Treating a Child with a Life-Threatening Condition

Pediatric palliative care has been frequently misconstrued as pediatric end-of-life or hospice care, where the goal of comfort is not instituted until curative therapy has failed.\(^1\) Traditional goals of care focus solely on cure, addressing symptoms, such as pain, dyspnea, fatigue, poor appetite, and other non-pain-related distressing symptoms, only when death is imminent.\(^2\)

Pediatricians have been challenged to lead a shift in the paradigm: Palliation can be combined with curative treatment from the time of diagnosis of a child’s life-threatening or life-shortening condition. We review the educational, systems-based, legal, and ethical bases for the recent evolution for pediatric palliative care services.\(^3\) We also examine the current evidence that the implementation of pediatric palliation concomitant with curative treatments may be viewed as an efficacious treatment for a diversity of life-threatening conditions.\(^3,5\)

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EDUCATIONAL OBJECTIVES
1. Describe the recent revolution in pediatric palliative care services.
2. Determine the educational, legal, and ethical basis for recent changes in pediatric palliative care services.
3. Review current evidence demonstrating the efficacy of delivering pediatric palliative care in concert with ongoing curative treatments.

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CASE EXAMPLES

Case 1

J was a 4-year-old boy with refractory pre-B cell leukemia, which was treated by the pediatric oncology team since diagnosis. Despite aggressive treatment, sustained remission was never achieved. Although the oncology team recommended stopping chemotherapy, his family insisted on continued attempts at cure and requested transfer to a facility in another country, which offered non-traditional chemotherapeutic agents.

Before transfer could be arranged, he had a sudden deterioration, requiring transfer to the pediatric intensive care unit (PICU) and died shortly thereafter. Independently, his grandmother contacted a social worker from a local hospice; a member of the hospice team arrived soon after the child's death. The hospice team provided bereavement support to the family in the months after the death.

Case 2

A was a 15-month girl with mitochondrial encephalopathy, lactic acidosis, and stroke-like episodes (MELAS) syndrome. She had progressive neurologic deterioration, leading to recurrent episodes of apnea, with hypercarbia. After repeated admissions to the PICU, twice requiring mechanical ventilation, she was maintained on Bi-PAP, oxygen, and enteral feeding by gastrostomy tube. The wishes of the parents were to have their daughter home, with no further PICU admissions, and a do-not-in-tubate, do-not-resuscitate order.

A referral to hospice was made, with the challenge that they had never cared for a child at home in a rural area. The hospice medical director was not comfortable with addressing the care and needs of this young child, nor had any of the hospice nurses any pediatric experience, and the authorization for a pediatric Bi-PAP machine was denied.

The child’s pediatrician agreed to assume the hospice care, arranging for telephone support from the child’s subspecialists (eg, infectious diseases, intensive care, neurologist, pulmonologist, and palliative care). The palliative care team, along with the regional home health agency, worked with the local hospice and state agencies to secure home equipment, which included the Bi-PAP machine, suction, oxygen concentrator, and feeding supplies. The toddler was transferred home, under the parents’ care, and survived for several months without another hospital stay.

EPIDEMIOLOGY

In 2007, 29,241 infant deaths occurred. Of these infants, 12,692 died within the first year of life, with 4,572 deaths occurring in the first month of life. In 2007, 23,976 children aged 1 to 19 years died, with 17,351 of these deaths occurring from chronic, controlled conditions. The population of children with complex chronic conditions die in the hospital (81%), while 17% die at home. A child’s race, ethnicity, and region of home residence are significantly associated with death occurring at home (ie, white and Western region of the US); children with malignancy, neuromuscular, metabolic, or congenital/genetic complex chronic conditions were associated with the greatest likelihood of death having occurred at home.

The population of children with complex chronic conditions, whether they are residing in the community or in the hospital, is the current focus of many pediatric palliative care teams.

BARRIERS TO IMPLEMENTATION OF PEDIATRIC PALLIATIVE CARE

Many explanations are possible to relate why pediatric end-of-life care has been so focused on curative therapy at all cost. One major cause has...
been the lack of acceptance of palliative care by the lay public as well as the medical establishment.9 Physicians have been trained with a curative focus; some perceive a patient’s death as a failure of medical care. Symptoms, in the medical model, are clues to a diagnosis rather than targets for treatment. Academic medical centers and parents have needed to balance cutting-edge clinical research in incurable conditions with the choice to enroll children in clinical trials in the last weeks or days of their lives.10

Training resources in pediatric palliative care during residency began with the introduction of Initiative for Pediatric Palliative Care (IPPC), launched in 1998 as a research, quality improvement, and education effort aimed at enhancing family-centered care for children living with life-threatening conditions. IPPC’s comprehensive, interdisciplinary curriculum (available at www.ippcweb.org) addresses knowledge, attitudes and skills that health care professionals need to serve children and families better.

IPPC was initiated as a project of the Center for Applied Ethics (CAE), a division of Education Development Center Inc. (EDC), working in collaboration with the National Association of Children’s Hospitals and Related Institutions (NACHRI); the Society of Pediatric Nurses (SPN); the Association of Medical Schools Pediatric Department Chairs (AMSPDC); and the N.Y. Academy of Medicine (NYAM). IPPC’s broad-based collaborative initiative has included children’s hospitals, pediatric units in general hospitals, and hospice or home care programs that serve children living with life-threatening conditions and their families.11-13

The Medicare hospice benefit, which provides end-of-life care for adults (when curative therapy is abandoned and life expectancy is less than 6 months), does not fit most needs of children with life-limiting conditions. Aggressive treatments, such as chemotherapy, antimicrobial agents, and even surgical procedures, may be a part of the goals for the child, family, and pediatric medical team, to achieve palliation of symptoms, quality of life, and maintaining hope of response in the face of disease. There are limitations of most hospice services for home care, leaving few options for families wanting to take children home (see Sidebar 1, page 260).14-17

**MIXED MESSAGES**

Mixed messages from health care providers also create confusion for families. Prognostication has been difficult for children due to limited reliable information that was often old and based upon few cases (see Sidebar 2). Even when prognosticating data may be clear, physicians may disagree among themselves about prognosis and what should be told to the parents.18 There may be a significant delay between when the physicians recognize the failure of curative therapy and when the parents ultimately express their understanding.19

Further, parents and even clinicians may avoid recognizing the dying process because the child has repeatedly survived nearly fatal episodes.20 Perception of hope for transplant, fear of taking away hope, and parents lack of understanding, unwilling to accept mixed messages from health care providers may explain why pediatric palliative care. Care teams may struggle to communicate a shift or change, realizing this may change the hope expectation for a family (changing “what they hope” for “to taking away hope”).20

In Case 1 at the beginning of this article, the family was introduced to hospice late in the process of treatment of this child.3 Bereavement services were offered to his family, although there was minimal preparation for or anticipation of death. The ideal pediatric palliative care model would be that the oncology and palliative care teams

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**SIDEBAR 2.**

**Challenges in Prognostication in Pediatric Chronic Care**

**Mixed Messages to Families about Care**
- Aggressive measures taken despite limited response
- Lack of communication among subspecialists

**Confusion about Prognosis**
- Limited reliable information may be outdated and based upon few cases
- Disagreement among health care providers
- Parental, family denial about news

**Delay in Recognition of Failure of Curative Therapy**
- Short-term problems addressed, no big perspective taken
- Health care team(s) unwilling to accept failure (eg, impending death)
- Parents lack of understanding, unwilling to accept

**Persephone Syndrome**
- Family and health care team avoid recognizing the dying process because child has repeatedly survived nearly fatal episodes

**Delay in Initiation of Conversations about Palliative Care**
- Child, families fear candid talk signals loss of hope
- Concern talk communicates defeat or giving up
- Fear of taking away hope21
- Discomfort with unfamiliar/unknown prognosis

**Confusing Future while Waiting for Organ Transplantation**
- Aggressive treatment of symptoms while on list
- May be unclear direction of care if patient’s condition worsens, and there is no organ available
- Balance of hope for transplant, fear of death while waiting
- May receive care in more than one tertiary care center (referring center and transplant center)

*Source: Klein SM.*
being present at the time of diagnosis. Discussions would include conversations on diagnosis, prognosis, and treatment options with the oncology team, while reviewing expected symptoms during the treatment and strategies for addressing each symptom with the palliative care team.

Many existing pediatric subspecialists already work as a team (eg, oncology, cystic fibrosis, neonatology) and address symptom management. The advantage of involvement of a pediatric palliative care team from the beginning is the familiarity with the child and family and the lack of an abrupt signal that a shift in care is happening, when consultation of the palliative care team begins.

In reality, pediatric tertiary care centers do not usually include the palliative care team at the time of diagnosis; many palliative care teams are introduced mid-way or later through a process, when the clinical course is shifting toward goal-of-care and symptom-management, with progression of the disease process to death.

**A DIFFERENT MODEL**

It may be agreed that introduction of the palliative care team earlier in the process for the child in Case 1 may not have changed the outcome or the family’s wishes for transfer.

Depending on the needs of the family, however, an outpatient pediatric palliative care team may have employed a child life therapist or expressive art therapist to work with the patient and siblings; enlisted a social worker to make sure the parents are juggling their many new responsibilities; hired a nurse to draw blood, inquire about pain, nausea and fatigue; and consulted a chaplain to assess and support the family’s spiritual or religious well-being.

As the disease process progresses, the family would continue to build relationships with the oncology and palliative care teams, would discuss results of treatment, review symptoms, and provide support. When the curative treatment begins to fail, the oncology and palliative care teams would jointly shift goals of care.

Case 2 is an example of a shift in intensive care treatment of a progressive neurologic condition to home-based care. This toddler received aggressive care in the PICU, which was in line with the initial wishes of the mother, while her father expressed wishes to withdraw care. After consultation, the palliative care team worked over a series of weeks establishing a relationship with the parents, clarifying the prognosis, and defining and aligning each parents’ goals for their child. The mother’s goal was to take her daughter home, to which the father agreed. This was accomplished with the coordination of the pediatrician, hospice, and the state agencies.

Care of children with many other chronic conditions can follow the model of shifting aggressive care to a symptoms-based approach when dealing with issues such as congenital heart disease, neurologic conditions, genetic disorders, cystic fibrosis, oncology, and nephrology. The advantage of a palliative care approach is the comprehensive team management of disease symptoms, as well as to the psychological, social, and spiritual needs.

Ideally, a sudden switch to palliation would occur, in the event that a cure is not possible. The palliative care team can act as a helpful, impartial support, taking a more prominent role in care, as the care shifts to comfort. Whether the patient is cured of illness or dies, the palliative care team will have worked alongside the primary team to complete the most basic principle of palliation: to relieve physical, social, emotional, and spiritual suffering.

The pediatric palliative care team may participate in complex decision-making to provide a framework for the medical indications, patient preferences, quality of life, and contextual issues that surround forming goals of care. The primary medical and palliative care team, with the family and child, may develop the goals of care based upon what is most important to achieve. Although some of the goals may be an eventual cure, there will also be goals based upon symptom control and psychosocial needs. Goals of care should be customized and placed in the context of the current clinical situation.

**CHANGING GOALS**

As health care needs change, so may the goals of each patient’s care. One of the important questions that can be answered through this process is, “What can we do?” This question places a strong emphasis on solutions rather than focusing on “What can’t we do?” Often parents and providers become stuck in the “there is nothing we can do” frame of mind and greatly benefit from learning some of the simple things that can be done.

Part of the goals of care discussion should include advanced care planning. Advanced directives are more than do-not-resuscitate and do-not-intubate orders. The directives can include school attendance, comfort measures, decisions about returning to the inpatient or intensive care settings, and even questions about feeding and hydration. These conversations are difficult and often painful. However, in some families, they can provide tremendous comfort to families as the child declines. These discussions are much more difficult when postponed to the last days of life.

Another strategy that can help families accept palliative care is that symptom control should be emphasized even when curative treatment is the goal. Many children suffer from symptoms that can be eased with palliative techniques and do not need to be reserved for the dying. Again, the focus should always be on what “can” be done, and away from what “cannot.” Family support groups can be helpful in providing examples of successes.

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with palliation. Parents who share their struggles with curative therapy can also relate how palliative therapies improved quality of life regardless of the outcome of attempts at cure.

In addition, families are prone to feeling alone; these groups can provide comfort and solidarity. Siblings are especially susceptible to loneliness and feelings of abandonment. Participation of the siblings within child life activities, attendance at camps, and finding support groups or family activities is encouraged and can have long-lasting benefits for these surviving children. In addition, some of the burden can be lifted from the parent who may feel overwhelmed by the competing needs of their dying child, spouse, other children, extended family, and friends.

Two other aspects of care often neglected are alternative non-pharmacologic therapies and spiritual care. Examples of non-pharmacologic care to provide symptom relief include mindfulness, aromatherapy, acupuncture, and massage. Depending upon the values and beliefs of the family, spiritual care can have a role in the care of the dying child. Many families may work closely with their own spiritual supports; however, there is a growing trend of hospital chaplains receiving specialized training in caring for dying children, and they may be a resource not only for the family and the hospital staff, but also for the family’s own clergy.

Location of care may be negotiated. For some families, the hospital is a sanctuary and the preferred source of care. However, other families prefer home care, even if some types of care are not possible. Depending on his or her age, the child often has specific opinions about where he/she would like to die and should be included in the discussion. Each decision made should be the one that best suits the individual needs of the child, the family, and the community resources.

**ETHICAL CONSTRUCTS**

The general approach to decision-making for a child under palliative care assumes that the parents will act as surrogate decision-makers for their child. Thus, parents proceed in a manner that would be in line with their child’s own decision, if the child had capacity. These decisions are understood to be congruent with the values and beliefs most important to the family and thus would be shared by the child.

The best interest standard pertains to decisions that have weighed risks and benefits as they specifically apply to the child. Best interest can be fairly subjective, and the views of the various stakeholders involved in the care of the child can be very different. This can lead to tremendous conflict among the family, the child, and the health care team.

Futility may be prone to subjective bias among various stakeholders. Quantitative futility looks at chances of success of treatment and often uses 1% success as a standard. However, quantitative futility is often difficult to prove. Qualitative futility is focused upon quality of life and is dependent upon individual values and beliefs.

There are several court decisions that help frame the rights and responsibilities of parents, adolescents and providers. The regulation often misunderstood is that of Baby Doe, an amendment in 1984 of the Child Abuse Prevention and Treatment Act (CAPTA) (see page 254, Sidebar 1, in accompanying article). The regulation allows physicians to withhold treatment for infants younger than 1 year if certain conditions (including being irreversibly comatose) are met, and treatment would only prolong the dying process or be futile. The intent was to make sure that infants who were disabled would not be denied appropriate treatment solely based upon disability. However, the regulations do not mandate that infants with disabilities must receive treatment even if the treatment would be nonindicated for a non-disabled infant.

Questions of adolescent authority in their own self-determination become more complex. By definition, to make medical decisions, one must be competent, and traditionally, this has included being at least 18 years of age. However, in many situations, minors have demonstrated great insight into their own care and have been afforded some decision-making rights. Many states recognize “emancipated minors” as being able to make competent decisions.

Some authors have suggested the idea of “functional competence,” which could be seen in minors as young as 14 years. By definition, functional competence is shown through the ability to reason, understand, choose voluntarily, and appreciate the nature of the decision. The courts have been slower to adopt this developmental and ethical construct of adolescent competence and have generally approached medical decision-making in this age group on a case-by-case basis.

Lastly, the child’s own opinion may be overlooked within the conflict between families and providers. Although the court has not had consensus on an approach to adolescent decision-making, pediatricians have been more supportive of involving the wishes of the child in the overall goals of care. Ethics committees can be very helpful in dissecting the issues and even mediating discussions with families.

**RELIEF OF SUFFERING**

There may be several parties who suffer when a child is dying. First of all, children are capable of suffering from pain, anxiety, and loneliness. A terminally ill 3-year-old child may acquire awareness and concepts of death far in advance of healthy peers. A child may experience suffering due to concerns about the family and the fear of being a burden; this is no different from what dying adults may experience.

Parents, on the other hand, suffer in different ways. First, parents grieve for their living child. They may fear that their child is suffering, but they also fear about living in a world without their child. Second, marital discord is a frequent occurrence;
acute and chronic illness in a child can create tremendous stress on all relationships.

Different emotions and experiences from a similar situation are common, sometimes making it challenging for parents to sympathize with each other. “Why is (s)he not over it yet? I am!” may be a spousal complaint. Strong emotional factors about the child’s illness may cloud parental decisions, as well as their perception of their own or their child’s suffering. Parents may feel guilty from decisions about withdrawing care; on the other hand, deciding to prolong their child’s life, at the cost of quality of life, may create guilty feelings, as well.40

Health care providers may also suffer during the care of dying children. The longer providers have been involved with a child, the more attached they may become to the child and family. In addition, close bonds can develop that may cross professional boundaries. Lastly, moral distress can occur when practitioners’ beliefs clash with the decisions made by families. Practitioner burnout is a significant risk for those working with dying children (see accompanying article, page 266).41

BREAKING DOWN BARRIERS

Progress in the field of palliative care for pediatricians makes this model more of a reality when caring for children with chronic disease. Children’s Hospice International (CHI), with technical assistance from the Centers for Medicare and Medicaid Services (CMS), Members of the US House of Representatives, and health care leaders, developed the Children’s Hospice International Program for All-Inclusive Care for Children and their Families (CHI PACC) to overcome existing barriers to appropriate care for children with life-threatening conditions and their families. Several states have adopted the CHI PACC program, with others to follow. The tenets of CHI PACC include four goals:

- To offer a comprehensive continuum of care for children with life-threatening conditions and their families from the time of diagnosis with hope for cure through bereavement follow-up if cure is not attained.
- Families of children with life-threatening conditions are not forced to choose between curative care and hospice/palliative care, but instead choose the CHI PACC model that allows palliative care to be offered along with disease treatment.
- The CHI PACC model provides early and continual intervention and case management functions to prepare families and health care providers to provide support for the seriously ill child.
- With respite care, provided by the CHI PACC model, parents are able to continue functioning in the community.42,43

This program is intended to provide funding for children to enroll in services by removing some traditional barriers; there would be no requirement for a life expectancy of less than 6 months, and curative treatments do not have to be abandoned to be eligible. These two factors are intended to allow parents to enroll children in hospice services earlier in the course of illness without fearing that they are giving up hope for a cure.44

Another factor helpful in the acceptance of palliative care is the academic recognition that has grown in the past several years. The certificate of “Added Qualifications in Hospice and Palliative Medicine” is now a sub-board of pediatrics; internal medicine; anesthesiology; emergency medicine; family medicine; obstetrics and gynecology; physical medicine and rehabilitation; psychiatry and neurology; and surgery. The number of available fellowships in the United States in palliative care has grown to 73 active programs, five of which are in pediatrics.44 Several peer-reviewed medical journals have dedicated sections to palliative care. Centers of Excellence have been identified that act as role models for other institutions looking to develop palliative care programs.45

Perhaps even more importantly, parents and children have developed a strong voice advocating for palliative care. The Internet has become a valuable resource for parents reaching out to other parents, with support groups and informational websites prevalent.25,46 Parents have authored articles relating their experiences, and frequently families share their stories at national meetings. The lay press even seems more interested in the human aspect of pediatric palliative care.47,48 Parents also have clarified their wishes regarding prognosis disclosure as well as the need for reasonable hope as a way to ease suffering.49,50

ROLE OF THE PEDIATRICIAN

Pediatricians experience pediatric death in their practices and must become familiar with this not-so-uncommon process. The pediatrician can be a tremendous source of comfort to the child and family, possessing a relationship and the ability to guide the family through the difficult process and maze of decision-making.11,51 The pediatrician can become the coordinator of care, facilitating communication among the various services. Communication has been reported by parents as the key determinant in high-quality physician care.52 Pediatricians may have the biggest effect by just “being with (the) dying.”53 Finally, for many children with complex, chronic disease, the day-to-day issues of comprehensive primary care are best served by their primary care providers.

CONCLUSION

More than just end-of-life symptom management, a pediatric palliative care interdisciplinary team attends to pain and symptom management throughout illness, establishes and revisits goals and location of care, and is attuned to the physical, social, emotional and spiritual sufferings of the patient, family, and professional caregivers.
Pediatric palliative care specialists have been challenged to lead a paradigm shift: palliation can be combined with curative treatment from the time of diagnosis of a child’s life-threatening or life-shortening condition.

REFERENCES