you assess whether Gina has pain? (Choose the best response.)

A. Ask one of her parents.
B. Move her to see if you can reproduce the pain.
C. Using a scale, ask her to indicate the amount of pain that she has.
D. Look at her facial expression for signs of pain.

Correct Response and Analysis

The correct response is C. Patient self-report is the gold standard for pediatric pain measurement. However, it is helpful to use all sources of information, including family members' perceptions and facial expressions or other body language.

Question Two

Which of the following pain assessment tools could be used for Gina? (Choose all that apply.)

A. A 0 to 10 numeric scale, as in the adult population
B. A faces scale, such as the Bieri or Oucher Scale
C. A behavioral observation scale
D. Hester Poker Chips

Correct Response and Analysis

The correct answers are B and D. The observation scales are used for infants and nonverbal children. The 1 to 10 scale is too abstract for a 5 year old.

Pediatric Pain Management

Factual information can help to counteract misconceptions that impede best practice.[55] The basic principles of pharmacologic pain management for adults are also applicable to children. Variations exist, for example, calculating the initial starting dosage based on weight; however, the principles of pain assessment, dosage titration, and anticipating and managing side effects are the same.

FACT 1. The risks of respiratory depression and other opioid-induced side effects are generally no more frequent or profound in children than adults.

The incidence of significant opioid-induced respiratory depression and other opioid-induced side effects in children older than 3 months is no greater than that for adults, at a rate of .09%.[68,69] Infants younger than 6 months of age are initially dosed at 25% to 33% of the usual mg/kg dosage used for older, opioid-naive children. Subsequent titration to symptom relief can provide effective and safe pain control when accompanied by careful monitoring of level of consciousness and respiratory rate. As with adults, other potential side effects, such as constipation or pruritus, should be anticipated and treated. Concern about possible side effects from the use of opioids should not prevent adequate treatment of pain.

Children may be less willing to continue taking a medication that causes temporary discomfort even if they are experiencing pain relief. Side effects must be anticipated and managed aggressively.

FACT 2. There is no evidence that treatment of pain with opioids causes addiction in children.

Children and adults suffer needlessly because of misconceptions about addiction, tolerance, and dependence. The misconceptions are so common that they must be proactively addressed and reviewed with all healthcare professionals, patients, and families.

Addiction is a primary, chronic, neurobiologic disease, with genetic, psychosocial, and environmental factors influencing its development and manifestations. It is characterized by behaviors that include one or more of the following: impaired control over drug use, compulsive use, continued use despite harm, and craving.[70] Addiction to opioids is very uncommon with individuals being treated with opioids for pain relief.[71-73] There is evidence that increased prescribing of opioids for pain actually decreases the incidence of addiction to opioids.[74] It is important to reassure patients with a previous or current history of substance abuse that their pain will still be addressed and managed effectively. Close monitoring is also essential as addiction to cocaine or alcohol increases the risk of opioid abuse.[75]

When a patient with addiction requires pain treatments with opioid analgesics, a contract between the patient and the care provider can help delineate mutual expectations, which should be discussed and respected.[76] Contracts can include conditions such as the following:

- All prescriptions will be written by one individual and filled by one pharmacy
- No allowance will be made for "lost" prescriptions or weekend or evening refills.

The difficult but manageable situation of treating pain for patients with addiction is well delineated in the reference *Substance Abuse Issues in Cancer Patients*.[77,78] Health professionals caring for children of families with histories of substance abuse report the existence of an unwritten code of ethics that indicates that the family member with the history of substance abuse will not use their child's analgesics for themselves.[79] However, a written contract may also be beneficial in this situation.

Physiologic dependence is a state of adaptation that is manifested by a drug-class-specific withdrawal syndrome that can be produced by abrupt cessation, rapid dose reduction, decreasing blood level of the drug, or administration of an antagonist.[70] Withdrawal may be accompanied by psychological distress. Withdrawal from opioids is easily avoided by gradually tapering the dos-

age downward, once the indication for the medication has resolved, while monitoring the patient for symptoms of withdrawal, notably, for some or all of the following: anxiety, tachycardia, sweating, rhinorrhea, and diarrhea.

Tolerance is a state of adaptation during which exposure to a drug induces changes that result in a diminution of one or more of the drug's effects over time.[70] Evidence indicates that the most common cause of increased dosage requirements to control pain is advancing disease, not tolerance. After stable pain relief is achieved, patients generally can be managed with regular doses of oral, long-acting medication until the disease advances. When patients with chronic pain develop tolerance, it can be managed by increasing the opioid dosage or changing to a different opioid (opioid rotation).[80] Because of concerns about tolerance, many physicians save the use of opioids for the very end of a patient's life. The presence of unrelieved pain, not the patient's prognosis, should determine when opioids are used.

FACT 3. Medications to effectively manage pain in children are available.

Ninety percent of pain in children can be controlled with systemic opioids and adjuvant analgesics without using extraordinary opioid dosages or invasive procedures, such as epidural or subarachnoid analgesic administration.[51,52] Extraordinary opioid requirements refer to dosages in the range of 100 times the usual postoperative infusion of 30 mcg/kg/h morphine equivalent. When required, invasive procedures can be used to control most pain that cannot be managed otherwise. In very rare cases, sedation at the end of life can be used to manage severe refractory pain or other distressing symptoms, such as dyspnea.[81-83]

For more in-depth discussions of pediatric pain management, see references 74, 77-82. *Pain, Pain, Go Away: Helping Children with Pain* is a helpful resource on pain management for family members of a child with cancer.[84]

For more information on pain assessment and management in adult populations, see UNIPAC 3.

General Principles of Pain Management in Children

- Diagnostic procedures and medical therapies, including IV insertions, continue to be a significant cause of pain experienced by children with complex, chronic conditions.
- Children with pain at the time of a cancer diagnosis generally experience excellent pain resolution with initiation of treatment for the disease (eg, chemotherapy and radiation).[85] Disease-associated pain can be a prominent feature during recurrence and the terminal phase of cancer.[3]
- Other conditions, such as HIV/AIDS, traumatic injury, and severe cerebral palsy, may cause pain throughout the course of care.
- Ongoing pain should be treated with regular, scheduled, around-the-clock medications.
- Breakthrough pain (sudden increases in pain, escalating pain, or movement-related pain [incident pain]) should be anticipated. Dosages of 5% to 15% of the total daily dosage of an opioid offered every 1 to 2 hours are recommended.
- Pain relief is achievable without having to resort to "extraordinary" dosages or routes of administration in at least 90% of children with disease-related pain due to advanced cancer.[51,86]
- Nonsteroidal antiinflammatory drugs (NSAIDs)* are generally avoided in children with low platelet counts, such as those with cancer or HIV/AIDS.[84]
- Pharmacologic management of pain should always be used in combination with nonpharmacologic therapies.
- Newborns have been shown to experience decreased morbidity and mortality when given adequate and appropriate analgesic therapy.[54]

Editors' note: The US Food and Drug Administration (FDA) has added "black box" warnings to several medications commonly used by hospice and palliative care practitioners. "Black box" warnings are designed to highlight the potential for rare but serious medical complications such as stroke or myocardial infarction associated with the use of these drugs. Hospice and palliative care practitioners should be aware of these FDA "black box" warnings and the risk-benefit ratio of these medications and alternatives for an individual patient. The short prognosis and critical importance of symptom relief for many hospice and palliative care patients may justify the use of these medications despite such risks.

- Many of the medications used for pain and symptom control are not well studied in the pediatric population.

Nonpharmacologic Management of Pain and Other Symptoms

Nonpharmacologic interventions, including physical measures and cognitive and behavioral techniques, can alleviate physical and psychological distress in children with life-limiting illnesses. Guided imagery or hypnotherapy can decrease breakthrough pain or breathlessness when waiting for pharmacologic agents to take effect. Distraction and relaxation can help both patients and caregivers maintain a sense of control and decrease the perceived intensity of symptom exacerbations. Physical techniques (eg, massage, therapeutic touch, acupressure and acupuncture) are effective for some patients. Such measures become increasingly important and meaningful to patients and family members as other interventions and monitoring are discontinued. However, nonpharmacologic interventions should be accompanied by effective pharmacologic therapies when indicated and desired by the patient. (See page 68, Preventing and Managing Procedure-Related Pain.)

Pharmacologic Management of Pain

Analgesics for Mild Pain

For mild pain, oral medications such as acetaminophen or ibuprofen can be used (see **Table 6**). Nonopioid analgesics are effective for relieving mild pain; however, a "ceiling effect" limits their usefulness. Dosages higher than those recommended increase the risk of toxicity without any increase in analgesic benefit. Acetaminophen may cause liver toxicity and NSAIDs cause platelet dysfunction, gastritis, and renal toxicity. Ibuprofen can interfere with the antiplatelet effects of low-dose aspirin.[87]

Analgesics for Mild-to-Moderate Pain

For mild-to-moderate pain, an oral opioid such as codeine can be added to the acetaminophen or ibuprofen or used alone. The dosage range for codeine is 0.5 to 1.0 mg/kg/dose every 4 hours. Dosages higher than 1.0 mg/kg contribute to an increased side-effect profile with no substantial increase in pain relief. If pain relief is inadequate using a 1.0 mg/kg/dose of codeine, it should be discontinued and an opioid for moderate-to-severe pain should be started. Alternatives include hydrocodone, 0.1 to 0.2 mg/kg/dose every 4 hours (available in the United States only in a fixed-dose combination with acetaminophen or ibuprofen), or oxycodone, 0.1 mg/kg/dose every 4 hours (available as a single agent or in a fixed-dose combination with acetaminophen or ibuprofen).

When using fixed-dose combinations, it is important to monitor the total dosage of the nonopioid medication. If the maximum allowable dosage does not achieve effective pain relief, switching to a pure opioid with a separate dosage of an NSAID or acetaminophen is the recommended approach. Concurrent administration of an opioid and an NSAID is often more effective than using an opioid alone. However, this approach requires evaluation of the child's willingness to take multiple medications. Some clinicians prefer to use a single-agent opioid when children present with moderate pain to allow for adequate dose titration.

Tramadol has been used effectively for dental procedures for patients between 4 and 7 years old,[88] and, apart from a rapid rise in serum concentrations, appears to have similar pharmacokinetics to those seen in healthy adults.[89]

Analgesics for Moderate-to-Severe Pain

If a child has no previous adverse experience with morphine, it is the opioid of choice for moderate-to-severe pain. Most physicians are familiar with morphine, it is inexpensive, and it is available in a variety of formulations. Alternatives to morphine include hydromorphone, oxycodone, oxymorphone, fentanyl, and methadone. Morphine and oxycodone are available in both immediate-release and sustained-release oral preparations. Fentanyl is available as parenteral, transdermal, or transmucosal preparations. Methadone is a long-acting opioid available in a liquid preparation, which makes it especially useful for children. When opioid side effects cannot be managed or become intolerable, rotating opioids (changing from one opioid to another) is recommended.

Table 6. Pediatric Palliative Medicine Formulary

Class and Drug	Route	Pediatric Dosage (mg/kg/dose, unless otherwise stated)	Adolescent Dosage (mg/dose)	Interval (hours)	Maximum Dosage	Comments
Nonopioid Analgesics (fever, mild pain)						
Acetaminophen	PO	10-15	650-1,000	4-6	75 mg/kg/day or 4 g/day	
	PR	Loading, 40-45; then 10-15	650-1,000	4-6		Rectal absorption variable
Ibuprofen	PO	5-10	200-800	6-8	3.2 g/day	Antiinflammatory
Naproxen	PO	5-10	250-500	8-12	1,250 mg/day	Antiinflammatory
						Older than 2 years
Choline magnesium trisalicylate	PO	7.5-15	500-1,500	6-8	60 mg/kg/day	Antiinflammatory
						Avoid in varicella or flu-like illness; risk of Reye's syndrome
Aspirin	PO	10-15	650-1,000	4	60-80 mg/kg/day	Antiinflammatory
					4 g/day	Avoid in varicella or flu-like illness; risk of Reye's syndrome
Opioid Analgesics (moderate-to-severe pain, dyspnea; titrate dosage to effect; for infants younger than 6 months, start at ¼ to ⅓ of recommended dosage)						
Codeine	PO	0.5-1	15-60	4-6	2-6 years: 30 mg/day 6-12 years: 60 mg/day	
Hydrocodone	PO	0.1-0.2	5-10	4-6	6 doses/day because of ceiling of coformulated drug	Available only in fixed-dose combinations FDA rule 10/1/07 removes unapproved preparations from market
Oxycodone	PO, SL	0.05-0.2	5-10	3-6		

Table 6. Pediatric Palliative Medicine Formulary

Class and Drug	Route	Pediatric Dosage (mg/kg/dose, unless otherwise stated)	Adolescent Dosage (mg/dose)	Interval (hours)	Maximum Dosage	Comments
Morphine	PO, SL	0.15-0.3	5-10	3-4		Dosage is for immediate-release preparations only; sustained-release dosing is determined based on stable chronic 24-hour requirement
	IV, SC	0.05-0.1	2-5	2-4		
	Infusion	0.03 mg/kg/hour	1.5 mg/hour	Continuous		
Hydromorphone	PO	0.03-0.08	1-4	3-4		Less pruritis than morphine
	IV, SC	0.015-0.07	1-2	3-4		IV solution may contain benzyl alcohol and/or parabens; do not use in neonates
	Infusion	0.006 mg/kg/hour	0.3 mg/hour	Continuous		
Fentanyl	IV	0.5-2 mcg/kg	25-75 mcg	30 minutes		Caution: Rapid infusion may cause chest wall rigidity.
	Infusion	1 mcg/kg/hour	25-100 mcg/hour	Continuous		
	Transdermal	See comment	See comment			Lowest-dose patch 12.5 mcg/hour
						Dose selection based on chronic stable opioid requirement and equianalgesic potency (tolerant to minimum 60 mg oral morphine equivalent per day)
						Age greater than 2 years
	Oral transmucosal	5-15 mcg/kg			400 mcg/dose	For procedure-related pain
Methadone	PO	0.1-0.2	5-10	Initial doses, 4-6; then 6-12		Long-acting opioid available in oral liquid form
	IV	0.1	5-8	4-8		Titrate slowly because of accumulation and delayed sedation

Table 6. Pediatric Palliative Medicine Formulary

Class and Drug	Route	Pediatric Dosage (mg/kg/dose, unless otherwise stated)	Adolescent Dosage (mg/dose)	Interval (hours)	Maximum Dosage	Comments
Local and Topical Anesthetics (procedural pain, adjuvant analgesic)						
Lidocaine with bicarbonate (buffered)	SC infiltration				3 mg/kg lidocaine	
Lidocaine 2.5% and prilocaine 2.5% cream (EMLA)	Topical	1 g/10 cm^2	2 g/10 cm^2	30-60 minutes before procedure	**0-2 months:** 1 g × 1 hour **3-11 months:** 2 g × 4 hours **1-5 years:** 10 g × 4 hours **6-11 years:** 20 g × 4 hours	Apply to intact skin; cover with occlusive dressing Methemoglobinemia risk in infants younger than 3 months, premature infants, prolonged exposure time, G6PD deficiency
Lidocaine cream 4% (ElaMax;LMX4) 5% (LMX5)	Topical	Apply to area smaller than max for weight			**Weight < 10 kg:** 100 cm^2 **10-20 kg:** 200 cm^2	
Lidocaine 5% extended-release patch	Topical	N/A	1-3 patches on for 12 hours	12 hours per day	Apply for only 12 hours out of 24-hour period	Patch size may be cut prior to removal of release liner
Lidocaine 70mg/ tetracaine 70mg extended-release patch	Topical	1 patch	1 patch	20-30 minutes before procedure	1 patch x 30 minutes	Older than 3 years Apply to intact skin Do not cut patch Used patch retains 90% of drug; dispose properly to prevent ingestion by children or pets
Tetracaine gel 4%	Topical	1 g	1 g	30 minutes before procedure		Not available in the US Less pain with IV start compared to EMLA With or without occlusive dressing

Table 6. Pediatric Palliative Medicine Formulary

Class and Drug	Route	Pediatric Dosage (mg/kg/dose, unless otherwise stated)	Adolescent Dosage (mg/dose)	Interval (hours)	Maximum Dosage	Comments
Capsaicin	Topical	0.025% 0.075%	0.025% 0.075%	Three to four times daily		Initial discomfort (burning) when applied; consider preapplication of topical lidocaine preparation
Benzodiazepines (agitation, anxiety, seizures, insomnia)						
Clonazepam	PO	0.01	0.25-0.5	8-12	0.1-0.2 mg/kg/day > 10 years: 20 mg/day	Long-acting Titrate by 10%-25% every 2 or 3 days For myoclonus or anxiety
Diazepam	PO	0.04-0.2	2-10	6-12	**< 5 years:** 5 mg/dose	Medium-acting
	IV	0.04-0.2	2-10	2-4	**> 5 years:** 10 mg/dose	
	IV	Seizure: 0.2-0.5	5-10	May repeat in 15 minutes		
	PR	0.5 (2-5 years old) 0.3 (6-11 years old)	0.2 mg/kg/dose	May repeat in 4-12 hours		Onset of action: 2-10 minutes IV preparation may be given PR; additional dose of 0.25 mg/kg in 10 minutes if needed Rectal gel formulation—round dose to nearest available dosage strength Suppositories may be compounded by pharmacy

continued

Table 6. Pediatric Palliative Medicine Formulary

Class and Drug	Route	Pediatric Dosage (mg/kg/dose, unless otherwise stated)	Adolescent Dosage (mg/dose)	Interval (hours)	Maximum Dosage	Comments
Lorazepam	PO, SL, PR	0.02-0.05	0.5-4	4-12	4 mg/dose	Medium-acting
	IV	0.05-0.1	0.5-1.5	Seizure: may repeat in 10-15 minutes	4 mg/dose	Useful for anticipatory nausea
Midazolam	PO	0.25-1	10-20	Single dose	20 mg/dose	Short-acting
	IV	0.025-0.1	0.5-2	May repeat dose in 2-3-minute intervals	10 mg *total* dose	Sedation for procedures—monitor appropriately Administer IV dose over 2-3 minutes
Prokinetic Antiemetic						
Metoclopramide	PO	0.1-0.5	5-15	6	0.8 mg/kg/day; 15 mg/dose	For nausea from gastic stasis or opioids.
	IV, SC	0.1-0.5	10-20	4-6		Chemotherapy antiemetic dose 1-2 mg/kg/dose; premedicate with diphenhydramine 1.0 mg/kg Well tolerated PO, SC, IV
Antihistamines (nausea, vomiting, pruritis)						
Diphenhydramine	PO, IV	0.5-1	10-50	4-8	100 mg/dose	Can relieve dystonia from phenothiazines or haloperidol
Hydroxyzine	PO, IV	0.5-1	25-100	4-6	600 mg/day	

Table 6. Pediatric Palliative Medicine Formulary

Class and Drug	Route	Pediatric Dosage (mg/kg/dose, unless otherwise stated)	Adolescent Dosage (mg/dose)	Interval (hours)	Maximum Dosage	Comments
Promethazine	PO, PR, IV	0.25-1	12.5-25	4-6		Age older than 2 years
						Like other phenothiazines, can cause dystonia
						Potency IV=PO=PR
Neuroleptics (nausea, vomiting, delirium)						
Haloperidol	PO, IV, SC	0.01-0.05	0.5-5	4-12	0.15 mg/kg/day	Age older than 3 years
						For delirium or nausea
						Watch for dystonia
Haloperidol	IM	1-3 mg (6-12 years)	2-5	4-8		IM special circumstances only
Chlorpromazine	PO	0.5-1	10-25	4-8	**< 5 years:** 50 mg/day	
	IV, IM	0.5-1	25-50	4-8	**5-12 years:** 75 mg/day	
	PR	1	50-100	6-8		
Prochlorperazine	PO	0.1-0.5	5-10	6-8	15 mg per day	Age older than 2 years
	PR	0.2-0.5	25	8-12		
Risperidone	PO	Do not use	0.25	12		Age older than 9 years
Serotonin (5-HT3) Receptor Antagonists (nausea, vomiting)						
Ondansetron	PO	Body surface area: **< 0.3 m²:** 1 mg/dose **0.3-0.6 m²:** 2 mg/dose **0.6-1 m²:** 3 mg/dose **> 1 m²:** 4-8 mg/dose	4-8	6-8		Generic available Alternative dose by age: 4-11 years, use 4 mg three times daily; 12 years and older, use 8 mg three times daily

Table 6. Pediatric Palliative Medicine Formulary

Class and Drug	Route	Pediatric Dosage (mg/kg/dose, unless otherwise stated)	Adolescent Dosage (mg/dose)	Interval (hours)	Maximum Dosage	Comments
	IV	0.15	0.15	6-12		For highly emetogenic chemo, give 0.45 mg/kg/dose up to max 32 mg/dose 30 minutes prior
Granisetron	PO	0.04	1-2	12-24	2 mg/day	Older than 4 years
	IV	0.01-0.02	0.01-0.02 mg/kg/dose	8	2 mg/day	Older than 2 years
Dolasetron	PO	1.2-1.8	100	Single dose	100 mg/dose	
	IV	0.35-1.8	12.5-100	Single dose	100 mg/dose	
Corticosteroids (antiinflammatory, nausea, vomiting, pain, anorexia)						
Dexamethasone	PO, IV, SC	**Antiinflammatory:** 0.08-0.3 mg/kg/*day*	0.5-9 mg/day	6-12		For nausea, anorexia, or pain from intra-cranial pressure or hepatic distension
		CNS edema: 1-2 mg/kg/dose, then 1-2 mg/kg/day				Watch for mood swings, psychosis, or long-term side effects.
Methylprednisolone	PO, IV	0.5-2 mg/kg/day	2-60	12-24	60 mg/day	
Prednisone	PO	0.5-2 mg/kg/day	5-60	12-24	80 mg/day	
Anticholinergics (secretions)						
Scopalamine	Transdermal	Older than 6 years: ½ patch	1 patch (delivers 1 mg over 72 hours)	Every 3 days		Place behind ear Movement-related nausea Rattling secretions
	IV, SC	0.006	0.2-1	6-8		

Table 6. Pediatric Palliative Medicine Formulary

Class and Drug	Route	Pediatric Dosage (mg/kg/dose, unless otherwise stated)	Adolescent Dosage (mg/dose)	Interval (hours)	Maximum Dosage	Comments
Hyoscyamine	PO, SL	2-12 years old: 0.0625-0.125 mg/dose	0.125-0.25 mg/dose	4	2-12 years old: 0.75 mg/day	For troublesome secretions
					> 12 years old: 1.5 mg/day	
Glycopyrrolate	PO	0.04-0.1	1-2	4-8	Max 1-2 mg/dose, 8 mg per day	For troublesome secretions
	IV, SC	0.004 – 0.01	0.01 mg/kg	4-8		
Psychostimulants (opioid sedation, depression)						
Methylphenidate	PO	0.1 mg/kg/dose or	5-10	Morning and noon	0.5 mg/kg/day	Can exacerbate anxiety or agitation
		2.5-5 mg *dose*			60 mg/day	
Dextroamphetamine	PO	2.5-5 mg *dose*	5-10	1-2 doses per day	40 mg per day	Same indications and cautions as above
Tricyclic Antidepressants (neuropathic pain, depression)						
Amitriptyline	PO	0.1-2	10-50	Single or divided doses	5 mg/kg/day	Older than 6 years
					200 mg/day	Antidepressant dose higher than analgesic dose
						Increase dose gradually every few days
						Most anticholinergic
Desipramine	PO	1-5 mg/kg/day	25-100 mg/day	Single or divided doses	150 mg/day	Older than 6 years
						Increase dose gradually every few days

continued

Table 6. Pediatric Palliative Medicine Formulary

Class and Drug	Route	Pediatric Dosage (mg/kg/dose, unless otherwise stated)	Adolescent Dosage (mg/dose)	Interval (hours)	Maximum Dosage	Comments
Nortriptyline	PO	1-3 mg/kg/day	1-3 mg/kg/day	Divided doses	**Child:** 10-20 mg/day	Older than 6 years
					Adolescent: 30-50 mg/day	
					Adult: 75-100 mg/day	
Anticonvulsants (seizures, neuropathic pain)						
Phenytoin	PO	5-10 mg/kg/day	100	Two to three times daily	600 mg/day	
	IV-loading	Seizure: load 15-20	Seizure: load 10-15 mg/kg		1000 mg/day	IV rate max 1 mg/kg/min (ie, 15-20 minutes)
						Useful in pain crisis
	IV	5-10 mg/kg/day	100	Two to three times daily	600 mg/day	
Fosphenytoin	IV	Seizure: load 10-20 (PE)	Seizure: load 10-20 mg/kg (PE)			Dose calculated in phenytoin equivalents (PE)
	IV, IM	N/A	4-6 mg/kg (PE)	Three times daily		IV rate max 3 mg/kg/min (ie, 5-7 minutes)
Carbamazepine	PO	**< 6 years:** 10-35 mg/kg/day	100-200	Twice daily	**6-15 years:** 1,000 mg/day	Increase dose gradually
						Watch for marrow suppression
		6-12 years: 10-30 mg/kg/day			**> 15 years:** 1,200 mg/day	Suspension should be divided four times daily

Table 6. Pediatric Palliative Medicine Formulary

Class and Drug	Route	Pediatric Dosage (mg/kg/dose, unless otherwise stated)	Adolescent Dosage (mg/dose)	Interval (hours)	Maximum Dosage	Comments
Gabapentin	PO	3-5 (initial)	300 (initial)	Three times daily (see comment)	300-mg initial dose 3,600 mg/day	First 3 days: give one dose on first day, 2 doses on second day, 3 doses on third day (maintain)
Phenobarbital	PO, PR, IV	**Neonate:** 3-5 mg/kg/day **Infant:** 5-6 mg/kg/day **1-5 years:** 6-8 mg/kg/day **6-12 years:** 4-6 mg/kg/day	> 12 years old: 1-3 mg/kg/day *or* 10-100 mg/dose	Once or twice daily		
	IV	Seizure: load 15-20 mg/kg/dose; additional 5 mg/kg/dose every 15-30 minutes to max of 30 mg/kg	Seizure: load 15-20 mg/kg/dose; additional 5 mg/kg/dose every 15-30 minutes to max of 30 mg/kg		30 mg/kg/dose	
Cholinergic Agent (urinary retention)						
Bethanechol	PO	0.15-0.2	10-50	Repeat hourly until void, then three to four times daily	50 mg total dose (initial titration)	For urinary retention Watch for nausea

Table 6. Pediatric Palliative Medicine Formulary

Class and Drug	Route	Pediatric Dosage (mg/kg/dose, unless otherwise stated)	Adolescent Dosage (mg/dose)	Interval (hours)	Maximum Dosage	Comments
	SC	0.03-0.04	2.5-10	Repeat every 15-30 minutes until void, then three to four times daily	10 mg total dose (initial titration)	
Miscellaneous						
Octreotide	SC,IV	0.001-0.01 mg/kg/day	0.1-0.6 mg/day	Continuous or two to three times daily		Antisecretory agent for bowel obstruction
						Titrate up by 0.0003 mg/kg every 3 days
						Limited data in children
Baclofen	PO	3-5 mg dose	5	Three times daily	40 mg/day (2-8 years)	Age older than 2 years
					60 mg/day (8-80 mg/day (adult)	Titrate dose up every 3 days
						For nerve pain or spasticity
						For intrathecal route, consult specialty pain service
						Watch for withdrawal symptoms—seizure, anxiety
Mexiletine	PO	1	200-300	Three times daily	900 mg/day	Adjuvant analgesic
						Increase by 1 mg/kg every 2-3 days as tolerated to relief
						Monitor blood levels, EKG
						Case report: 75 mg once daily, increased over 3 weeks to 150 mg dose three times daily

Table 6. Pediatric Palliative Medicine Formulary

Class and Drug	Route	Pediatric Dosage (mg/kg/dose, unless otherwise stated)	Adolescent Dosage (mg/dose)	Interval (hours)	Maximum Dosage	Comments
Biphosphonates (hypercalcemia, bone pain)						
Pamidronate	IV	1-2	60-90	Infuse over 2-24 hours every 1-4 weeks		Child dose extrapolated from adult experience
						Monitor serum electrolytes, calcium, phosphorus
Alendronate	PO	0.07-0.14	5-10	Daily to		Child dose extrapolated from adult experience
		0.5-1	35-70	Weekly		Watch esophageal irritation

Sources: Micromedex® Healthcare Series [Internet database]. Greenwood Village, CO: Thomson Healthcare. Updated periodically; Robertson J, Shilkofski N (eds.). Harriet Lane Handbook: A Manual for Pediatric House Officers, 17th ed. Philadelphia, PA: Mosby: 2005; Taketomo CK. Pediatric Dosage Handbook 2002-2003, 9th ed. Lewisville, TX: JA Majors Co.; 2002; Quill T, Holloway RG, Stevens Shah M, Caprio TV, Storey P. Primer of Palliative Care, 4th ed. Glenview, IL: American Academy of Hospice and Palliative Medicine; 2007; Hospice Pharmacia Pediatric Pharmaceutical Care Tool Kit, 1st edition (unpublished).

Continued from page 38.

Gina's Case Continues

Gina spends a comfortable week at home on an increased dosage of codeine, 1 mg/kg, along with acetaminophen, 15 mg/kg (300 mg acetaminophen) every 4 hours, and a laxative regimen. Gina's mother calls to say that Gina is reporting severe pain in her knees and hips. Her mother knows she cannot exceed the current dosage of medication because the maximum dosage of acetaminophen is already being used. Gina rates her pain as 8/10 on the Bieri Faces Scale.

Question Three

Which of the following is the best intervention to manage Gina's pain? (Choose one response.)

A. Continue the codeine and start oral morphine (starting dosage as indicated in Table 6).
B. Increase the codeine by 50% to 1.5 mg/kg/dose PO every 4 hours, keeping the acetaminophen at the same dose.
C. Increase the frequency of dosing of the codeine from every 4 hours to every 2 to 3 hours.
D. Continue the acetaminophen, discontinue the codeine, and start morphine 3 mg PO every 4 hours with a breakthrough dose of 2 mg every 1 hour as needed.
E. Discontinue the codeine and use one of the opioids from the agonist-antagonist group.

Correct Response and Analysis

The correct answer is D. The 3 mg scheduled morphine dosage was calculated by multiplying Gina's weight of 20 kg with a conservative starting dose of 0.15 mg/kg/dose (see Table 6). The breakthrough dosage is between 5% and 15% of the total scheduled dosage of morphine that Gina is receiving in 24 hours.

Routes of Medication Delivery

Clinicians must become familiar with converting dosages of a medication when changing route of administration. See Table 6 (page 44) for appropriate starting dosages by route.

Oral Route

The oral route is the preferred route because oral medications are

- generally less expensive than other options
- available in a variety of preparations
- relatively easy to titrate
- administered without advanced skill
- easily provided in a variety of settings
- less frightening to most children.

Oral medications must be presented in ways that make them acceptable to a child. Important considerations are palatability, size of tablets, volume of solution, and frequency of administration. Until recently, many oral medications were available only in tablet form, which makes this route difficult for children who are unwilling or unable to swallow tablets. Various solutions can now be mixed with flavored vehicles to make them more palatable. Many children like grape or cherry acetaminophen solution, which can be mixed with an opioid as long as the acetaminophen dosage does not exceed 15 mg/kg/dose every 4 hours (maximum dose of 75 mg/kg/24 hours). Maple syrup, ice cream, snow cone flavorings, and applesauce can also be used. To increase palatability, some pharmaceutical companies have begun manufacturing better tasting medications and have developed concentrated solutions for children requiring high dosages. Some facilities have access to a compounding pharmacist who can develop special preparations.

Frequency of medication administration can be problematic. For example, prescribing medication on an every-4-hours schedule is not very practical for anything other than short-term use. Fortunately, many analgesics are now available in sustained-release preparations. Some of the newer preparations, such as some sustained-release morphine capsules, can be opened without affecting their sustained-release properties,

unlike tablets, which lose their sustained-release capabilities when crushed or dissolved. The capsules' contents, referred to as sprinkles or beads, can be mixed with soft foods, but the beads cannot be chewed. A sustained-release sprinkle of hydromorphone is available in Canada and is awaiting approval in the United States.

The oral route is not the preferred route when

- the child prefers a different route
- aspiration is a significant concern, as might be the case with severe neurologic impairment
- the gastrointestinal tract is dysfunctional
- the child is experiencing a severe pain crisis, in which case the parenteral route is preferred for extremely rapid titration
- no long-acting preparation is available for long-term use.

Parenteral Route

Parenteral routes should be used when rapid titration is required, as in a severe, acute pain crisis or when a child is unable to tolerate the oral route. Several types of devices, including syringe drivers and patient-controlled analgesia (PCA) pumps, offer parenteral delivery of opioids and other medications. Such devices can deliver a continuous infusion, intermittent boluses, or both. Codeine cannot be given intravenously because of the risk of hypotension, but it can be given subcutaneously. Because PCA devices provide rapid analgesia, they are particularly helpful when patients experience incident pain (movement-related breakthrough pain). In addition, they give the child and family control over the administration of medication, which in and of itself improves pain relief and reduces anxiety.

Subcutaneous

If parenteral opioids are required, subcutaneous (SC) administration is usually easier to manage than repeated IV insertions in children without indwelling central venous access.[90] The dosing and efficacy of SC delivery are approximately equivalent to IV delivery.[91] Intermittent or continuous SC delivery, or a combination of both, is possible using a small-gauge (25 g to 30 g) indwelling butterfly needle, most often placed in the upper arm, leg, or abdominal wall and sometimes on the chest wall. However, care must be taken when placing needles in the chest because pneumothoraces have occurred on rare occasions. Although some clinicians suggest changing needles every 5 to 7 days, or sooner if redness occurs, many change the needle only when needed (eg, when skin irritation occurs). Alternatively, plastic catheters, typically used for SC administration of insulin or heparin, can be used for administration of opioids and other analgesics. Because of the availability of many highly concentrated parenteral opioid preparations, volume limits generally do not pose a problem unless very high dosages are needed for pain control. In this circumstance, highly concentrated hydromorphone may be particularly valuable.

Intravenous

For children who require IV medications but have difficult venous access, consider a permanent or semipermanent catheter, such as a peripherally inserted central venous catheter (PICC) line or other central venous device such as a Hickman or a port. These are appropriate when other intravenously administered treatments, such as blood products or parenteral nutrition, will continue. If such a catheter is in place, it can be used for all parenteral medications.

Intramuscular

The intramuscular (IM) route should not be used for administration of analgesics. Absorption is erratic, and children may decline analgesia if this route is used. There is generally no justification for IM administration of pain medication.

Rectal Route

Children generally dislike the rectal route, but it can be helpful for short-term administration when a child is unable to tolerate oral analgesics because of vomiting or swallowing difficulties. Although few dosage strengths are readily available in suppository form, oral preparations of sustained-release or immediate-release opioids can be administered rectally at the usual oral dosing intervals, which provides effective and easy administration. Compounding pharmacists can also prepare tailor-made sustained-release suppositories of many nonopioid medications used in symptom management, but documentation of efficacy is rarely available. Neutropenia and thrombocytopenia, commonly found in patients with diseases such as hematologic malignancies, are relative contraindications

to the rectal route because of the risk of infection (neutropenia) and bleeding (thrombocytopenia).

Transdermal Route

Fentanyl patches deliver medication transdermally, which can be helpful for children who are unable to take oral medications or for those with problematic parenteral access. The pharmacokinetics of transdermal fentanyl have not been studied in children, so additional care must be taken for this population.[92] Although patches range from 12.5 to 100 mcg/hour of fentanyl, the lowest-dose patch may be still be too large to use for some children.

Transdermal fentanyl may be indicated when pain relief has been consistently achieved with a stable dosage of oral or parenteral opioids. Transdermal fentanyl should

- not be used for an opioid-naive patient
- not be used when rapid titration is required, as in the case of uncontrolled pain
- be accompanied by doses of an immediate-release opioid for breakthrough pain
- be used with caution in the presence of skin abrasions, heating pads, or febrile illness due to the possibility of increased absorption of drug in these settings
- never be cut; cutting the patch can create a "leading edge" of a very large bolus of opioid with a completely unpredictable quantity of opioid delivered.

When the patch is removed, drug uptake continues for 12 to 24 hours.

Transmucosal Route

For Procedure-Related Pain

In pediatric settings, oral transmucosal fentanyl has been used for managing acute procedure-related pain (eg, lumbar punctures, bone-marrow aspirations, suturing lacerations). Oral transmucosal fentanyl is available in dosages as low as 100 mcg/unit. For procedure-related pain, dosages usually range from 5 to 15 mcg/kg. Because procedure-related pain generally occurs in an acute-care setting in opioid-naïve patients whose pain is short-lived, transmucosal preparations of fentanyl for managing procedural pain should be used only when carefully supervised by a knowledgeable clinician with experience in airway management and life-support techniques.

For Breakthrough Pain

The use of oral transmucosal fentanyl for managing breakthrough pain has been studied in the adult population and has been approved for children. Oral transmucosal fentanyl is available in lozenges ranging from fentanyl 100 mcg to 1,600 mcg per lozenge and in buccal tablets containing 100 mcg to 800 mcg. Analgesia begins within 5 to 10 minutes and peaks at 20 to 25 minutes. The rescue dosage must be determined by starting with the lowest dosage, 100 mcg, and titrating to relief. Buccal tablet dosages will be lower than that of transmucosal lozenges. As with other medications, the rescue dosage of oral transmucosal fentanyl for breakthrough pain is based on the patient's dosage of regularly scheduled pain medications.

Epidural and Intrathecal Routes

Epidural and intrathecal (intraspinal) routes are used when fairly localized pain persists and dose-limiting side effects prevent escalation of systemic analgesia. Indwelling catheters can be implanted if prolonged use is anticipated. These invasive methods, performed by specially trained clinicians, are sometimes needed for children with severe localized pain.

Generally, an opioid is used in intraspinal delivery systems (epidural, intrathecal) in combination with a local anesthetic or another agent such as clonidine or baclofen.[93] When converting the opioid dosage from systemic to intraspinal, considerable caution is required because of the wide variability of responses. Approximately one-tenth of the IV dosage is used epidurally, in one method, with an additional ten-fold dosage reduction when converting from the epidural route to the intrathecal route.[94] The transition from systemic to regional opioid analgesia requires particular vigilance because acute respiratory depression may occur even in an opioid-tolerant patient when the pain stimulus is attenuated or removed, illustrating the intense central respiratory drive stimulation that pain represents. A dose reduction of a systemic opioid in the range of 75% can prevent respiratory depression while usually avoiding opioid withdrawal. Some clinicians decrease the systemic opioid while titrating the epidural or intrathecal opioid dosage upward.

Calculating Starting Opioid Dosages

See Table 6 for guidelines on calculating approximate, weight-based starting dosages of an opioid for an opioid-naive child. *These dosing guidelines are only starting dosages and should not limit upward titration if necessary for relief. See special considerations below when starting opioids for neonates.*

As with adults, the goal of pain relief for children is to maximize comfort and maintain function to the greatest extent possible. Starting with an adequate dosage of medication and rapidly titrating to relief are important steps to achieving pain control. When pain is allowed to recur regularly or the child has to ask for medication and then wait for relief, pain is exacerbated and feelings of anxiety and mistrust are heightened. Additionally, more medication is generally needed to "chase after pain" than to prevent it.

Except in situations of very intermittent, infrequent pain, an opioid should be initiated on a scheduled, around-the-clock basis. Initially, dosages of an immediate-release preparation should be given every 4 hours, with rescue dosages for breakthrough pain administered as needed. When possible, the same opioid should be used for both the scheduled dosages and the

CLINICAL SITUATION

Dexter

Dexter is a 6-year-old boy with spastic cerebral palsy, minimal relational potential, and bilateral hip dislocation. His hip arthritis seems to be causing moderate pain, which is indicated by facial grimacing and moaning as well as bruxism when he is moved for diaper changes. His parents report that this pain does not appear to be as severe as when he had his scoliosis repair. You decide to treat Dexter's mild-to-moderate pain with an opioid and acetaminophen (15 mg/kg/dose). Dexter weighs 20 kg.

Question One

What opioid dosage should you start with?
A. Codeine, 30 mg PO every 4 hours, which is in the lower dosage range for an adult
B. Codeine, 5 mg PO every 4 hours, because his pain is mild-to-moderate
C. Codeine, 10 mg PO every 4 hours
D. Codeine, 10 mg PO as needed

Correct Response and Analysis

The correct answer is C. Because Dexter weighs less than 50 kg, the recommended initial starting oral dose of codeine is 0.5 to 1.0 mg/kg every 4 hours around the clock. A conservative 0.5 mg/kg multiplied by Dexter's 20-kg weight equals a 10 mg starting dose. Other appropriate opioid options are hydrocodone or oxycodone. See Table 7 for approximate starting dosages for those opioids based on the child's weight.

Question Two

At what frequency should the codeine be given to Dexter?
A. As needed
B. Every 4 hours around the clock
C. Every 6 hours initially, then reassess the interval
D. Every 2 hours to achieve more immediate relief

Correct Response and Analysis

The correct response is B. Because Dexter's arthritis is not expected to resolve, pain medication should be given at regularly scheduled times, not as needed. Pain that occurs on anything more than an occasional basis should be treated on a regular, around-the-clock schedule, not intermittently. Dosing intervals are determined by the medication's duration of action.

Question Three

Which of the following are reasons for choosing the oral route? (Choose as many as are appropriate.)
A. The oral route is easily accessed in a variety of settings.
B. The oral route is less expensive than other options.
C. The oral route may be less frightening to the child than other routes.
D. The bioavailability of oral opioids for children exceeds that documented for adults.

Correct Response and Analysis

The correct answers are A, B, and C. A variety of preparations other than tablets is available and acceptable to children.

rescue dosages for breakthrough pain. This approach facilitates opioid-dose titration based on frequency of rescue dose requirements. Orally administered medications for breakthrough pain are dosed approximately every hour as needed. Intravenous dosages can be given every 15 minutes as required. If a child requires frequent rescue dosages (generally more than four to eight in a 24-hour period), both the scheduled dosages for background pain and the rescue dosages for breakthrough pain should be increased.

Use of Opioids in Neonates

Infants younger than 3 months are more vulnerable to opioid-related respiratory depression because of the following factors[95]:

- Immature liver function may increase the duration of action of opioids, requiring longer dosing intervals.
- Decreased clearance by the kidneys may also increase the duration of action of opioids.
- A high ratio of body water to fat increases the volume of distribution of the drug, thereby increasing its duration of action.
- Highly perfused tissues, such as brain, heart, and viscera, make up a larger proportion of body mass, increasing the amount of the drug delivered to the cerebral circulation.
- Reduced levels of circulating albumin and glycoprotein can increase the ratio of unbound (active) to bound (inactive) drug, increasing the effect of each dose.
- A reduced ventilatory response to low O_2 and high CO_2 can predispose the patient to inadequate gas exchange.[96]

This risk can be managed easily with conservative initial dosages and good monitoring. The initial dosage of opioids for children 6 months and younger is approximately 25% to 33% of the usual childhood dosage on a mg/kg basis. Once the effect of the initial dosage has been evaluated, the opioid should be titrated to symptom relief using the same incremental parameters as used in older children and adults.

Calculating Continuous Opioid Infusions for Children

Calculating Opioid Infusions for Opioid-Naive Children

- An appropriate starting infusion rate for an opioid-naive child with continuing pain is 0.03 mg/kg/hour of morphine (0.01 mg/kg/hour for neonates), with a rescue dose for breakthrough pain available every 15 minutes.
- Another option is to begin with bolus opioid dosages every 4 hours to achieve pain relief and then to divide the every-4-hours dosage by 4, which can then be delivered as the hourly infusion rate. However, the more consistent pain relief achieved with constant infusions may allow a lower average hourly milligram dosage that achieves the same degree of relief.

Calculating Opioid Infusions for Children Already Receiving Opioid Therapy

When calculating opioid infusions for children who are already receiving opioid therapy, one option is to calculate the rate based on the child's total previous 24-hour opioid requirement (ie, the around-the-clock baseline dose plus any rescue or breakthrough doses). For a child on enteral opioids, use the equianalgesic table (**Table 7**) to convert the enteral dose to a parenteral dose.

Opioid Titration

Titrating opioids is an important pain-management technique used daily in hospice and palliative care settings. Opioid dosages can be increased by 25% to 100% or more each day, as needed, until pain is relieved. There is no ceiling or maximum recommended opioid dose. Children in the terminal phase of life may require and tolerate extraordinarily large dosages of opioids.[97] Dosages as high as several hundred milligrams every 4 hours may be needed to relieve severe pain.

During the titration process, the physician must remember that immediate-release morphine is a 4-hour drug that reaches its peak effect after approximately 45 to 60 minutes when taken orally, after 15 to 30 minutes when delivered SC, and after 5 to 15 minutes when given IV. When pain is not relieved within these peak times,

Table 7. Equianalgesic Dosages of Opioids (For starting dosages, see Table 6.)

Drug	Oral	SC/IV	Common Formulations / *Cautions*
Codeine	65 mg	IV not available	Oral tablets (15, 30, 60 mg)
			Fixed dose combinations (codeine/acetaminophen):
			Oral liquid (12.5/125 per 5 mL)
			Tablets (15/325 mg, 30/325 mg, 60/325 mg)
			Monitor total acetaminophen dose.
			Use Caution in patients with renal failure.
Morphine	10 mg	3.5 mg	Immediate-release tablets (10 mg, 15 mg, 30 mg)
			Oral solution (2 mg/mL, 4 mg/mL)
			Conc. (20 mg/mL) can give buccally
			Sustained-release tablets (15 mg, 30 mg, 60 mg, 100 mg, 200 mg) every 12 hours
			Rectal suppositories (5 mg, 10 mg, 20 mg, 30 mg)
			Use Caution in patients with renal failure.
Oxycodone	6.5 mg	Not available	Immediate-release tablets (5 mg)
			Immediate-release liquid (20 mg/cc)
			Sustained-release (10 mg, 20 mg, 40 mg, 80 mg) every 12 hours
			Percocet (oxycodone/acetaminophen): 2.5/325 mg, 5/325 mg, 7.5/500 mg, 10/650 mg)
			Monitor total acetaminophen dosage.
Hydromorphone	2.5 mg	0.5 mg	Immediate-release tablets (2 mg, 4 mg, 8 mg)
			Immediate-release liquid (1 mg/cc)
			Acceptable for patients with renal disease.
			High equianalgesic potency.
Methadone	Oral morphine : methadone ratio varies markedly across dosage range. Consider consult.		Tablets (5 mg, 10 mg, 40 mg)
			Liquid (1 mg/cc, 2 mg/cc, 10 mg/cc)
			Long variable T½. Small dosage change makes a big difference in blood level.
			Always write/advise "hold for sedation."
			Consider consult for high-dose conversion, IV conversion, or if prescriber is inexperienced.
			Acceptable for patients with renal disease.

an additional dose should be given. During the titration process, no ceiling exists on the number of times that a dose may be titrated. During a 24-hour period, some patients may require more than a 100% increase over their initial daily baseline dose.

Titration of Oral, Rectal, and Transdermal Opioids

Titration depends on the level of pain severity, half-life of the medication, and a patient's renal and hepatic function. In general, increase a dose by 25% to 50% for mild-to-moderate pain and 50% to 100% for moderate-to-severe pain. Most short-acting medications (eg, morphine, oxycodone) can be increased safely every 2 hours, but long-acting preparations (eg, sustained-release morphine, oxycodone) are increased every 24 hours. Transdermal fentanyl patches can be adjusted every 72 hours. Methadone dosages are usually escalated no more frequently than every 4 to 7 days (see Rotating to Methadone in Unipac 3, page 48). Persons with renal insufficiency or hepatic disease may require a smaller increase in dose or longer titration interval (see Special Considerations in Unipac 3, page 42).

Titration of Intravenous or Subcutaneous Infusions

If the patient's level of pain control is insufficient, the basal dose should be escalated by 25% to 100%, depending on pain severity. In addition, the patient may require a "loading" dose, which is generally 2 to 3 times the original basal dose, prior to increasing the infusion rate.

For example, a patient on a morphine infusion of 2 mg/hour has severe, uncontrolled pain. A loading dose of 4 mg IV followed by an increase in infusion rate to 4 mg/hour should be prescribed.

NOTE. During the entire titration process, continuing reassessment is needed. Pain that does not respond to opioids may need to be treated with adjuvant drugs or other specific remedies. Nonphysical pain must be treated with compassionate listening and involvement of other members of the interdisciplinary team. (See UNIPAC 2.)

Managing Breakthrough Pain

Breakthrough pain is associated with inadequate analgesic dosages, escalating disease burden, and pain on movement (ie, "incident pain"). The principles for managing breakthrough pain for children are the same as those for adults. Whether pain is being managed with an around-the-clock immediate-release opioid or a sustained-release preparation, additional rescue dosages should be readily available for pain that "breaks through" the scheduled dosing.

Breakthrough pain is treated with an immediate-release preparation, preferably using the same medication as scheduled. This approach eases titration and makes identification of the source of potential opioid-related side effects easier. The rescue dosage for breakthrough pain is calculated as a proportion of the scheduled (baseline) daily opioid requirement. The dosing interval for rescue dosages is based on the specific agent's pharmacology specifically, the time to peak effect. For orally administered immediate-release morphine, the time to peak effect is approximately 1 hour. If ineffective relief is achieved at the time of peak effect, the rescue dose can be repeated at that time.

Any of the following methods are appropriate for calculating a rescue dose based on the baseline requirement:

- 5% to 15% of the 24-hour opioid requirement
- 50% to 200% of the hourly dosage if the child is on a continuous parenteral infusion
- 25% to 50% of the 4-hour oral or parenteral dose.

Rescue dosages for breakthrough pain generally should be available every hour as needed when using the oral route, every 15 minutes as needed IV, and every 30 minutes as needed SC. In some instances, depending on the time to peak effect and the patient's need for rapid titration, more frequent rescue dosing is reasonable.

Children taking a sustained-release preparation sometimes experience more pain before receiving the next dose of medication. Although this usually means the dose is too low, it may also indicate that the child requires a shorter interval between administration. Smaller, more frequent dosages using the same total mg/24-hour dosage may correct the problem. For example, if a child receiving 120 mg of sustained release morphine every 12 hours experiences generally good pain relief and minimal side effects but experiences pain about 8 to 10 hours after receiving a dose

of sustained-release medication, the schedule could be changed from 120 mg every 12 hours to 80 mg every 8 hours. If the child is experiencing side effects from the sustained-release opioid at the time of the medication's peak plasma concentration, it may be helpful to continue the same total 24-hour dose but to prescribe smaller doses more frequently. An example would be a child experiencing nausea 4 hours after the administration of a sustained-release opioid.

Children on an every-4-hours medication schedule may benefit from receiving 1.5 to 2 times their usual dosage at bedtime, which may reduce the need to wake them to take their medication during the night. Widespread experience with this regimen suggests it is safe and does not carry undue risk of respiratory depression. However, a clinical trial evaluating 20 adults who took twice their usual dosage of immediate-release morphine at bedtime found no better analgesia and increased xerostomia and vivid dreams.[98]

Incident pain can be particularly problematic. It occurs with movement and is typically short-lived but severe. The dose of analgesia adequate to relieve the brief, intense episodes of pain on movement may lead to oversedation when the incident pain resolves, since the background pain is not as severe as the pain with movement. Nonpharmacological strategies to help prevent incident pain include stabilizing the painful area with positioning and bracing. Before movement, boluses of a parenteral opioid, often by using a PCA, may be of benefit. Oral transmucosal fentanyl citrate (OTFC), a rapid-onset, rapidly metabolized medication, may offer effective pain control for incident pain. (See UNIPAC 3 for more information on managing breakthrough pain.)

Conversion to Long-Acting Preparations

When stable pain relief is achieved, conversion to a sustained-release preparation is recommended. For example, divide the previously prescribed 24-hour dosage of morphine into two or three larger dosages instead of continuing with six smaller dosages. Dosages of most sustained-release preparations should be given every 8 to 12 hours, depending on clinical response. After con-

version to a long-acting preparation, rescue dosages of an immediate-release, short-half-life opioid should be readily available for breakthrough pain (eg, morphine or hydromorphone). Examples of long-acting preparations includ

- sustained-release morphine: Oramorph SR (usual dosing interval, 8 to 12 hours), MS Contin (usual dosing interval, 8 to 12 hours), Kadian (usual dosing interval, 12 to 24 hours), Avinza (usual dosing interval, 24 hours), generic morphine SR (usual dosing interval, 8 to 12 hours)
- oxycodone (Oxycontin; usual dosing interval, 8 to 12 hours)
- oxymorphone (Opana ER; usual dosing interval, 8 to 12 hours)
- transdermal fentanyl (Duragesic or generic patches; usual dosing interval, 48 to 72 hours)

Methadone

Methadone is considered a long-acting opioid because of its very long biologic half-life. Metaanalysis of randomized trials suggests that methadone's safety and effectiveness is similar to that of morphine with adult patients.[99]

However, the analgesic half-life of methadone, particularly at the start of therapy, does not generally correspond with its long biologic half-life (it exhibits a wide range of approximately 6 to 80 hours). In addition, the cross-tolerance between methadone and other opioids is dramatically incomplete despite the dose conversion recommended in some tables, many of which are based on single dosages rather than chronic administration. The range of methadone-to-morphine dosage ratios varies from 1 : 5 up to higher than 1 : 88. Clinicians must use great care when dosing and monitoring methadone. It can be extremely useful for some patients, but it should be prescribed only by clinicians with experience using this particular opioid. Physicians can ensure a greater margin of safety by converting only the sustained-release component of the regimen and using breakthrough-pain medication with a short half-life until consistent pain relief is achieved (at least 3 days) using the same methadone dosage. Close follow-up is essential. See UNIPAC 3 for more information on methadone dosing.

Gina's Case Continues

Gina is cared for at home with scheduled doses of morphine, 3 mg every 4 hours (0.15 mg/kg multiplied by her 20-kg weight) and rescue doses of morphine, 2 mg every hour as needed. For the past several days she has used six rescue doses of morphine (2 mg/dose) to control her pain. To simplify her dosing schedule, she is placed on sustained-release morphine, 15 mg every 12 hours. This dose was calculated by adding Gina's total morphine intake over a 24-hour period—3 mg multiplied by 6 doses plus 2 mg multiplied by 6 doses—or 30 mg per day, and then dividing the total daily dose into two doses of 15 mg. An immediate-release analgesic must be readily available for breakthrough pain; that is, pain that is escalating, is inadequately treated, or occurs on movement (incident pain).

Question Four

Which of the following is an appropriate dosage of an immediate-release analgesic for Gina's breakthrough pain?
A. Immediate-release morphine, 3 mg PO every 1 hour as needed for pain
B. Immediate-release morphine, 3 mg PO every 4 hours as needed for pain
C. Codeine, 1 mg/kg PO every 1 hour as needed for pain
D. Codeine, 1 mg/kg PO every 4 hours as needed for pain

Correct Response and Analysis

The correct response is A. To control Gina's breakthrough pain, it is best to use an immediate-release preparation of the same opioid as that being used to manage her background pain. Codeine is not an appropriate choice for breakthrough pain for a child who requires regular dosages of morphine. Gina's baseline dose for managing background pain is sustained-release morphine, 15 mg every 12 hours. An appropriate rescue dose for breakthrough pain is morphine, 3 mg PO (10% of her 24-hour morphine requirement), given every hour if needed.

Opioid Tapering

If a child who is receiving large dosages of opioids undergoes a treatment or procedure that successfully relieves pain (eg, insertion of an epidural device or a nerve block), vigorous opioid tapering may be needed (reductions of 75%). Less dramatic but similar results are sometimes seen when an effective adjuvant analgesic is added. Even when patients are considered opioid tolerant, dose-related toxicity may manifest acutely when the pain stimulus is suddenly removed. Manifestations of withdrawal are unlikely to occur with a 75% taper under these circumstances, but careful monitoring for sweating and agitation should be maintained.

Some patients require continuous opioid therapy for 1 week or longer for a painful condition that resolves. In this case, the opioid must be tapered to prevent withdrawal symptoms. Dosages can be reduced by 10% to 20% every 1 or 2 days.[100] Once a dose of 0.6 mg/kg/day (if less than 50 kg) or 30 mg/day (if greater than 50 kg) of oral-morphine equivalent is reached, the opioid can be discontinued. If the patient experiences withdrawal symptoms (eg, anxiety, tachycardia, sweating, rhinorrhea, diarrhea), a higher dosage should be reinstituted until these symptoms abate, then more gradual tapering is initiated. Some individuals may require a higher dosage than the last one for which no withdrawal symptoms occurred to reverse withdrawal symptoms.

Gina's Case Continues

When Gina got the flu that was going around her school, she became unable to tolerate her oral morphine because she was vomiting and had diarrhea. She regurgitated about one-third of the medication.

Question Five

Of the following choices, which is the best option for providing ongoing pain relief?

A. Administer her morphine rectally using the same sustained-release medication and the same dosage.

B. Convert her morphine to a parenteral dose, and administer it intramuscularly.

C. Convert her morphine to a parenteral dose, and administer it subcutaneously or intravenously.

D. Insert a nasogastric tube and administer her morphine through the tube.

Correct Response and Analysis

The best response in most circumstances is C. Gina can receive morphine subcutaneously or intravenously if she has an indwelling line, as do many children who have been recently treated for cancer. Opioids, even oral sustained-release preparations, can be administered rectally and are effective as long as they are in contact with the rectal mucosa. However, this route is probably best avoided in Gina for four reasons: (1) she has neutropenia (her WBCs are predominantly blasts), (2) her platelet count is low, (3) she dislikes the rectal route, and (4) diarrhea precludes the rectal route as an option. Rectal administration puts her at risk of bleeding and infection. The intramuscular route should never be used to provide analgesia. Similarly, other potentially objectionable routes (eg, nasogastric tube) should not be used unless they are in place for some other reason.

Question Six

Gina had been taking sustained-release morphine in capsule form, 15 mg PO every 12 hours. Her pain had been well controlled until the past 24 hours, when she required five additional rescue doses of immediate-release morphine, 3 mg PO for breakthrough pain, after vomiting her sustained-release morphine. What should Gina's new baseline (around-the-clock) dosage of parenteral morphine be? (Choose all that apply.)

A. Morphine, 1.7 mg IV every 4 hours

B. Morphine, 5 mg IV every 4 hours

C. Morphine, 0.4 mg hourly as a continuous infusion

D. Morphine, 1.5 mg IV every 12 hours

Correct Response and Analysis

The correct response is A or C. When changing from oral to parenteral opioid, the current 24-hour dose requirement is usually determined by adding the regularly scheduled dosages and any rescue dosages and then consulting an equianalgesic table. The ratio of oral morphine to parenteral (IV/SC) morphine is approximately 3 : 1 for patients like Gina who require regular dosing.

Gina's daily morphine dose was 15 mg multiplied by 2 doses—30 mg per day (baseline dose)—plus 3 mg multiplied by 5 doses—15 mg (rescue doses)—for a total 24-hour dose of 45 mg. Because there is no change in agents, a simple arithmetic conversion of oral morphine to parenteral morphine (3 : 1) is applied. Thus, 45 mg oral morphine divided by 3 is 15 mg of parenteral morphine per day, which could be given every 4 hours (6 doses/day) or as an hourly infusion (24 doses/day). Fifteen milligrams of parenteral morphine per day divided by 6 equals 2.5 mg of parenteral morphine every 4 hours; 15 mg of parenteral morphine per day divided by 24 equals 0.625 mg of parenteral morphine per hour.

However, it is important to adjust Gina's dose downward; she most likely required rescue doses because she vomited some of her medication. In this circumstance, it is reasonable to start with an SC or IV bolus dose about one-third less than was calculated above (multiplied by 0.67) and then titrated to relief. Parenteral morphine, 2.5 mg every 4 hours, multiplied by 0.67 is 1.675 mg (or about 1.7 mg) of parenteral morphine every 4 hours. Parenteral morphine, 0.625 mg per hour multiplied by 0.67 is 0.41875 mg (or about 0.4 mg) of parenteral morphine per hour. Gina should be watched closely and the dose titrated as needed from here.

Opioids to Avoid in Pediatric Palliative Care

Meperidine is a short-half-life opioid used for moderate-to-severe pain. Its active metabolite, normeperidine, not only has a very long half-life (about 30 hours) but is also toxic to the central nervous system and can cause irritability, insomnia, myoclonus, and seizures. Although toxicity is possible at any dosage, it is most likely to occur when meperidine is used at higher dosages, if a patient has renal or hepatic insufficiency, or because of accumulation after repeated dosing for more than 2 or 3 days. Meperidine has an atropine-like structure and can cause tachycardia. It is also a negative inotrope and can have fatal interactions with monoamine oxide inhibitors (MAOIs).

Propoxyphene, used for mild-to-moderate pain, is available alone or as a fixed-dose combination with aspirin or acetaminophen. In these formulations, the propoxyphene dosage provides minimal analgesia. Propoxyphene's active metabolite, norpropoxyphene, is toxic to the central nervous system and accumulates with repeated dosing. Norpropoxyphene's effects are similar to normeperidine's effects.

Managing Opioid-Related Side Effects

When initiating opioid therapy, clinicians should review the child's history for any prior opioid experience or side effects. The physician's discussion with the child and family should include common opioid side effects, such as constipation, increased sleepiness, nausea, or mild itching, and the plan to prevent or relieve these side effects. Most side effects resolve within several days, leaving the child able to tolerate the opioid very well. Constipation is the exception and requires treatment throughout the course of opioid therapy. Some patients and family members mistakenly attribute side effects, such as itching or nausea, to allergy. True opioid allergies are rare and often manifest as bronchospasm (wheezing), hives, or anaphylaxis, not as isolated pruritis or nausea. It is critical to anticipate and manage opioid-related side effects because children are more likely than adults to refuse a medication that causes distressing side effects even when experiencing effective pain relief.[43] Trust in the practitioner is an essential ingredient for success.

When a symptom is clearly opioid related, it is important to remember that side effects are drug specific and dose related. Most opioids are metabolized by the liver and excreted by the kidneys. For a patient with renal insufficiency, there is an increased risk of neurotoxicity with morphine, probably due to accumulation of its centrally acting metabolite, morphine-6-glucuronide.[101] However, opioid metabolism can also be altered by significant hepatic compromise, which can lead to higher drug levels.[102] Lowering the opioid dosage, if pain is well controlled, or rotating (changing) opioids are management options. Opioid rotation may be the best option when a side effect is especially troublesome, when it requires time to resolve, or when the child is unable or unwilling to take another medication to counteract the opioid-related side effect.

Adjuvant Medications

Adjuvant medications have coanalgesic properties even though their primary indications are for the treatment of conditions other than pain. They are generally added

to opioid analgesia to enhance pain relief or allow a reduction in opioid dosages, reducing concomitant side effects. (See Table 6.) Identifying a patient's specific pain syndrome is important to determine which adjuvants might be most helpful. Adjuvants with differing mechanisms of action can be combined for added benefit.

In palliative care settings, adjuvants offer the potential benefit of providing relief for more than one symptom. The type of pain the patient is experiencing and the adjuvant's side-effect profile is largely what determines which adjuvant medication a physician should choose. For example, the analgesic and sedating properties of tricyclic antidepressants can help a child with neuropathic pain who is also experiencing insomnia. Some adjuvants are relatively specific, and others, such as steroids, are beneficial for several types of pain and other symptoms. Steroids can relieve bone and neuropathic pain, capsular-distension-related pain, vomiting due to increased intracranial pressure, or partial bowel obstruction. They can also improve appetite and a patient's general sense of well-being.

Adjuvants for Specific Pain Syndromes

Pain is generally classified into two broad categories: nociceptive, which includes somatic and visceral pain, and neuropathic pain. A patient may experience several types of pain concurrently. For example, a collapsed vertebra can cause bone pain but may also cause distorted neural foramen with neuropathic pain from nerve impingement, as well as somatic pain from muscle spasm associated with nerve inflammation. Each type of pain may require a different treatment.

Nociceptive Pain

Somatic pain. Somatic pain is caused by injury or inflammation of soft tissue or bone. Somatic pain tends to be well localized and is often described as sharp. Standard analgesics (nonopioid and opioid) are effective for the treatment of somatic pain.

Bone pain may require a more complex treatment plan. Diffuse bone pain is seen in children with marrow expansion from hematologic malignancies, such as leukemias, and from other cancers of childhood that invade bone marrow, such as neuroblastoma. Focal bone pain due to localized or metastatic bony lesions is less common in children than in adults but can occur with sarcomas, such as osteogenic and Ewing's. Focal pain due to skeletal morbidity, including fracture, vertebral collapse, or osteonecrosis of the femoral head, may occur because of treatment-related factors, such as prolonged steroid use, and with conditions associated with osteoporosis, such as muscular dystrophy or cancer.[103]

Nonsteroidal antiinflammatory drugs (NSAIDs) are effective adjuvants for the management of bone pain. Their use may be contraindicated in children with low platelet counts because of chemotherapy or marrow-invasive malignancy. Steroids are potent antiinflammatory agents. Bisphosphonates (alendronate, pamidronate) help reduce skeletal morbidity and provide analgesia by decreasing osteoclast activity. Their expense and the nature of some of the regimens (parenteral infusion of pamidronate every 3 weeks) may be prohibitive, but some children clearly seem to benefit.[104] Selective radiotherapy may be appropriate for sites of focal pain.

Visceral pain. Visceral pain is caused by distension (eg, bowel obstruction, capsular distension). It tends to be poorly localized and may be referred to distant sites, such as shoulder-tip pain from hepatic distension. Steroids can be useful for treating visceral pain in combination with opioids.

When severe pain crises occur, a large, one-time dosage of a steroid can be very helpful. Corticosteroid therapy involving short-term administration of high dosages can be appropriate for selected patients with severe, progressive pain. For children, an initial dose of approximately 1 mg/kg of methylprednisolone or dexamethasone is followed by tapering to a lower daily maintenance dosage. Guided imagery can also be used for some recurrent abdominal pains in children.[105]

Neuropathic Pain

Neuropathic pain is caused by disordered function of the nervous system anywhere along its course, from the periphery to the cerebral cortex.[106] A patient's description of the pain is often diagnostic. Neuropathic pain is commonly described as *burning, pins-and-needles* pain, or as sudden, sharp, and shooting pain. Sensory abnormalities such as numbness or hyperalgesia or other neurologic deficits may be found on examination.

Neuropathic pain typically requires treatment with opioids in combination with adjuvants. Sequential trials of various adjuvants are sometimes necessary to achieve relief. Methadone is a preferred opioid because it blocks NMDA receptors known to mediate neuropathic pain. Tricyclic antidepressants are useful adjuvants at dosages lower than those required for treating depression. Anticonvulsants such as gabapentin or carbamazepine are useful adjuvants, although the potential bone-marrow suppression associated with carbamazepine limits its usefulness for children with hematological comorbidities. Topical lidocaine patches have also proven to be beneficial.[107] Systemic local anesthetic agents, such as oral mexiletine or IV/SC lidocaine, can be useful for managing neuropathic pain despite their primary indication for cardiac arrhythmias.[108]

Preventing and Managing Procedure-Related Pain

Effectively preventing and managing procedure-related pain is vital whether the procedure occurs once or repeatedly throughout the course of an illness. The child's first experience with a procedure will significantly affect his reactions to subsequent procedures, making meticulous attention to pharmacologic and nonpharmacologic analgesia essential. The basic management principles listed here also apply to disease-related pain.

- Address both the psychological and physical components of pain by including pharmacologic and nonpharmacologic interventions.[109]
- Ask about the child's past experience with procedure-related pain—what helped and what didn't. Explain to the child why the procedure is being performed.
- Encourage the use of topical local anesthetics.

Pharmacologic Measures to Relieve Procedure-Related Pain[110]

Sucrose and the act of sucking provide an analgesic effect for newborn infants for minor procedures such as heel-sticks (**see Table 8**). Sucrose is available as a 12% solution. For term infants, the sequence is as follows: (1) slowly give 1.5 to 2 cc PO over 2 minutes, (2) wait 2 minutes before starting the procedure, and (3) if needed, give the remain-

Table 8. Sucrose Administration for Pain	
Gestational Age	**Dosage**
< 28 weeks	0.2 cc swabbed into mouth
28-32 weeks	0.2-2 cc depending upon suck/swallow
> 32 weeks	2 cc

ing 0.5 cc of sucrose on a pacifier or on a gloved finger. Analgesia should persist for up to 8 minutes. The dose may be repeated once. Sucrose works best for neonates but may be tried for infants as old as 3 months.[111]

EMLA (eutectic mixture of local anesthetics: 2.5% lidocaine and 2.5% prilocaine), a topical anesthetic cream, should be used when a procedure involves penetration through the skin, such as routine venipuncture, insertion of an intravenous line, access to a percutaneous central line, or administration of L-asparaginase or other treatments by intramuscular or subcutaneous injection. EMLA may also be helpful for neuropathic pain from cutaneous causes (eg, herpetic neuralgia).

EMLA is applied to the skin and then covered with an occlusive dressing for a minimum of 1 hour, but to a maximum of 4 hours; it is then wiped off, and the procedure is carried out. Adequate anesthesia is achieved in 1 hour, but the anesthetic effect will increase until 3 hours have passed if the cream is left in place. For injections, the cream should be left on for more than 2 hours (except as indicated in **Table 9**). Once the cream has been removed, the anesthesia will persist for 1 or 2 hours. Vasoconstriction is an occasional problem with EMLA use. This may be reversed by applying a warm pack to the site before the procedure. EMLA should only be applied to intact skin. EMLA may be used for infants who are 32 weeks of age and older. One risk of EMLA is methemoglobinemia. Infants, especially those being treated with other methemoglobin-inducing agents (sulfa drugs) or with G6PD deficiency, are more likely to have this complication.

Tetracaine gel 4%, applied 45 minutes before the procedure, was shown to be significantly more effective than EMLA for 60 minutes in a randomized trial of sixty 3- to 15-year-old children requiring venous canulation[112] but was not effective for infants receiving peripherally inserted central venous catheters.[113]

Table 9. EMLA Dosing per 24-Hour Period

Age and Weight	Max Dose	Max Application
0 to 3 months or up to 5 kg*	1 g	1 hour
3 to 12 months and greater than 5 kg	2 g	4 hours
1 to 6 years and greater than 10 kg	10 g	4 hours
7 to 12 years and greater than 20 kg	20 g	4 hours
* Only appropriate for infants older than 32 weeks' gestation		

Vapocoolant sprays cause a transient freezing of the skin surface. The anesthetic effect begins immediately after application but lasts less than 1 minute. It may be applied by direct spraying or by the application of a saturated cotton ball. Vapocoolant should only be applied to intact skin. If using the spray technique, hold the spray can 3 to 9 inches away from the site. Spray the vapocoolant onto the desired area for up to 10 seconds or until a white frost is immediately noted. Avoid getting the spray into the patient's face. Immediately swab the site with alcohol and perform the procedure. If using a cotton ball, place a cotton ball in the medicine cup. Spray the vapocoolant into the cup for 10 seconds. Apply the cotton ball to the desired area using forceps and hold it in place for 15 seconds. Perform the procedure immediately afterwards.

ELA-max is a 4% lidocaine cream. It is applied for 30 to 60 minutes before a painful procedure. It does not contain prilocaine, so methemoglobinemia is not a concern. Use of an occlusive dressing is not necessary but may help to hold the cream in place.

LET consists of lidocaine 4%, epinephrine 0.1%, and tetracaine 0.5%. It is suitable to use before laceration repair in children older than 6 months. Onset of action is 15 to 30 minutes, and duration of anesthesia is 45 to 60 minutes. First, apply LET with a cotton swab to the edges of the wound. Then, place a cotton ball saturated with LET into the wound and secure it with tape. If necessary, ask the parent or guardian to hold it in place with a gloved hand. LET may not be used on fingers, toes, earlobes, or the glans of the penis.

Local infiltration with buffered lidocaine stings less than unbuffered lidocaine. Buffered lidocaine should be made close to the time of the procedure with 1 part sodium bicarbonate to 9 parts lidocaine (with-out epinephrine) (0.1 mEq/mL). Let the alcohol dry. Use a small gauge needle (25 to 30 gauge). Inject the local anesthetic slowly over a couple of minutes. When injecting multiple times, try and inject through previously anesthetized areas. Buffered lidocaine may be better tolerated when the superficial tissues have been previously anesthetized using EMLA or vapocoolant. If marcaine is used, it should not be buffered because it will precipitate out.

Conscious sedation is useful for painful invasive procedures such as bone-marrow aspiration or lumbar puncture. Policies and credentialing are regulated by the healthcare institution. Become familiar with the administration of opioids and benzodiazepines before attempting to use them for managing procedural pain. Ensure adequate monitoring by skilled personnel other than the person performing the procedure. A healthcare professional skilled in airway management should be present when opioids or sedatives are used for conscious sedation. Procedural pain is brief and acute, with little or no time for gradual titration. Practitioners must be familiar with dosing on a mg/kg basis for children, and they must be skilled in the management of pediatric airways. Procedural and chronic pain management for children are described in the publication *Cancer Pain Relief and Palliative Care in Children,* published by the World Health Organization.[5] The American Academy of Pediatrics Committee on Drugs has published an excellent resource describing the personnel and skill sets necessary for safe and effective management of procedural pain titled *Guidelines for Monitoring and Management of Pediatric Patients During and After Sedation for Diagnostic and Therapeutic Procedures.*[25]

Nonpharmacologic Management of Procedure-Related Pain and Distress

Use cognitive and behavioral interventions along with pharmacologic interventions; both are more effective when used together than when used alone.[114-116] There is increasing interest in complementary and alternative medicine approaches to the management of pain in pediatrics.[117] Simple measures include the following:

- Provide honest, developmentally appropriate information about what to expect, tailored to the individual child's coping strategies. (Some children prefer preparation just prior to a procedure to reduce anxiety; others like detail provided well in advance of the event. Preparation can include why a particular test is being done, how it will be done, where it will be done, and who will be in the room.)
- Talk to the child before and after the procedure about what has been helpful, then implement the child's suggestions.
- Give the child choices whenever possible (eg, time, site, or location of procedure).
- Include the child in decision making based on his or her willingness and desire to participate. For example, offer options and ask how he or she wants to proceed. Children can make decisions about
  - interaction during the procedure, including explanations of what is going on
  - use of distractions, such as small pop-up books, a magic wand, a small container of bubbles, or headphones with a choice of tapes or CDs. (Music did not supplement analgesia for dental procedures in a series of 45 4- to 6-year-old children having a dental procedure, but it also was not harmful.)[118]
  - use of physical measures, such as gentle touch or massage
  - telling or reading the child's favorite story or singing his or her favorite song.
- Presence of a parent or other family caregiver who the child knows, likes, and trusts is the one thing children find most helpful when coping with a procedure.
- Provide parents with information about concrete ways to support their child during the procedure.

Other nonpharmacologic techniques requiring more training include the following:

- guided imagery[55,114-116,119] (child life and physical therapists are often trained in these skills)
- hypnotherapy, which has been effective both for pain[116,120-124] and dyspnea[125] in some children. For more detail, refer to *Hypnosis and Hypnotherapy with Children*.[119]

These techniques can be used when the procedure may cause anxiety but is not painful or when pain is induced but cannot be well managed with current techniques, for example, blood samples obtained by lancing the fingertip.

CLINICAL SITUATION

Case Example: Nonpharmacologic Therapy for Pain[5]

A 3-year-old boy with acute lymphoblastic leukemia requires intravenous vincristine therapy. Previously, he cried and had to be held down when intravenous treatments were started; now he is whimpering and clinging to his mother. EMLA was applied to his arm 2 hours ago. His mother is anxious but cooperative, having been coached by a child-life therapist, who has also explained to the child what will happen and how it will feel in words that he understands. In the waiting room, he is given soap solution and a wire loop for blowing bubbles. His mother shows him how to make the bubbles. The boy enjoys this and plays while the intravenous line is being prepared. Mother and child then go into the treatment room and the boy continues to blow bubbles while the puncture site is prepared and the tourniquet is applied. He chooses to sit on his mother's lap during the procedure and is encouraged to "blow away the hurt" as the needle is inserted. His mother and all the medical staff praise him for being brave. When he tires of blowing bubbles, his mother reads him his favorite story.

Assessing and Managing Selected Nonpain Symptoms

Pediatric Symptom Assessment and Management

The principles for assessing and managing nonpain symptoms parallel those of pain assessment and management. A decision to investigate the specific etiology of any symptom should be based on the goals of care and the relative burdens and benefits of the investigation, including the likelihood that determining the precise etiology would result in interventions to improve comfort that would not otherwise be employed. In most cases, the likely cause of a symptom is obvious, and therapy can be initiated empirically. Many therapies not specific to a particular etiology are effective nevertheless.

Managing Selected Nonpain Symptoms
Constipation

Constipation is such a common opioid-related side effect that a bowel regimen should be started whenever opioid therapy is initiated. Constipation is also a common cause of abdominal pain, nausea, and vomiting for patients with life-limiting illness. The goal of treatment is reestablishing the patient's usual bowel habits.

A stool softener (eg, docusate) and a stimulant laxative (eg, senna or bisacodyl) should be included in the treatment plans for most patients who are receiving opioid therapy. Because opioid-related constipation does not resolve over time and worsens as opioid dosages escalate, changes in opioid dosages should always be accompanied by changes in the bowel regimen. Agents should be chosen according to the patient's preference, with the exception of psyllium and other fiber-based compounds. The effectiveness of these bulking agents depends on adequate fluid intake, which makes them inappropriate choices for ill, anorectic patients. Both stool softeners and stimulant laxatives usually must be increased as the opioid dosage increases. Osmotic laxatives such as sorbitol, lactulose, or polyethylene glycol are also effective and may serve as good, second-line choices. One study suggested that polyethylene glycol produced better results and fewer side effects than lactulose in children.[126] Pilot studies suggest that probiotic mixtures may also be useful for constipation in children.[127] Painful or invasive interventions should be avoided when possible. For example, enemas are invasive, can contribute to the risk of infection in the neutropenic patient, and can precipitate bleeding in a patient with thrombocytopenia. However, they may be necessary and effective for patients with neurologic conditions associated with poor gut motility.

CLINICAL SITUATION

Mohammed

Mohammed is a 15-year-old adolescent with Duchenne's muscular dystrophy. He recently fell while being transferred out of his wheelchair and sustained soft-tissue injury to his back, requiring acetaminophen, codeine, massage, and heating pads for relief. When he began taking acetaminophen and codeine, he was placed on a stool softener, docusate 100 mg, and a stimulant laxative, senna 1 tablet daily. He has not had a bowel movement (BM) for 2 days, and his usual habit is a daily BM.

Question

What would you recommend for a bowel regimen for Mohammed now?

A. Add psyllium, 1 to 2 tablespoons daily, to his current regimen.

B. Begin daily enemas if there is no spontaneous stool.
C. Wait to intervene until he has not had a BM for 4 days.
D. Increase the doses and/or frequency of his stool softener and stimulant laxative and titrate until daily bowel movements are achieved.

Correct Response

The correct response is D. Psyllium is not likely to be effective and can cause impactions. Enemas are uncomfortable and invasive. Waiting may only make the situation worse. Doubling the above doses will likely be most effective. An occasional dose of an osmotic agent like sorbitol or polyethylene glycol in such situations is also likely to be effective.

Nausea and Vomiting

Nausea and vomiting are common symptoms in childhood illness and may be multifactorial in etiology. Nausea is the unpleasant subjective feeling that one needs to vomit. Usually mediated by the autonomic nervous system, it is caused by rapid distention of an abdominal viscus. Less frequently, it is caused by direct stimulation of medullary receptors. Vomiting is the coordinated forceful ejection of stomach contents through the mouth and nose. The vomiting center receives input from peripheral stimuli and from other areas of the brain: the chemoreceptor trigger zone (CTZ), the cerebrum and limbic system, and the vestibular system. Evaluation includes an assessment of the severity and the potential causes of a child's nausea and vomiting. When possible, reversible causes are treated promptly. For instance, the nausea of acute gastroenteritis can often be treated with oral or intravenous fluids instead of antiemetics.[128]

Regardless of the etiology of nausea and vomiting, the following physical and environmental measures should be offered:

- Avoid odors the child identifies as noxious or nauseating (eg, certain foods or perfumes).
- Use the child's favorite scent in the room (eg, a citrus or other fruit-based scent).
- Ensure good ventilation.
- Encourage upright posture for 1 hour after eating.
- Serve food and drinks in small and appetizing portions.
- Try complementary techniques, such as acupressure provided by Sea-Bands. The bands are applied over the wrists with the small projection placed over the acupressure site associated with nausea and vomiting. Although typically used for motion sickness, studies document benefits in other situations, such as chemotherapy-induced nausea and vomiting.[129]

Pharmacologic management—both prevention and treatment—may require multidrug therapy. Antiemetics work at specific sites of action; using two or more antiemetics from different classes with different sites of action has additive benefits and is a rational approach to managing severe nausea and vomiting. The predominant etiology guides the choice of antiemetic. See Table 6 for antiemetic dosing suggestions.

- When nausea is temporally associated with meals, a prokinetic agent such as metoclopramide can help reverse delayed gastric emptying. (Be prepared to treat possible dystonia with 1 mg/kg of diphenhydramine.)
- Ondansetron, a 5-HT3 receptor antagonist, is helpful for many types of nausea, especially chemotherapy-induced nausea and vomiting.
- Vomiting may occur in anticipation of a hospital or clinic visit for chemotherapy. Effective treatment of this nausea is essential to prevent chronic syndromes of vomiting.[130] Anticipatory vomiting may be effectively treated with lorazepam <u>plus</u> ondansetron and cognitive interventions.[130]
- When vomiting is movement related, a trial of a scopolamine patch or meclizine may be effective.
- Although dexamethasone or other steroids are helpful for all types of nausea, they may be of particular benefit in the presence of increased intracranial pressure and partial bowel obstruction. When determining the utility of steroids, particularly if long-term use is being considered, the unwanted side effects, such as change in appearance, acne, fluid retention, hyperglycemia, proximal weakness, demineralization of bones, and mood changes must be factored into the decision.
- When nausea or vomiting is reported at the peak onset of action of a short-half-life opioid, a reduction in the bolus effect may be achieved by changing to a sustained-release oral preparation or to a continuous parenteral infusion. An antiemetic can be added directly to the opioid infusion (see UNIPAC 4). Changing to an alternative opioid is indicated when nausea or vomiting is not relieved by these adjustments.

When nausea and vomiting are of uncertain etiology, the side effects associated with specific antiemetics may dictate which agent is chosen. For example, when insomnia or a mild degree of nighttime confusion is present, a neuroleptic agent is preferred over one of the antihistamines, but an antihistamine would be more appropriate if pruritus is a problem. The cannabinoids dronabinol and nabilone are effective antiemetics for adults receiving chemotherapy but

are often associated with dysphoria, paranoia, hallucinations, and hypotension.[131]

Figure 5, from the American Medical Association's Education for Physicians on End-of-Life Care Project, highlights several possible etiologies of nausea and vomiting as well as sites of pharmacologic action for relief.

Figure 5. Pathophysiology of Nausea and Vomiting
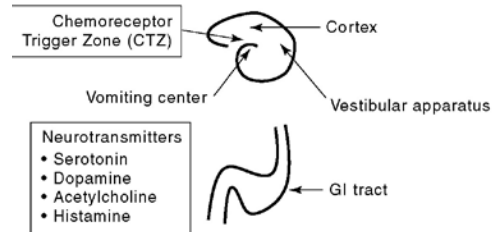
From The EPEC Project Web site, www.epec.net.[132] © by the Education in Palliative and End-of-Life Care Project. Reprinted with Permission.

CLINICAL SITUATION

Brian

Brian is a 3-year-old boy with a brainstem ependymoma. Chemotherapy and surgical resection have failed to arrest the tumor growth, and Brian is expected to die within the next 2 to 4 months. He has experienced increasing intracranial pressure (based on fundoscopic examination and symptomatic worsening) and resultant headaches, which are being treated with dexamethasone and morphine. He is vomiting frequently, but this symptom has improved somewhat with regularly administered metoclopramide. He had previously been treated with diphenhydramine without relief. He and his parents are very distressed by his ongoing vomiting and request help.

Question

Appropriate choices include (Choose all that apply)

A. hydroxyzine
B. dronabinol
C. dimenhydrinate
D. haloperidol.

Correct Response and Analysis

The correct answers are B and D. Adding an antiemetic from a different class (eg, dronabinol or haldoperidol) can be helpful. Hydroxyzine or dimenhydrinate are unlikely to be effective because both are antihistamines and have the same mode of action as diphenhydramine, which has already been unsuccessful for Brian. Agents working by different pathways should be added to the decadron that has been beneficial rather than substituting one for the other. His dexamethasone dose can also be escalated, depending on expected length of life, side effects experienced, and Brian's preference.

Pruritus

Pruritus associated with opioid therapy is generally not immune mediated; rather, it is caused by a non-specific histamine release. Children are often unwilling to take another medicine to counteract side effects. Trying a different opioid may be more appropriate than adding another medication. Additional options include local measures, such as skin emollients, soft bed sheets, nonirritating detergents for washing clothes and sheets, and pharmacologic interventions such as antihistamines.

If nausea is also a problem, antihistamines may be effective for both. Doxepin is an antidepressant with histamine-blocking properties that may be helpful if pruritus is difficult to treat.[133] Doxepin is also useful for treating neuropathic pain. Dosing starts at 0.2 mg/kg PO at bedtime, and is titrated to effect with a usual maximum dose of 150 mg/day. Blood levels may help guide therapy in much the same way as they do for anticonvulsants. Topical application of 5% doxepin has also been described for pruritus, although it must be restricted to relatively small areas (not greater than 10% of the body's surface area).[91,134] Other antidepressants and antipsychotics have been used to treat pruritis from a variety of causes.[135]

CLINICAL SITUATION

Sammy: Addressing Breathlessness

Sammy is 10 years old. He has a progressive neurodegenerative disease and is acutely ill with cough and fever. His physician suspects aspiration pneumonia. Sammy appears distressed and breathless despite nasal oxygen, although his caregivers are unable to confirm this from verbal self-report. He is not receiving any medications other than anticonvulsants and metoclopramide. He has a gastrostomy tube.

Question One

Of the following, which is the best treatment for Sammy's fever, cough, and breathlessness?

A. Acetaminophen for comfort from fever and antibiotic therapy administered through an IV that was inserted for that purpose
B. Acetaminophen for comfort from fever and antibiotic therapy plus an opioid for his breathlessness, administered through an IV that was inserted for that purpose
C. Acetaminophen for comfort from fever and an opioid for his breathlessness, administered through his gastronomy tube
D. Acetaminophen for comfort from fever, antibiotics, and an opioid for his breathlessness, administered through his gastronomy tube
E. All of the above

Correct Response and Analysis

The correct response depends on the goals of care, which must be clarified with Sammy's family and the healthcare professionals who know him best. Will Sammy be able to rally from this event and resume his previous state of health? Is his baseline quality of life acceptable to him and his family? What are the relative risks of each intervention and the anticipated benefits? Are the family's expectations realistic? Useful questions for framing the discussion include those in Juanita's clinical situation on page 22.

Regardless of other goals of care, enhancing the patient's comfort is always required. In every case, treatment of breathlessness is necessary. Treatment of fever is directed to relief of associated discomfort. Antibiotic use depends on the perceived benefits and burdens of treatment from the patient's and family's perspectives and on Sammy's determination of his quality of life. Considering the way Sammy may be feeling because of infection, it could be reasonable to start antibiotics. If infection is present, antibiotics may reduce his lung congestion, dyspnea, and fever, which would make him feel better. However, if using antibiotics results in more discomfort, such as repeated IV insertions, the hoped-for potential benefits of therapy should be carefully weighed against the potential burdens. Gastronomy-tube administration of medication is one alternative

and withholding antibiotics is another. Regardless of the goals of care vis-á-vis prolonging life, aggressive treatment of his dyspnea is required for comfort.

Opioids are first-line agents for relieving dyspnea. An opioid could be initiated, along with the antibiotics, followed by reassessment and escalation of symptom-

atic treatment as needed. Positioning may be helpful if he develops increased secretions. Reducing fluid intake could decrease secretions but may make them thicker, resulting in problems in mobilizing and clearing them.

Breathlessness

Breathlessness, or dyspnea, is a subjective phenomenon. Like pain, its severity is judged by patient self-report using assessment scales. Tools appropriate to the child's developmental level are necessary. Current options include a categorical scale such as "a little" or "a lot"; a faces scale, like the 0-to-10 Bieri rating scale for younger verbal children; or a 0-to-10 scale for older children. Specific scales for measuring dyspnea in children include pictorial scales for throat closing, chest tightness, and effort.[136] These scales appear to be most useful for children between 8 and 18 years of age. Correlation between ratings of dyspnea by hospice patients themselves and by their caregivers have been found to be "weak but significant".[137]

A decision to investigate the specific cause of dyspnea should be based on (1) whether the cause is obvious and treatment empiric, (2) the current overall goals of therapy, (3) the degree of invasiveness or potential discomfort associated with proposed diagnostic interventions, and (4) the relative benefits and burdens of potential interventions. For example, a chest X-ray is noninvasive and may lead to treatments that improve the patient's quality of life. A blood test is more invasive and distressing but also may lead to treatments that improve the patient's comfort, such as a transfusion to relieve anemia. On the other hand, a nasopharyngeal aspirate for a viral culture is invasive, distressing, and will not lead to therapeutic interventions that improve comfort; therefore, it should not be done. All tests and interventions must be considered in light of their potential impact on a specific child.

Nonpharmacologic interventions include physical measures such as position changes, cool cloths, fans to blow air toward the patient's face, and using the child's favorite scent in the room. Indoor air quality is

also a concern, as air pollution is now affecting even neonates.[138]

Pharmacologic measures include those directed at the cause of the dyspnea (eg, antibiotics to treat pneumonia) and nonspecific treatments such as oxygen and steroids. Pharmacologic interventions include the following:

- Supplemental oxygen that is titrated to comfort rather than oxygen saturation. If supplemental oxygen fails to increase comfort, it can be discontinued.
- Steroids can be useful for relieving symptoms associated with disseminated leukemic infiltrates or metastases to the lungs from solid tumors.
- Opioids are the main agent for symptomatic relief.
- Benzodiazepines can help relieve anxiety associated with dyspnea.
- Sedation at the end of life may be required for severely dyspneic patients when discomfort cannot be relieved by other means, even if the intervention may further compromise alertness or gas exchange.[81,82] (See www.aahpm.org/positions/sedation.html and UNIPAC 4.)

Respiratory Depression

Although the risk of respiratory depression due to opioids is much feared, it is grossly overestimated. Pain is a very effective respiratory-drive stimulant. When pain is relieved, obvious changes in the child's breathing pattern may occur that are unrelated to respiratory depression. Opioid-induced respiratory depression is preceded by increasing sleepiness and then a decreased level of consciousness and a slowly decreasing respiratory rate, with apnea following later. Apnea does not occur suddenly. Good monitoring by parents, nurses, and other caregivers should detect signs of respiratory depression, which

also may be an indicator of disease progression in a child with a life-limiting illness. Withholding the next opioid dose and instituting a dose-reduced opioid regimen are considerations, depending on the overall goals of care and the presence of distressing symptoms or their likely recurrence. It is important to understand that in the final phase of a terminal illness, irregular respiration is anticipated; therefore, withholding opioids or administering naloxone is inappropriate in most cases. Ensuring the child's comfort is the most important goal.

Guidelines for the use of naloxone are included here with the warning that very few instances exist in which its emergent use is indicated in the practice of palliative care. Naloxone should be avoided because it may precipitate acute, severe withdrawal symptoms; a pain crisis; and other adverse effects, such as pulmonary edema. Naloxone could be considered appropriate if a gross miscalculation of an opioid dosage causes a patient to be unresponsive with significant hypoventilation and puts the child at significant risk. In such instances, dilute 0.4 mg (1 mL) of naloxone in 9 mL of saline or sterile water to provide 40 mcg/mL. Then give 1 to 2 mL (or approximately 1 to 2 mcg/kg for greater accuracy) IV/SC at frequent intervals (every 2 to 3 minutes) until reversal of hypoventilation is achieved; be sure to avoid reversal of analgesia. Repeated doses will be needed until the time of peak effect of the opioid overdose has passed.

Myoclonus

Myoclonus is defined as involuntary muscle contraction during consciousness. Most healthy individuals experience myoclonus when drifting off to sleep. High dosages of opioids or long-term opioid therapy may precipitate myoclonus because of the accumulation of opioid metabolites. However, myoclonus can also be an idiosyncratic reaction to opioids unrelated to dosage or duration of therapy. When problematic, myoclonus can be treated by changing to an alternative opioid or by adding a benzodiazepine, particularly clonazepam (see Table 6).

Urinary Retention

Urinary retention can be caused by any opioid given by any route, but it occurs more frequently with regional anesthesia techniques such as epidurals. Interventions include

- applying external bladder pressure (Credé)
- starting a low-dose naloxone infusion for patients with a regional block to relieve urinary retention without reversing analgesia
- providing intermittent bladder catheterization (If catheterization is required more than a few times, consider an indwelling catheter.)
- adding bethanechol to stimulate effective bladder contractions.

CLINICAL SITUATION

Gina's Case Continues

Gina, the 5-year-old girl with relapsed leukemia, rates her pain as 6 out of 10 on the Bieri Faces Scale. You want to treat her pain but are concerned that she is likely to experience side effects from more pain medication. One of your colleagues advises you to be very cautious about using opioids because children are at increased risk of respiratory depression. You wonder if your colleague's concern is warranted.

Question Eight

When receiving opioids, are children at increased risk of respiratory depression and other side effects compared to adults? (Choose the best response)

A. Yes, all children other than teenagers have an increased incidence of respiratory depression with the use of opioids for pain.
B. No, the risk of respiratory depression from opioid use is the same for patients of any age.
C. No, the risk of opioid-induced respiratory depression in childhood is increased only for infants younger than 6 months.

Correct Response

The correct answer is C.

Seizures

Seizures may or may not result in distress for patients; however, clinicians should err on the side of treatment because seizures may be difficult for family members and other bystanders to witness. Because family members, friends, and staff will remember the events surrounding a child's death for the rest of their lives, a child's final days and hours need to be as peaceful and symptom-free as possible.

As death approaches, children with no previous history of seizures may experience them because of the disease process, fluctuations in serum glucose, calcium or sodium or other biochemical abnormalities, or hypoxia. In some cases, if no obvious disease-related cause exists, if death is not imminent, and if a work-up involves a single venipuncture or blood sample, it may be appropriate to check for treatable causes such as hypoxia.

Generalized seizures should usually be treated with anticonvulsants. After an acute episode has been treated with either lorazepam or diazepam, it may be helpful to load the child with parenteral phenytoin, valproate, or phenobarbital. Recurrences are usually suppressed with with lamotrigine, topiramate, or valproate.[139] Focal seizures can be effectively treated with (in alphabetical order because there are no comparative trials after a first seizure) carbamazepine, clobazam, gabapentin, lamotrigine, oxcarbazepine, topiramate, or valproate.[139]

CLINICAL SITUATION

Sammy's Case Continues

Sammy experiences some resolution of his cough and apparent respiratory distress with a combination of antibiotics and opioids. However, 4 days later his respiratory status deteriorates with no evidence of a potentially reversible cause. The frequency of his seizures increase despite his regular anticonvulsants: phenytoin and clonazepam. He is unconscious and death is thought to be imminent within the next 48 hours.

Question Two

Which of the following are the best approaches for managing Sammy's seizures? (Choose all that apply)
A. Because Sammy is unconscious and death is imminent, treating Sammy's seizures is inappropriate.
B. Although Sammy is unconscious, his seizures should be treated.
C. After drawing blood to check a level, Sammy could be reloaded with phenytoin.
D. Lorazepam or diazepam could be used in addition to his usual anticonvulsants for breakthrough seizures.

Correct Response and Analysis

The correct responses are B and D. The usual doses of Sammy's anticonvulsants should be increased to prevent further seizure activity. Because Sammy is close to death, he could be reloaded empirically with phenytoin without subjecting him to a blood test. He will also need medication to manage breakthrough seizures, which are likely to increase in frequency. Dosages of lorazepam or diazepam should be calculated on a mg/kg basis and should be readily available (see Table 6). Treating Sammy's seizure activity will benefit his entire family.

Insomnia

Children with serious or advanced illness may develop restlessness, insomnia, and sometimes nightmares, particularly when the environment is dark. The following nonpharmacologic strategies may be effective in reassuring an ill child and decreasing anxiety and nightmares:

- Move the child to familiar surroundings.
- Institute or resume a comforting nighttime routine.
- Read a nightly bedtime story.
- Furnish items that provide a sense of security, such as a favorite blanket or toy.
- Decrease unnecessary clutter to avoid potentially frightening shadows.
- Ensure adequate sleep by minimizing nighttime medications, labs, treatments, and monitors.

Simple insomnia may respond to hypnosis with school-age children.[140] Melatonin has proved useful in sleep disorders for neurologically disabled children.[141]

Somnolence

When starting opioid therapy, the child and family should be informed that the child may initially feel sleepier than usual. This effect generally subsides over several days. The increased sleep may be restorative, compensating for sleep interrupted by distressing symptoms. Occasionally sedation persists. Consider either disease progression or other contributing factors such as hepatic or renal dysfunction that prolongs the effects of the medication.[102]

If somnolence persists and is bothersome to the patient, but the intensity of pain precludes lowering the opioid dosage, adjuvants such as gabapentin for neuropathic pain or steroids for bone pain can be added. These agents act as coanalgesics and usually allow a reduction in the opioid dosage while improving or maintaining pain control. These coanalgesics can be added to counteract sedation, creating a larger therapeutic window for opioid administration. Alternatively or concomitantly, a psychostimulant like methylphenidate can be added.[142] Psychostimulants should be avoided for patients with a history of hypertension, seizure, anxiety, or agitation. Psychostimulants can also relieve depression more rapidly than tricyclic antidepressants.[143]

Delirium

Delirium, an acute change in mental status due to an underlying medical illness, is understudied, underrecognized, and undertreated in children. It can occur with people of any age, and some case reports indicate delirium in children as young as 6 months.[144]. Very little literature exists about delirium in children with life-limiting illness. In the adult population, the incidence of delirium near the end of life has been reported to be as high as 85%. A recent study of critically ill children in a pediatric ICU reported the incidence of delirium at 5%. This was considered an underestimation because only patients receiving psychiatric consultation were assessed for delirium. Of 877 admissions, 61 psychiatric assessments were performed and 40 cases of delirium were diagnosed.[145] (Please see UNIPACs 4 and 9 for a full discussion of the principles of diagnosis and treatment of delirium in adults.)

Studies have shown that the same mental-status changes that define delirium in adults also occur in children. There is some suggestion that certain mental-status changes occur more frequently in children, and others occur more frequently in adults.[146] When applying the diagnostic criteria to the evaluation of delirium for children, a few adaptations need to be made based on the child's age and developmental level. Inability to maintain attention, one of the key features of delirium, is manifest in infants and toddlers as difficulty engaging with others. It is manifest in children and adolescents as distractibility and inability to focus, which is the same way it manifests in the adult population. In addition, when orientation is tested in young verbal children, they can only be expected to be oriented to person and place. They have not developed enough of an understanding of months, days of the week, or dates to be able to answer questions about orientation to time.[144]

As soon as delirium is diagnosed, possible reversible causes should be promptly considered. When necessary, antipsychotic medications can be used safely and effectively in children to manage the symptoms of delirium. Fortunately, treatment with agents like risperidone rarely causes tardive dyskinesia or other movement

disorders in children.[147] Most of the patients diagnosed with delirium in the ICU study described above were treated with haloperidol or risperidone. Only two children who were treated with haloperidol experienced an acute dystonia, and the symptom responded well to treatment with biperiden. There were no other major adverse side effects of the medications. All of the children recovered from the delirium; five died from their underlying disease.[145]

It is very important to diagnose and treat delirium when it occurs in children because it can cause significant distress for this population. Having hallucinations and feeling disoriented can be very scary. Being unable to maintain attention can interfere with the ability of the child to participate in the activities that they normally enjoy. In addition to being distressing for the effected child, delirium can also be distressing for families and caregivers who see the child become confused or agitated. Furthermore, the burden is increased because it can be very exhausting to care for a child who is delirious. The identification and treatment of delirium can lead to improved quality of life for children, their families, and their caregivers.

Restlessness, agitation, and confusion can escalate with disease progression. Agitation may be a manifestation of undertreated pain, particularly for children with cognitive impairment, or these symptoms may be related to cumulative drug metabolites. Depending on the relative benefit-burden ratio, some clinicians initiate a trial of hydration.[148] A sedating medication may also be indicated to relieve agitation and restlessness. If agitation is mild or accompanied by anxiety, hydroxyzine or diphenhydramine may be beneficial, but it may also increase delirium, urinary retention, or confusion.[149] Because benzodiazepines can produce paradoxical agitation in children, a neuroleptic such as haloperidol or chlorpromazine may be a better choice. If sedation is the goal, chlorpromazine is more effective than haloperidol. Newer atypical antipsychotics may have a lower incidence of dystonia but have not been proved to be superior in other ways.[150]

Some patients who are receiving opioids, benzodiazepines, antihistamines, or barbiturates become agitated rather than sedated. This occurs more commonly in young children and may be related to the nonopioid medication, which should be discontinued or changed as the first intervention. If agitation occurs for a child taking an opioid or other centrally-acting medication, renal and hepatic function should be checked when appropriate. Reducing the opioid dosage or lengthening the dosing interval can be considered if pain is likely to remain under control.

Bleeding

Bleeding is a feared complication of some diseases and conditions. When bleeding is anticipated, the physician should gently and clearly discuss the possibility ahead of time with the child's family and caregivers. Information should include the possibility that a bleed may be the terminal event. Rapid responses should be planned in advance. For example, dark towels should be on hand to reduce the visual impact, and rapid-onset medication to sedate the child should be readily available. Midazolam can be helpful; doses begin at 0.05 mg/kg IV/SC and can be repeated every 5 minutes. Factors influencing the use of sedatives include the child's state of consciousness at the time of the bleed and the level of distress experienced by family members and staff when witnessing the event.

Helpful measures to prevent subsequent bleeds include tranexamic acid, 25 mg/kg/dose PO or 10 mg/kg IV four times a day. This drug has few adverse effects other than nausea. Potential renal impairment is a relative contraindication to be considered in light of the overall goals of care. Another potentially helpful agent is aminocaproic acid, 100 mg/kg/dose every 6 hours, with a maximum daily dose of 30 grams. The oral and IV doses are equivalent. Topical agents include a solution of aminocaproic acid applied as nose drops or as a mouth rinse. Thromboplastin tissue factor and apotonin can also be used.[151] It is generally advisable to avoid steroids and NSAIDs because they increase the risk of bleeding in children with potential bleeding complications.

Anorexia and Cachexia

Many children experience anorexia and cachexia throughout the course of their illness and, in particular,

at the end of life. Metabolic abnormalities associated with renal or hepatic failure, as well as the underlying disease such as cancer or HIV/AIDS, may be the underlying cause. Children may have received nutritional supplementation during the entire course of their disease, in some cases for the majority of their lives. Interventions usually focus on providing appetizing, small portions of the child's favorite foods and avoiding strong odors (see UNIPAC 4). Some medications, such as steroids and megestrol acetate, help stimulate appetite. Generally, steroids are not recommended for use solely as appetite stimulants because of their distressing side effects, particularly with prolonged use. Megestrol acetate is generally well tolerated if the child agrees to take the medication. Dosages for children range from 160 to 800 mg daily.[152]

When artificial nutrition and hydration are no longer meeting their therapeutic goals, the family may benefit from gentle exploration of their beliefs and wishes about withholding or discontinuing artificial hydration or nutrition. In some cases, family members initiate the discussion. In any case, withholding or withdrawing artificial nutrition and hydration can be an emotionally charged and ethically complex situation. Family members and all involved staff must be included in developing a plan of care, and all parties must be clear about the intent, ethical justification, and legality of such options.

Providing calories and fluids is viewed as a basic requirement by some caregivers, even when a child is dying. However, artificial nutrition and hydration are medical therapies; they are not required if they fail to accomplish the therapeutic goal. Physicians should provide information about the effects of either administering or forgoing artificial nutrition or hydration in terms of their effects on the patient's comfort. For example, providing artificial nutrition through a nasogastric tube may cause abdominal distension and emesis rather than comfort or prolonged life. In addition, nasogastric tubes can be quite irritating for some children and may interfere with the family's ability to interact with their child. On the other hand, enteral feeding may provide comfort for some children. Infants may enjoy sucking on a pacifier. Parenteral feeding can cause symptomatic distress in the face of cardiac or renal failure, and it can prolong life in some situations. However, the question remains whether prolonging life is a beneficial outcome of care, particularly in the face of permanent unconsciousness.

Compassionate discussions and exploration of values can help family members make more realistic and beneficial decisions about artificial nutrition and hydration. See the article "Forgoing medically provided nutrition and hydration in pediatric patients"[153] and UNIPAC 6 for more information.

Intestinal Obstruction

Malignant bowel obstructions are unusual in children because most childhood malignancies are hematologic in origin. Solid tumors (eg, osteogenic and Ewings sarcomas, brain tumors) are rarely present in a child's abdominal cavity.

Bowel obstruction may occur at the end of life, causing distension, cramping, nausea, and vomiting. Options should be reviewed in light of the goals of care, the child's life expectancy, and the child's quality of life with and without surgical intervention.

Surgical interventions should rarely be considered for many reasons, including the likelihood that the postoperative recovery period may exceed the overall life expectancy. Other interventions, such as the insertion of nasogastric tubes, are based on what is most acceptable and comfortable for the patient.

Options for symptom management include the following:

- Anticholinergic agents, such as glycopyrrolate or scopolamine, reduce secretions and bowel contractility. Antiemetics and systemic analgesics for colicky pain are important components of bowel-obstruction management. They can be delivered effectively at home through continuous IV or SC infusions.[154]
- Octreotride, a somatostatin analogue, has been used primarily for the adult population for the medical management of malignant bowel obstruction.[155] Limited data support its use in children to decrease intestinal secretions associated with certain hormonally active tumors.[156]

- Endoscopic stenting may be useful in some situations.[157,158]
- Use of nasogastric tubes should be based on the child's tolerance, which varies tremendously. Some children use nasogastric tubes for supplemental feeding throughout therapy, and others cannot tolerate them.
- A venting gastrostomy should be considered, if necessary, to control vomiting.[159]
- Surgery may be considered, depending on the child's life expectancy, individual preferences, and carefully identified benefits and burdens related to the child's quality of life.[160]

Eating and drinking are problematic for patients with bowel obstruction. With education, most families understand the burdens associated with continued attempts to feed their child enterally, particularly when they observe the abdominal distention, discomfort, and vomiting that can result. However, in some instances—depending on life expectancy, quality of life, continued therapeutic benefit, and the child's and family's wishes—artificial nutrition or hydration may be appropriate and consistent with the overall goals of care.

Assessing and Managing Refractory Pain and Other Symptoms

Assessing Refractory Symptoms

Most symptoms experienced by dying children are manageable with standard therapeutic interventions.[51] Although the incidence of refractory symptoms in children is low, vigorous attempts must be made to control them as soon as possible because of their severe impact on the child's and family's quality of life. Pain and other symptoms are classified as refractory when, despite the best efforts of clinicians with expertise in palliative medicine, adequate relief is unachievable while simultaneously maintaining consciousness.[81] Before pain or other symptoms are considered refractory, a physician with expertise in palliative medicine should review all preceding interventions and their outcomes and suggest any additional maneuvers that could be attempted within a reasonable time frame. Because specialists in pediatric end-of-life care are rare at the present time, telephone consultation may be necessary.

Symptom-related distress should be assessed from the perspectives of the child, family, and caregivers. Any proposed treatments must be acceptable to the child and to his or her parents.[82] Treatment is directed primarily by the child's self-reports of distress. When a child is unable to communicate, careful observations by a caregiver who is familiar with the child should guide treatment. In some cases, the child is treated for presumed distress. On occasion, the child is treated to alleviate the family's and staff's distress. For example, noisy breathing may be treated for an unconscious child at the end of life. Although the child is probably not aware of the noise, family distress is often quite extreme, justifying the use of anticholinergic drying agents such as scopolamine or glycopyrrolate.

Managing Refractory Pain and Other Symptoms

Sedation at the End of Life

In the past, sedation at the end of life has generally been referred to as terminal sedation. However, that phrase is being abandoned because of the erroneous implication that the intent of sedation is to hasten death. Misconceptions about intent may preclude parents from being comfortable with an option that may be in the best interests of their child.

Ethical Considerations Related to Sedation at the End of Life

Doing everything possible to free children from intractable pain and other intractable symptoms is a moral imperative. Sedation at the end of life can be initiated when symptoms remain refractory despite the use of appropriate intensive interventions.

Acknowledging the distress of a child's parents, other family members, and caregivers is a moral duty. Parents, other family members, or caregivers may believe an unconscious child is experiencing distress when moaning or breathing noisily. Unconscious children may exhibit certain body movements or positions (such as dystonic posturing or stridulous breathing in children with central nervous system pathology) that seem to indicate distress. It is important to acknowledge manifestations of the disease process and to describe their etiology to the child's family. In addition, the clinician should explain that it is unlikely the child is experiencing discomfort when unconscious. However, it is also reasonable to treat the child empirically for presumed distress. Treatment with analgesics or sedatives should be accompanied by explanations that, although the child's breathing pattern may not change, the medication will help to relieve any possible discomfort. Such explanations and interventions generally help alleviate the family's distress.

Honoring the child's appropriate role in making decisions is an essential component of ethical decision making. Children younger than 6 years generally lack the cognitive and emotional maturity to make informed choices. Children 6 to 12 years old are developing the necessary capacities for making medical decisions but may not be mature enough to provide the final decision. Child assent with parental permission and authorization

is considered necessary in this age group. Adolescents are generally presumed to be competent to make informed choices. Respectful appreciation of their wishes is key to making ethical decisions at the end of life.[15,47,161]

Effective communication and clear, compassionate explanations of the goals of sedation and its effects on the child are ethical imperatives. It is critical to be clear and honest when describing to the child (if appropriate, based on the child's developmental level and situation), to the family, and to other caregivers what measures will be taken to ensure sedation and how they will effect the child. The misconception that intensive symptom management, especially sedation at the end of life, is synonymous with euthanasia must be dealt with clearly and compassionately. The intention of sedation at the end of life is not to hasten death; rather, it is to relieve patients from suffering associated with uncontrollable pain and other distressing symptoms. A child's unresponsiveness when intentionally sedated is sometimes distressing for parents. Rather than immediately discontinuing the intervention, which is likely to return the child to unbearable pain or other severe distress, parents should be gently reminded and reassured that sedation is the only intervention that has provided relief for their child.

CLINICAL SITUATION

Lea

Lea is an 8-year-old Korean girl with end-stage rhabdomyosarcoma and profound breathlessness from extensive lung metastases. She conserves her energy by speaking in one-word answers, is profoundly sleep deprived, and has not had any oral intake for the past 24 hours. She rates her breathlessness as 10 out of 10.

On examination, Lea is bent over her bedside table, gasping for air, breathing at a rate of 48 breaths per minute. Within an hour of administration of a dose of parenteral morphine, Lea rates her dyspnea as 4 out of 10 and can speak in sentences. She asks to go to the playroom and is active and comfortable the entire evening.

The following day Lea reports severe and worsening dyspnea. The morphine is titrated in approximately 50% increments to symptom relief, and lorazepam is given for anxiety. As Lea becomes progressively more cyanotic and less responsive, her family cannot tolerate their inability to communicate with her. They attribute her deterioration to the morphine and ask to have the infusion stopped.

Questions for Reflection

Identify several issues that arise when clinicians are faced with symptoms that cannot be adequately controlled at the end of life without sedation.

Several Responses

- Have all other reasonable avenues been explored?
- What can be done when symptoms cannot be relieved while simultaneously maintaining consciousness?
- When should communication about management of refractory symptoms occur and with whom?
- What are some of the communication issues that need to be considered when sharing information about refractory symptoms with the patient, family, and staff?
- What ethical and medical or legal issues are associated with managing refractory symptoms and providing sedation at the end of life?
- What are the benefits and burdens of decreasing the medications used for sedation and control of dyspnea?
- Whose needs are paramount?
- Are the medications causing Lea's deterioration?
- How would you respond to Lea's parents' request?
- What might have prevented this situation?

NOTE. This clinical situation was adapted with permission from Kenny NP, Frager G. Refractory symptoms and terminal sedation of children: ethical issues and practical management. *J Palliat Care.* 1996;12(3):40–45.[82]

Preparing for Total Sedation

As soon as clinicians realize a patient may develop refractory symptoms, it is important to begin preparing the patient and family. Clinicians must initiate compassionate, recurring discussions about treatment options, particularly when a situation actually arises. After the physician, in consultation with palliative medicine specialists or someone with the required skill sets,[81,82] has determined a child is suffering from a refractory symptom, the option of providing a trial of sedation should be discussed with the healthcare team, the family, and the child (if developmentally appropriate). The physician should convene a team conference, initially without the parents' participation. The healthcare team needs to achieve consensus about the goals of care before the family's involvement. The healthcare team must also discuss and clarify concerns and possible misconceptions. Information is then shared with the family, whose questions and concerns should be fully addressed. Caregivers and family members should understand the following:

- The child cannot be kept both awake and in a relative state of comfort.
- Sedation at the end of life is the only effective means of providing comfort at this stage.
- Such sedation will result in loss of consciousness.
- Family members and staff should feel free to voice any concerns at any time; concerns should be fully addressed with compassion.
- If sedation at the end of life is chosen, all involved staff members must understand the principles involved and follow established guidelines.[81,82,162]

After the presence of a refractory symptom has been confirmed through consultation with a specialist in palliative medicine or someone with similar expertise, and the process of sedation in the terminal phase of life has begun, it generally continues until the child dies. Occasionally, a family may request that sedation be withdrawn so they can interact with their child. They should be reminded gently that sedation was started because the child could not be kept both conscious and free of significant distress. Withholding medication or decreasing the dosage is likely to result in the return of distress without improving the child's capacity to interact. It may be helpful to reemphasize the decision-making process and goals of care with the family. If the family continues to request the withdrawal of sedation despite repeated explanations and reassurance, a trial of lower dosages of medication is warranted, with the understanding that sedation will resume if the child appears distressed.

Even in a sedated state, children can live much longer than expected. Because caregivers may become very weary, getting adequate rest, food, and exercise and talking with a trusted listener are essential to the well-being of both the family and the staff.

Pharmacologic Principles for Providing Sedation at the End of Life

- Review the medication history to determine if the child has experienced any significant adverse effects, such as paradoxical agitation, with lorazepam or other benzodiazepines. Although agitation may be dose related, after it has occurred a different medication without the negative associations is preferable.
- The physician should have experience with the sedating drug in terms of dosing and titration.
- The addition of a sedative, such as a neuroleptic (chlorpromazine), a benzodiazepine (midazolam or lorazepam), or a barbiturate (phenobarbital) is indicated. When there is no history of adverse effects with a class of drugs, the selection of a sedating agent can be based on route availability, sedating properties, duration of action, and cost.
- Opioid therapy should continue at appropriate dosages. However, using an opioid as the sedating agent is unlikely to be successful, particularly when the child is opioid tolerant. The combination of opioid tolerance and heightened sympathetic stimulation because of symptom-related distress is likely to inhibit sedation. Escalation of opioid dosages with the intent of causing sedation may only exacerbate unwanted side effects, such as myoclonus, seizures, and delirium.
- Meperidine should not be used for sedation at the end of life or for relieving chronic pain because of

its potential for neurotoxicity from its metabolite, normeperidine.

- The physician should reassess the effects of sedation frequently and watch for breakthrough symptoms and adverse effects.
- When rapid titration is required to relieve severe distress in the terminal phase of life, parenteral dosing should be used, if possible.
- The clinician should seek consultation from individuals with expertise in caring for children at the end of life. (See AAHPM's position statement on palliative sedation at www.aahpm.org/positions/sedation.html.)

CLINICAL SITUATION

JB's Story: Communicating Bad News

JB is a 3-year-old boy who is experiencing difficulties with coordination and activity levels compared to his preschool peers. Genetic testing confirms a diagnosis of Duchenne muscular dystrophy (DMD). JB's parents are married and have one other child, a healthy 1-year-old daughter, Emily. Both children were the result of planned pregnancies. There is no family history of DMD and the family has never known a child who died. JB's parents have been invited to the physician's office to learn the diagnosis. Discussion with JB is planned following the meeting with his parents.

They are seated next to each other, about 2 feet from the physician. A social worker and JB's primary nurse sit nearby. Facial tissues are on a table, within reach of the parents.

Question One

When initially breaking bad news about the diagnosis to this family, which of the following approaches is likely to be most helpful? (Choose all that apply.)

A. Provide complete written and verbal information on the latest research on muscular dystrophy and give them a list of local support groups.

B. Reassure the family that a cure will be discovered before JB is threatened with death.

C. Advise the couple to request sterilization to avoid putting future children at risk for the illness.

D. Ask what they know about muscular dystrophy, briefly explain the illness, pledge your continued support, and agree to speak again in the next few days.

E. When talking with JB after the initial conference, address his fears and concerns and answer his questions in clear, age-appropriate language.

Correct Response and Analysis

The correct responses are D and E. Although maintaining a hopeful posture is desirable, ensuring that patients and families have accurate, realistic information is vital—it helps them make decisions consistent with their values. For more information about delivering bad news, see UNIPAC 5.

One of the first steps is to assess what JB and his family understand about the illness. Information about the illness, treatment options, the likely prognosis, and the probable effects of the illness on JB and his family should be provided gradually over the course of several visits. Such information is likely to help JB and his family develop some sense of control over an otherwise chaotic situation. However, neither the patient nor the family will be able to absorb much information during the first encounter. Breaking bad news should be viewed as a process, the first step of which is to name the illness. After hearing the diagnosis, shock and disbelief usually interfere with the patient's and family's ability to absorb other information. Throughout the entire multisession process of sharing information, it is important to continuously assess the patient's and family's understanding of information presented. Providing a tape of information-sharing meetings can be helpful.[163]

Making pronouncements about a family's future reproductive plans is not appropriate; however, providing information about the heritable nature of the disorder

is important. Family planning can be addressed in the following days or weeks, if the parents would like to discuss it.

Children know something is wrong when they are not functioning as well as their peers and must undergo numerous tests.[164] Determining what the child understands about the illness is a necessary first step. Helping the child express anxieties, fears, and other concerns is good medical practice.[165] When a child seems to be willing and able to talk about the illness, a nonthreatening and useful approach is to start by asking, "Can you tell me what you think is happening to your body?" Children then benefit from information tailored to their needs and degree of understanding,[166] which can only be accomplished by asking questions such as these:

- What do you want to know about what is happening to you?
- I wonder if there is something you are scared of?
- Is there something you wish you could talk about?

Children may be afraid to broach certain topics out of fear they will alienate or upset the adults upon whom they rely. However, children should be encouraged to ask questions with clear statements such as "Please don't be afraid to ask questions. When you ask questions, it helps me understand what you want to know." Again, the child's questions need to be answered directly in developmentally appropriate language.[10] Play therapy provided by a skilled clinician, such as a psychologist or child-life therapist, can help elicit the concerns of children with limited verbal skills or shyness.

The Case Continues

Two days later JB's parents meet with the physician again, this time with many written questions. They look as if they haven't slept well and have been crying. They are angry and defensive and are arguing with each other.

Question Two

When interacting with the couple, which of the following issues should be emphasized? (Choose all that apply.)
A. The disease is inherited through the maternal genes. To be supportive, the father should tell the mother that he forgives her.
B. Provide information about support groups for families facing this illness.

C. Ask them what has been happening at home over the past 2 days.
D. Emphasize that JB has a long time left to enjoy an excellent quality of life, which will be enhanced by family cohesiveness and meticulous symptom management.
E. Reassure them that you will be there to provide medical care and to help them adapt to this new reality.

Correct Response and Analysis

The correct responses are B, C, D, and E. Defusing blame is a critical part of providing information and support, particularly when dealing with inherited disorders. Parents do not want their children to die prematurely or to experience loss of function and disturbing symptoms; they want the best for their children. Neither parent purposely caused JB's illness, so there is nothing to forgive. When an illness becomes evident, parents must find new ways of "doing their best."

Support groups provide opportunities for patients and families to meet others who are adapting to an illness and creating solutions, thus decreasing their sense of isolation and helplessness. In support groups, experienced families help others by sharing ways of ensuring the best possible quality of life for their children. Because not all families experience support groups as helpful, other support modalities should be offered, for example, family counseling or "parent match-up" with parents facing similar experiences.

Demonstrating to families that you are aware of the illness's impact on the whole family and are willing to discuss it will encourage them to seek advice and support. Often, listening may be the most therapeutic intervention you can offer. Parents frequently need reassurance that they are doing a good job and have been good parents.[167,168]

Introducing Palliative Care

JB is now 6 years old. He is having trouble climbing up the stairs to his bedroom. He has no friends at school and his teacher chides him for being "lazy and slow." JB and his sister, Emily, have become very close. JB's parents have not yet participated in a support group and are feeling angry and isolated. They have chosen not to share

JB's diagnosis with school personnel. JB's mother seems withdrawn, and his father has adapted to the painful situation by traveling even more for his job. Although they mention feeling supported by their religious community, they also feel lost and are searching for ways to explain what has happened to them. Supportive and palliative care interventions have never been mentioned.

Question Three

Which of the following may be helpful interventions at this time? (Choose all that apply)

A. Family grief counseling

B. Physical therapy and occupational therapy for JB

C. Supportive, open-ended conversations with JB about his hopes, fears, frustrations, and ideas about his future

D. School interventions to educate the teacher, counselor, and class about JB's condition and how they can support him

E. Child-life interventions and volunteer support for JB and his sister

F. A practical assessment of home life by a social-service professional

Correct Response and Analysis

All of the responses listed above are correct. Palliative interventions provided by a team of healthcare professionals should include physical, emotional, spiritual, and social care to support the child and family. Palliative interventions and life-prolonging interventions should not be viewed as mutually exclusive.[4,6] The best approach is to incorporate supportive and palliative care interventions from the time of diagnosis, along with life-prolonging therapies for as long as is appropriate. This approach is the most likely to meet the multifaceted needs of families facing the ongoing crisis of a child's life-limiting illness.

Grief and Guilt

Grief occurs with any loss, including the loss of an expected healthy future.[169] The short- and long-term effects of unaddressed grief can be devastating, severely diminishing both the child's and family's quality of life during the remainder of the child's life and then affecting the family's bereavement.[170] Over time, as JB's functional capacity deteriorates, losses will accumulate; thus, early

and ongoing grief counseling will likely be extremely beneficial for JB and his family.

The parents of children with life-threatening conditions commonly experience guilt. They may ask themselves questions such as[16]

- Did I cause this during the pregnancy?
- Did I pass it on in my genes?
- Did I notice the problem soon enough?
- Will this happen to my other children?
- Am I being punished for something in my past?
- Am I ignoring the needs of my healthy children?
- Am I the only person in the world facing this kind of problem?
- Am I doing a good enough job of treating the symptoms?

Guilt needs to be acknowledged and discussed. Clear and supportive explanations can help prevent the kinds of self-destructive behaviors that JB's parents are exhibiting. For example, the physician can sit at eye level with JB's mother and say, "Some parents of children with DMD believe they are responsible for their children's condition. I wonder if you blame yourself for JB's condition?"

The physician should then wait for her response. If she agrees or if her response is uncertainty or silence, the physician can follow up with statements such as. "DMD is an inherited condition, but neither you nor your husband wanted your child to have it. With no family history, there is no way you could have known that he might get it. There would have been no reason for you to have a test to detect DMD. The best thing you can do now is to help JB live as fully as possible, to support him as a person, and to keep yourself and the rest of your family healthy and happy. I will help you accomplish these goals." After each statement, the physician should wait briefly for a response.

Even very young children who are ill recognize that something is wrong.[17,20] If their fears remain unaddressed, they will accumulate over time. JB and Emily need a caring adult to listen to their needs and concerns; however, they will only trust someone who is honest with them. Euphemisms are likely to be misunderstood by preschool and school-age children, who think concretely.[171] Simple, honest, direct language is the best way

of communicating. Instead of asking "How are you?" (answer, "Fine"), ask specific questions:

- What do you do that is fun at school?
- What are some of the things that are bothering you at school?
- Are you having problems walking up stairs?"

Physical and occupational therapy can help JB adapt to his physical limitations by showing him new ways of accomplishing tasks, helping him maintain his range of motion and flexibility, and helping to assess and treat pain.[172] Child-life interventions encourage children to express emotions through storytelling and nonverbal means, such as art and play therapy.

Support for siblings is critical.[173] Siblings spend more time together than do children and parents or any other family pairing. With appropriate emotional support, Emily may experience positive effects from JB's illness. Several studies demonstrate that siblings who were allowed to participate in caring for an ill sibling and who received effective emotional support experienced greater self-esteem and were more effective as adults than were children without the experience of a personal tragedy.[31,170,174] However, when parents are unable to provide adequate emotional support for their children, other adults must fill the gap. Adults who know the children well, along with reliable and consistent volunteers, can be a tremendous help.

Volunteers can also serve as sounding boards for parents who experience ambivalent feelings toward their ill child or medical interventions. The volunteers must be well trained, be free from personal unresolved grief issues, and have access to necessary family information to optimally help the family.

School and Community Issues

Schools can serve as valuable support systems or can become devastating social milieus for an ill child. The medical community should provide educational and supportive interventions that will benefit both the child and the school. For example

- Help the school develop a plan for responding to the child's medical emergencies—advance planning will decrease anxiety experienced by school personnel and increase their willingness to allow the child to attend school as long as the child desires.

- Explain that maximizing the child's potential will help them develop appropriate educational plans.
- After discussion with JB and his family, and only with their approval, explain the child's illness to school personnel and students. Other healthcare team members (such as the child-life worker, social worker, primary nurse, or psychologist) can talk with the ill child's peers. Schoolmates may fear JB, thinking that his condition is contagious. Clear, age-appropriate discussions can facilitate peer transition from a cycle of avoidance and harassment to one of support and caring.

Communicating with Children

During a conversation with his physician, JB (now 7 years old) reports several fears. He is afraid that he won't be able to go places, he wants to go to a different school where people will like him, and he wants to know if he will die. He also wants to know what he did to make his parents so sad.

Question Four

When responding to JB's concerns about dying, which of the following could be helpful? (Choose all that apply.)

A. Tell him that he is silly to worry; he will be able to go many places for a long time.
B. Explain that you don't know what will happen to him yet.
C. Reassure him that he is not dying now, but everyone dies sometime.
D. Explain that his illness will not allow him to live as long as many people, but he is not expected to die soon.

Correct Response and Analysis

The correct response is D. Because JB's mobility will be progressively limited, it is important to help him set a timetable to accomplish his goals and wishes. JB may have several wishes that are attainable; however, none of his important goals will be identified unless he is allowed to express them. JB's family and the medical community should make concerted, ongoing attempts to help him attain his goals. Wish-granting organizations are wonderful, but families, in cooperation with the medical community, religious groups, and other local organizations, can often help ill children accomplish a series of goals.

JB's worries are realistic. Instead of discounting his concerns, they should be acknowledged and addressed.[175] Changing schools is usually difficult. It is practical and reasonable to negotiate a time limit with JB for educating his classmates and determining if acceptable changes in their behavior will occur. If this strategy fails, school systems are often willing to "bend the rules" to accommodate the needs of a child with a life-limiting illness. What is not requested cannot be accommodated.

Although it is true that JB is not dying now and that everyone dies sometime, this response, or saying you don't know what will happen, discounts the seriousness of his question and ignores the possibility that he may have other, completely different concerns. Responding by saying, "What made you ask me that today?" may elicit responses such as, "My legs are hurting much more lately" or "I heard my mom and dad talking about someone's funeral." These issues can then be addressed appropriately. JB may be asking about his life expectancy so that he can gauge when to start giving possessions away or when to make known his preferences about funeral rituals. It is not unusual for children to have very specific requests regarding their funerals (eg, clothing they wish to be buried in, songs to be sung and by whom, prayers to be read). Teenagers commonly want to leave material legacies, such as poems they have written or videos of themselves. Affirming that JB is going to die earlier than his friends but not anytime very soon is honest and compassionate.[176]

Children notice their parents' grief and realize that their illness is one source of family distress. Denying the relationship between the illness and the parents' behavior will be perceived as dishonest. However, just as the parents did not intend to cause the illness, neither did the child ask to have it, so no fault should be assigned. Therapeutic responses include acknowledging the parents' sadness and their need for help, providing reassurance that they will get it, and providing reassurance that the child is a good person who deserves happiness.

Unfortunately, in the throes of their own grief, parents often neglect their other children. For the siblings' well-being, as well as for the well-being of the ill child, siblings should be included in discussions and in providing care whenever possible. They can serve as valuable resources for the ill child and the parents. Playing an active role in caring for the ill child also benefits siblings during bereavement.

The Case Continues

JB is now 10 years old and wheelchair-bound, but he is still able to breathe well on his own. With counseling and support, his home and school situations have improved greatly. His mother is attending a parent support group, and JB and Emily go on outings with a volunteer every other weekend. JB is talking about going to college but also talks about dying. Emily wants to know if she will still be a sister when JB dies.

Question Five

Which of the following are effective ways of supporting the children during their role crises? (Choose all that apply.)
A. Tell JB not to dwell on dying but rather to plan for the future and hope for a cure.
B. Ask JB how he feels about dying.
C. Before responding to Emily's questions, ask about the family's beliefs regarding an afterlife.
D. Reassure Emily that she will always be a sister and has been the best sister JB could have.

Correct Response and Analysis

The correct responses are B and D. Simultaneously planning for the future and for death is a healthy adaptation to uncertainty. Physicians can support JB by encouraging realistic goals, soliciting his preferences, and facilitating family discussions about death. Waiting until JB is closer to death to ask about his preferences may deny him the opportunity to express them at all. Both the medical community and parents often wait too long to initiate discussions about death-related issues; the child becomes incapacitated before making his wishes known. In addition, ignoring the fact that death looms large in the life of a chronically ill child denies both his personhood and his existence as an individual.[4]

Although children may share practical information about death-related issues (eg, songs to be sung at their funeral, videos they want to make), encouraging them to discuss more intimate issues about their death and its meanings can help resolve potential crises. Asking "How are you feeling about dying?" may elicit a response such

as "I am afraid." The physician can follow by saying, "Of what?" which may result in the response "Being forgotten." The caregiver should follow such a response with a discussion of how to leave a material legacy. If the caregiver's inquiry elicits a response such as "burning in hell," the child should receive spiritual counseling. A response of "suffocating" should be followed with information about the management of dyspnea and reassurance that the team will do everything possible to keep him comfortable.

Understanding the family's beliefs about an afterlife is important before discussing what happens to people when they die. However, Emily may be asking something entirely different. She probably wants reassurance that JB will continue to be her brother after he dies and that she will always be his sister. She is also likely to need reassurance that she has made a positive contribution to JB's life and has been a good sister. Such reassurance will help Emily adapt in a healthy manner to her ultimate loss.

The Case Continues

JB is now 17 years old and has been hospitalized three times for pneumonia in the last year. His speech is becoming very difficult to understand and he is no longer able to participate in physical activities. He is short of breath and chokes often. He rates his breathlessness as 5 out of 10. JB is frustrated by his communication difficulties. His physicians discuss mechanical ventilation with his parents, who agree to a tracheostomy and chronic ventilation. JB objects, saying he is not happy with his quality of life and does not want to prolong it.

Question Six

Which of the following statements about JB's rights in medical decision making are correct? (Choose all that apply.)

A. He is underage and his decisions may have no legal standing.
B. He has the right to influence his medical care, but his opinion is not determinative unless he is declared a mature minor.
C. He is incompetent to decide because the consequences of his decisions are not clear to him.
D. His parents' opinions regarding medical care must be sought.

E. He is depressed because he has been so ill; therefore, his opinions should be discounted.

Correct Response and Analysis

The correct responses are A, B, and D. Legally, JB's care is determined by his parents, as he is a minor. Ethically, as the person who is most intimately affected by the illness and as an older child, JB's opinions and preferences should play a large role in the decisions made. Temporary incapacity to make decisions is not sufficient justification for permanently discounting a child's opinions and preferences. Every effort should be made to determine JB's decision-making capacity and, if it is temporarily diminished, to restore it as quickly as possible. The importance of calm, repeated discussions about the expected course of an illness, potential treatment choices, and their likely outcome cannot be overemphasized.[177] If necessary, hospital ethics committees can help with achieving some consensus about the goals of care and making decisions.

Depression can impair a child's ability to make judgments. Because pain and other distressing symptoms contribute to depression, meticulous symptom control is required. Upon relief, a reassessment of previous decisions may be warranted. A trial of antidepressants may also be warranted. Anecdotally, methylphenidate may be particularly useful when the need to make decisions does not allow the several-week trial necessary for more standard antidepressants.[143,178]

The Case Continues

Morphine is started at 5 mg orally every 4 hours, with rescue doses as needed to relieve JB's dyspnea. After a couple of days of stable opioid dosing, he is switched to a long-acting morphine preparation of 15 mg twice a day with rescue doses of 3 mg of immediate-release morphine for breakthrough symptoms. Amitriptyline is started at bedtime to relieve depression, reduce secretions, and improve sleep.

Oxygen provides no benefit and is discontinued. JB agrees to a trial of nighttime continuous positive airway pressure (CPAP) and ventilation with a portable negative pressure ventilator (cuirass), but he finds them too burdensome and they are discontinued. Family coun-

seling sessions emphasize supporting JB and letting him know how important he is and how much the family loves and respects him. He spends extra time with his school friends. He is now much more comfortable; nevertheless, his request to forgo mechanical ventilation continues.

Question Seven

When assessing JB's decision-making capacity, it is important to determine that which of the following is correct. (Choose all that apply.)

A. His understanding that death is permanent.
B. His understanding of the risks and benefits of alternative treatments
C. His ability to balance a checkbook
D. His ability to live independently from his parents

Correct Response and Analysis

The correct responses are A and B. A determination of capacity to make medical decisions differs from a determination of capacity in other realms and also differs from "competence," a legal term that is age dependent (the person must be over 18 years of age). Medical-decision-making capacity rests solely on the ability to understand alternative treatments and their potential risks and benefits. If one of the potential risks is death, the person must clearly understand the irreversible nature of death. When determining medical-decision-making capacity, capacity in other realms is irrelevant (eg, the ability to live independently or balance a checkbook). For more information on medical-decision-making capacity, see UNIPAC 6.

Ethical and Legal Issues: Withholding and Withdrawing Life-Sustaining Interventions

JB's parents and physicians finally agree that he is not making a rash or immature decision based on a hurried or unrealistic assessment of his situation. They agree to forgo mechanical ventilation. Intensive symptom control measures will be continued and escalated as needed to ensure his comfort. The physician suggests a referral for hospice home care, and JB and his family agree. After JB and his family choose a hospice program, they work with the hospice team members to develop an interdisciplinary plan for home hospice

care that meets their specific needs. See UNIPAC 5 for more information about interdisciplinary teams.

Managing Refractory Pain and Other Symptoms: Sedation at the End of Life

JB asks what will happen if he is unable to breathe. He has heard about sedation at the end of life and asks if it applies to him. Such sedation involves the use of sedatives titrated to the point of unconsciousness; it can be used when all other measures to control pain or other symptoms (eg, dyspnea) are unsuccessful while simultaneously maintaining consciousness. See the section "Pharmacolgic Principles for Providing Sedation at the End of Life" on page 85.

Question Eight

Which of the following is a correct statement about sedation at the end of life? (Choose all that apply)

A. It is illegal because it is equivalent to euthanasia.
B. It is illegal if used for minors.
C. It is achieved by escalating the dosage of a sedative medication until the patient stops breathing.
D. It is an ethical intervention for patients with distressing refractory symptoms.

Correct Response and Analysis

The correct response is D. When opioids are titrated to relieve dyspnea, the goal is good symptom control while maintaining consciousness. Sedation at the end of life is rarely needed. Sedation is considered only when all other methods of controlling symptoms have failed to provide comfort. At that point, sedation may be initiated. The goal of sedation is to keep the patient comfortable until death occurs from an underlying disorder. Decisions about initiating or forgoing artificial nutrition or hydration are made separately. However, at the end of life, such therapies generally contribute to symptom distress and prolong the patient's discomfort rather than improve quality of life. The goal of sedation at the end of life is relief from suffering, not death.[81,82]

When respiratory compromise is the cause of impending death, relief of respiratory distress may inadvertently hasten death. However, because death is not the intended result and death is not intentionally hastened (most people live hours to days after sedation is initiated), sedation at the end of life is not

the equivalent of physician-assisted death or euthanasia. It is a legally and ethically permissible intervention. The patient's age is irrelevant; sedation at the end of life is appropriate for infants, children, and adults when pain, dyspnea, or other distressing symptoms are otherwise uncontrollable.

Anticipatory Grief

JB asks Emily to make a video of him and starts giving away his favorite CDs, books, and other items that he values. He also makes decisions about what he will wear at his funeral and who he will invite. He chooses prayers, music, and songs for his funeral and asks to see his minister. JB wants to continue going to school for as long as he can. He tells his friends he doesn't have much time left and begins inviting some of them to his funeral.

Question Nine

Which of the following is the correct interpretation of JB's interest in and concern about his impending death and funeral? (Choose all that apply.)

A. It is a normal, healthy response.
B. It is a symptom of increased depression and requires treatment.
C. It is a dramatic attempt to gain his parents' attention.
D. It is a symptom that he is romanticizing death.
E. It is a symptom of a spiritual crisis.

Correct Response and Analysis

The correct response is A. As children become aware of impending death, they often give away their belongings. Some children, particularly adolescents, want to create material legacies. Ensuring they have the opportunity to do so allows them to achieve final tasks of self-actualization, which are crucial for their well-being as well as for the well-being of survivors. Spiritual growth is common as life draws to a close. Far from representing a spiritual crisis, his request to speak to his minister may signal a need for affirmation or a desire to share new insights about life and death. Effective symptom control is essential during this time.

Sites of Care Delivery

JB develops pneumonia. He accepts antibiotics and cough suppressants but continues to decline ventilation. Oxygen now enhances his comfort. His fevers are controlled with ibuprofen, and his cough and dyspnea are relieved with opioid therapy through his gastrostomy tube using 15-mg sustained-release morphine twice a day and 3-mg immediate-release morphine every hour as needed for breakthrough dyspnea. However, his lungs do not improve, and JB realizes he will not survive. He says he always knew this day would come. JB's parents discover they cannot tolerate his dying at home. No palliative care or hospice inpatient facility is available. JB declines a nursing home, but he is willing to return to the hospital where he received care in the past.

Question Ten

Which of the following is a correct statement about entering the hospital? JB and his family must (Choose all that apply)

A. agree to mechanical ventilation
B. realize the hospital's main goal is saving lives; thus, JB is likely to be ignored if he chooses to forgo life-sustaining therapy
C. accept restrictions on visiting hours
D. forgo care by hospice personnel
E. understand they can continue following the hospice plan of care and receiving care from hospice team members.

Correct Response and Analysis

The correct response is E. JB and his family can decline any and all forms of life-sustaining treatment, regardless of care setting. However, JB's family should ask about the hospital's policies regarding end-of-life care. JB's status as a minor may invite more rigorous scrutiny to ensure that his "best interests" are being upheld. If the hospital's treatment team or any hospital personnel doubt that his best interests are being served, the palliative care team and hospice program's medical director and nursing staff should discuss the goals of care with JB's hospital-based physicians and nurses. If necessary, an ethics consultation can be requested.[177] Generally, ethics committees uphold the rights of a terminally ill child and family.

When patients with life-limiting illness receive care in an acute care hospital, their care may be viewed as a lower priority because cure is no longer possible. However, when a patient is dying, the goals of care appropriately shift from cure to providing excellent symptom control and ensuring a peaceful death. Achieving these goals will provide an affirming conclusion to JB's life by honoring his wishes and helping him and his family achieve their goals. Receiving less than attentive care is unacceptable in any care setting; therefore, palliative care specialists must support hospital personnel and educate them about the broader goals of medicine, the goals of palliative medicine, the transition from curative to palliative care, and the provision of palliative interventions. In many hospitals, palliative care specialists are part of the hospital staff.

Patients can enter a hospital for the final phase of life under the auspices of a hospice program's plan of care. Some hospice programs operate freestanding inpatient facilities; others contract with nursing homes or hospitals to provide end-of-life palliative care in a designated area staffed by hospice personnel. Others contract with a hospital or nursing home for flexible beds. This more common arrangement permits admission of hospice patients to any bed in the facility. Care is provided by inpatient-facility personnel and the hospice care team.

A facility's policies may become problematic in the flexible-bed situation. The hospice and palliative care program may have to arrange exceptions to rules that limit visiting hours or the ages or number of visitors. Hospitals have become more aware of the need for unlimited visiting hours for children and often have no difficulty accommodating such requests. Often, they can arrange for bedside availability of medications and allow family members to administer them, which optimizes response time for symptom control and ensures that patients and their family members retain as much control as possible. Unfortunately, bedside availability of opioids is not a possibility at all facilities. PCA pumps may provide the next best alternative solution.

Patient and Family-Centered Death in the ICU

Critically injured children who will not survive their injuries usually die in the ICU. These children, as well as critically ill premature neonates, should be provided with alternatives to isolated, highly technical deaths. A patient- and family-centered death allows the child to return to the role of son or daughter rather than remaining a patient during the last moments of life. Discussing the following maneuvers with the family can help ensure that dying children and their family members experience a patient- and family-centered death, whether death occurs in the ICU itself or in a separate space within the hospital:

- Remove monitors, urinary catheters, nasogastric tubes, and other clinical devices, but maintain an IV site in case it is needed for symptom management.
- Continue other supportive interventions, including music and touch, according to the family's wishes.
- Discontinue monitoring of vital signs and weight.
- Discontinue laboratory investigations and medications that do not provide comfort, such as antibiotics, enteral or parenteral feedings, and usually intravenous fluids.
- Discontinue mechanical ventilation.

The entire family and support community (if desired by the patient and family) can be invited to honor the child. Parents may want to observe or participate in rituals such as bathing the child; dressing the child in clothing the family's chooses; making hand- and footprints; preserving a lock of hair; joining in prayers, blessings, or chanting; taking turns holding or rocking the child; and taking video or still photographs. Staff are generally receptive to these supportive measures if they are aware of the family's wishes. Inexperienced staff may need assistance soliciting the parents' wishes. Giving examples of requests made by other families is one effective way of eliciting their wishes.

Pharmacologic paralysis should be reversed if previously used. When everyone is ready, the ventilator or pressors may be discontinued abruptly, with removal of all unnecessary medical equipment. An IV site should be maintained (or SC access established) to

rapidly manage any dyspnea or pain that occurs as the child dies. Many clinicians premedicate the patient with a sedating agent or an analgesic before discontinuing ventilatory support; others wait for signs of distress, which are not inevitable, and respond rapidly with parenteral means if symptoms occur. A compassionate patient- and family-centered death is possible, even for children who die in the ICU.[179] (See UNIPAC 9 for more information about the withdrawal of mechanical ventilation.)

The Clinical Situation Continues

JB asks Emily to stay with him at all times and requests a visit from his best friend, George. George's parents are hesitant; they are concerned that exposing George to a dying person may cause him harm. Although Emily will miss school, her parents allow her to remain with JB. At the suggestion of the hospice counselor, Emily brings photo albums from home and looks through them with JB, reminding him of good times they have had and assuring JB that he is loved and will be remembered.

JB requests a visit from the hospice chaplain and, after consultation, states that he is ready to die. He reassures his parents and sister of his love. He thanks them for their love and asks that they always remember him.

Question Eleven

When assessing whether the above psychosocial responses are adaptive or maladaptive, it is important to realize (Choose all that apply.)

A. George's parents should prevent him from visiting; it will only add to George's pain to see JB during this phase of his illness
B. children benefit from the opportunity to achieve closure; however, they should not be forced to visit if they do not want to
C. Emily should go to school to receive support from her friends and begin adapting to her new life without JB
D. the time that Emily spends with JB may be invaluable to her when adapting to life without her brother
E. JB's statement about being prepared for death is a veiled request for assisted suicide.

Correct Response and Analysis

The correct responses are B and D. Children grieve and benefit from the opportunity to achieve closure by spending time with the ill person, expressing feelings, remembering old times, providing care, and giving reassurances of remembrance. However, this must not be forced on a child who does not wish to participate. Studies indicate that bereaved siblings who provided care for a deceased child often develop an enhanced sense of self-efficacy and improved skills for adapting to loss.

It is not maladaptive to prepare to die. JB is manifesting self-awareness and maturity beyond his years, which is not unusual in children with chronic, deteriorating illnesses. His readiness to die provides an opportunity for reflection and contemplation about what is important in life.

Managing Symptoms at the End of Life

JB feels weaker and is more short of breath. He is sleeping more and is less attentive to his companions. He wants to discontinue the oxygen because it is irritating his nose and he dislikes the feeling of the mask on his face. He requests more morphine because it alleviates his sensation of suffocating even though it makes him sleepier. The resident physician expresses concern, but the palliative medicine consultant provides education about the appropriateness and acceptability of the intervention. JB's morphine is titrated upward in 50% increments for relief of his breathlessness. He becomes nonverbal. Whenever JB wakes, his parents, sister, and grandparents tell him they love him and that it is OK for him to go when he is ready. He smiles at them and closes his eyes. Six hours later, JB stops breathing.

Question Twelve

Which of the following responses best completes this sentence: By taking away the oxygen and increasing the morphine, the treatment team

A. committed euthanasia—the timing of the medication and the death prove the relationship
B. alleviated symptoms and provided an opportunity for a peaceful death to occur
C. caused his death by stopping the oxygen, which is always required for shortness of breath.

Correct Response and Analysis

The correct response is B. Any treatment that does not enhance a terminally ill patient's comfort is not required, even if it can potentially lengthen life by a few hours or days.[55] JB clearly indicated that the oxygen was uncomfortable and its burdens were outweighing its benefits.

Dyspnea and pain are both powerful respiratory-drive stimulants. When morphine is titrated to relieve symptoms, it rarely causes significant respiratory depression.[51,86] Because apnea is a uniform occurrence at the time of death, any attempt to attribute JB's death to pharmacologically induced apnea is unsupportable.

Some healthcare personnel are so fearful of being accused of inducing death that they withhold interventions that could alleviate a patient's suffering to avoid even the appearance of causing a patient's death. Some nurses withhold medication because they are reluctant to give the "last dose." Unfortunately, alleviating suffering is not emphasized as a major goal of medical intervention during healthcare training. Medical and nursing trainees should be carefully instructed about the importance of preventing and alleviating suffering—an honored and critical goal of medicine. It may also be more practical to order the opioid and anxiolytic medications to be administered by continuous infusions. Good communication is essential.

Bereavement: Family, Friends, and Medical Caregivers

When JB dies, his parents become very distressed. The crying grandparents calm them, and the hospice nurse and social worker remain in the room to provide comfort and support. Emily, who is now 15 years old, asks the social worker for permission to go to her friend's house.

Question Thirteen

In characterizing Emily's grief responses, which of the following is a reasonable analysis?
A. Emily is coping by engaging in age-appropriate activities.
B. Emily is callous and should be reprimanded.
C. Since JB's parents have been prepared for his death, their behavior is yet another manifestation of their poor coping skills and personality flaws.

Correct Response and Analysis

The correct response is A. See the sections Grief Support Through Chronic Illness (page 25), Assisting Siblings (page 25), and Bereavement (page 26) for discussion.

The Case Continues

The hospice social worker suggests that Emily may benefit from attending a grief support group for teenagers. She also suggests that JB's parents notify the school about his death. This will provide an opportunity for JB's friends and acquaintances to attend his funeral or plan an independent memorial service. Notification also will give JB's friends and acquaintances an opportunity to process the loss with their own families, their classmates, and the school counselor. JB's parents agree to this. The hospice social worker provides the school counselor with ideas and materials to help teachers, classes, and individual students process the loss. The school counselor sends a letter to the parents of all children attending the school suggesting ways to help their children with grieving and explaining that their children may want to talk more about death. She offers her services if children or families need it. She mentions that some children may benefit from attending a bereavement group specifically for children.

Question Fourteen

The school counselor's response is
A. very appropriate and likely to help the students and families affected by JB's death
B. overkill—most of the children will not be affected by JB's death; only those known to be affected should be contacted
C. overstepping her bounds; most parents know how to handle death and the responses to it.

Correct Response and Analysis

The correct response is A. Most families and teachers do not know how to help children process the death of a friend. In addition, many adults are unfamiliar with developmentally appropriate responses to death and may be shocked by a child's perfectly normal response. In most cases, parents appreciate assistance with helping their child cope.

It is not obvious which children will be affected by

JB's death. Some may have had a private relationship with JB or may have known him in nonschool contexts. Others may be close to Emily and identify with her distress. Still others may become frightened because JB's death means they could also die. Some children may fear JB's disease was communicable and they have been "marked" or exposed by contact with him. For these reasons, a broad-based approach is justified.

The Clinical Situation Continues

The nurses who cared for JB throughout his life request leave to go to his funeral. Granting their request will decimate the shift schedule.

Question Fifteen

The nursing supervisor should (Choose all that apply)
A. tell the nurses they may choose three nurses to represent them at the funeral
B. let them know she cannot accommodate the entire group and suggest that a memorial ceremony be arranged, possibly in the hospital
C. make every attempt to provide coverage for the day
D. tell them they are being immature and that good nurses get over these things; other patients need their expertise and care.

Correct Response and Analysis

The correct responses are B and C. Medical caregivers also grieve a patient's death and need time to process their losses. Studies indicate that healthcare professionals who care for dying children need approximately twice as long to recover from their losses as those who care for adults who die.[180] Providing them with opportunities to grieve their losses will help prevent burnout and turnover and prolong their ability to care for dying children. It is in the nursing supervisor's best interests to accommodate their requests. However, accommodation is not always possible. Alternatives, such as attending a memorial service in the hospital, may be a reasonable and acceptable substitute.

The Case Continues

JB's mother refuses to dismantle his room. At JB's next birthday, she bakes a cake for him and cries. She also cries at every holiday. His father begins to spend even more time away from home. The mother calls the hospice social worker because she cannot believe how unfeeling her husband is. JB's father complains that his wife will "not stop talking about JB" and says he is tired of her crying.

Question Sixteen

Which of the following should the social worker advise? (Choose all that apply.)
A. The husband needs counseling to determine why he cannot allow himself to grieve.
B. The wife is experiencing pathologic grief and needs counseling to determine why she cannot recover from her loss.
C. The wife must immediately dismantle JB's room so she can resolve her grief.
D. To preserve their marriage, JB's parents need to support each other's responses to grief, and counseling may be needed.

Correct Response and Analysis

The correct response is D. JB's parents are experiencing normal responses to the loss of a child. It is not unusual for parents to grieve in very different ways, with fathers being the stereotypical silent grievers and mothers stereotypically crying and talking a lot more about the loss. Encouraging tolerance of differences and suggesting methods of helping each another can be affirming and beneficial. Because the death of a child can be so devastating for parents, counseling may be helpful even in the absence of continuing difficulties. The following suggestions may promote mutual understanding:

- The quiet partner can help the expressive parter go to support groups or to the homes of friends, tolerate tearful phone calls to friends, and avoid ridiculing the expressive partner's emotional response.
- The expressive partner can appreciate that there are many ways of grieving, respect the quiet one's need for silence and solitude, and avoid questioning the depth or validity of the silent partner's grief.

The Case Continues

One year later, JB's class prepares to graduate from high school. They assemble a memory book for JB's family, which includes photographs of themselves with JB and written memories of things he said and did as their peer and friend.

Emily is socializing with friends and is involved with music activities. She sometimes talks with a friend and with her mother about her brother and his death. JB's father has started seeing a counselor referred to him by his employer.

JB's mother reports that until recently she has had good and bad days and sometimes even better days. She knows that she and the rest of the family will be forever changed by JB's life and death, but she is hopeful that with time she will not find his death as painful. However, she has experienced renewed grief during the past month and has been despondent for two weeks.

Question Seventeen

JB's mother is showing signs of
A. pathologic grief
B. a normal response to the loss of a child
C. a major depressive disorder.

Correct Response and Analysis

The correct response is B. Bereaved parents often experience renewed and intensified grief on occasions that would have been significant benchmarks in the life of their child. Events such as graduations, weddings, and the birth of a grandchild can rekindle intense grief. It is helpful to alert bereaved parents and siblings that exacerbations of grief may occur but do not reflect mental illness or instability. Over time, the pain will lessen, but it will never disappear entirely. Decreasing pain does not constitute diminishment of their love or abandonment of the deceased. They are not forsaking a loved one when they no longer dwell on their memories daily. Families need permission to adapt to their new reality and to move on with their lives.

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Pretest Correct Answers

1.	A		21.	D
2.	D		22.	A
3.	B		23.	A
4.	D		24.	B
5.	C		25.	B
6.	A		26.	C
7.	A		27.	A
8.	D		28.	D
9.	B		29.	C
10.	B		30.	A
11.	C		31.	C
12.	C		32.	D
13.	B		33.	C
14.	A		34.	C
15.	C		35.	D
16.	A		36.	B
17.	C		37.	B
18.	C		38.	A
19.	B		39.	C
20.	B			

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