

Because Statistics Don't Tell the Whole Story: A Call for Comprehensive Care for Children With Cancer

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Introduction

Families facing a childhood cancer diagnosis are catapulted into crisis mode. Everyday family life is whisked away and replaced by frightening diagnoses and medical language, life-or-death decisions, intrusive emergency hospitalizations of unknown duration, a steady stream of hospital visits, medications and procedures that must be handled with perfection, expert opinions that must be acted upon in haste, negotiations with employers for extreme absenteeism, special arrangements with school principals and teachers during long periods of quarantine from other children and friends, and financial burdens that are felt years beyond that dreadful day of diagnosis.

This is not a “new normal.” There is nothing “normal” about children being struck by serious illness. I know this well, from personal experience: my daughter, Alexandra, was diagnosed with medulloblastoma in 2011, just before we were to celebrate her fourth birthday. A childhood cancer diagnosis launches an unwanted journey with unrelenting potential for a bad ending. But there is a role for genuine hope in all aspects of the evolving experience. It begins with hope for cure or remission. If survival is possible, it can develop into hope for and a return to childhood, including reintegration into school. Yet many families come to learn that hope's constellations may change over time, particularly if disease progresses or recurs. In the face of such tragedy, with the horror of learning that death is certain, the hope may become one for perfect comfort to the end. No matter the course, the goal of providing care and comfort that supports a full life for all children with cancer must be supported and attainable.

With cancer, grief begins at the time of diagnosis, when the family learns that their child is sick and will experience necessary harm in order to survive. In the event that a child emerges cured of cancer, he or she will likely face permanent and lifelong damage from treatment, ranging from mild to

severe. Life will never be the same again, and the family's future shifts sharply from what was expected. When this grief and shift in expectations is addressed, it can be managed and the family can begin coping. As a cancer epidemiologist, I was better equipped than most parents to face down this crisis. But that realization frequently left me wondering: how did families with little medical knowledge or inflexible work schedules manage their fears and navigate the numerous daily unknowns? What is on the line is the life of your child. Worse yet, as we would be counseled in great detail, the cost of trying to save our young daughter would be certain and permanent cognitive devastation. Having to choose between treatment strategies that are terrible and terrible really presents no choice at all.

Cancer in children presents a different illness experience from cancer in adults and triggers fundamentally different needs in families. Unfortunately, however, because of a lack of dedicated pediatric cancer research support, clinicians are regularly forced to recycle adult treatment approaches in pediatric cancer and, with good intentions to protect the families, they often do so without adequately acknowledging the significant and potentially fatal problems with this course of action. It is firmly established that children are diagnosed with cancer types that differ from those in adults, and even cancers that are the same behave differently in a child. Unlike adults, children's bodies are still growing and developing during illness and treatment; therefore, cancer treatments will affect them unequally over their life compared with adults. The long-term and lasting implications of cancer treatment on a child's physical, emotional, and cognitive functioning are increasingly evident.^{1,2} Specialized medical care, and the research that guides it, must be tailored to address these unique circumstances, challenges, and family needs.

We have seen successes for some cancers, but others remain incurable and untreatable. And no matter the prognosis or survival statistic, the literature is clear that

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See referenced article on pages 83-103, this issue.

Dr. Cullen would like to acknowledge the contributions and encouragement in helping prepare this article from Rebecca Kirch (American Cancer Society [cancer.org]), Blyth Lord (Courageous Parents Network [courageousparentsnetwork.org]), and Joanne Wolfe, MD, MPH (Dana-Farber Cancer Institute/Boston Children's Hospital).

DISCLOSURES: Dr. Cullen is a cancer epidemiologist, mother, and American Childhood Cancer Organization (acco.org) board member.

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preventable suffering is a part of the experience that interferes significantly with the quality of life of these children and their families.³ The accompanying article by Ward et al reporting statistics for childhood cancer incidence, mortality, and survival is an essential inventory to help evaluate our progress and shortfalls.⁴ Importantly, for the first time in over a decade,⁵ instead of presenting these figures in the aggregate, these more comprehensive data tease out several specific cancer types to capture a clearer picture of the actual childhood cancer landscape today. These figures demonstrate tremendous variation in survival and success rates across the different cancers affecting children.

Moreover, while overall the cancer incidence numbers reported here may be considered comparatively small when measured against the much higher incidence among adults, cancer is still *the* leading cause of disease-related death (second only to unintentional injury in *all* causes of death) in children aged 5 years to 14 years.⁶ For families facing these unfortunate odds, characterizing childhood cancer as “rare” or survivable in “80%” of children is dismissive of the gravity of the impact on the young lives afflicted with cancer. Moreover, it fails to account for cancers that are terminal at the time of diagnosis or those that have widely varying behavior in terms of treatment response, and it fails to acknowledge that the state of the science for some childhood cancers has stagnated for decades. Finally, for some treatable cancers, the steep cost of survival is one of gross mental impairment, poor motor functioning, and highly compromised social well-being for decades of life after treatment. Associating aggregated information with the very unique cancers and circumstances of childhood cancer misrepresents the unyielding burden these children and their families must bear over their lifetimes.

As a cancer professional, I understand why cancer clinicians present the information as they do: they want to keep hope for survival and a positive future at the forefront. But as a parent, I know that parents want information and want to feel fully informed. They do not want false hope. And research with parents has borne this out.^{3,7,8} There are many childhood cancers that carry a grim prognosis from the outset. For cancers that do exhibit survival probabilities in the “80%” range, the percentage fails to acknowledge that the overwhelming majority of these children will have significant, chronic, and life-altering side effects as a result of their cancer treatment.

Supporting Families Facing Unthinkable Decisions

Clinicians often recognize and are inspired by the strength, resilience, and wisdom of children and parents facing life-threatening diseases such as cancer. Even very young children have remarkable insights and awareness about their condition. Although parents may not be experts in disease and medical decision-making, and may initially feel

overwhelmed with what they do not know, they are the exquisite experts on their children and also the keepers of their family’s needs and values. As such, parents deserve to be asked about their individual touchstones for ensuring their child’s well-being and about their care goals and values for their child and the whole family. Ensuring that parents have a voice and shared role in the critical decisions is especially vital for helping families live with the fewest regrets.

This focus on family engagement and support will require that we address the flaw inherent in today’s health system that emphasizes treating disease without requiring concurrent palliative care to also support the patient and family. Fortunately, the American Cancer Society’s advocacy affiliate, the American Cancer Society Cancer Action Network, has helped to lead efforts with multiple national partners to introduce and build support for quality-of-life legislative proposals in the US Congress and several statehouses as an important initial step in acknowledging that treating the pain, symptoms, and stress of serious illness, at any age and any stage, is as important as treating the disease.⁹ In addition, we must recognize that communication is central to the work clinicians accomplish. Good communication improves patient and family adjustment to illness, lessens pain and physical symptoms, increases adherence to treatment, and results in higher satisfaction with care.¹⁰ It also helps clinicians to enjoy and thrive in their work. To deliver truly person-centered and family-focused care, we must promote an oncology culture and associated academic training that emphasize clinical communication skills that equally balance honesty, empathy, and hope.¹¹ As recommended in the Institute of Medicine’s recent report,¹² providing communication skills training through cancer programs is essential support for clinicians who aspire to provide optimal care for their patients, particularly among clinicians who already recognize and trust that the awareness of children and their families may be even more realistic and transparent than their own. Certainly many pediatric cancer clinicians are already strong communicators who practice palliative medicine and know well how to frame their conversations, especially the difficult ones, with parents. However, given the sensitive nature and life-changing content of their work, *all* cancer clinicians need training to be strong communicators who understand the benefits of palliative care.

Palliative Care as a Lifeline to Quality of Life

Childhood cancer requires a parent to helplessly stand by while bearing witness to a succession of physical and emotional insults. In the case of Alexandra’s cancer and its treatment, she bore craniotomy scars, baldness, pallor, hydrocephaly-induced seizures, a cerebral shunt, severe

TABLE 1. Benefits of Pediatric Palliative Care

- Relieves the child's disease symptoms, such as pain, shortness of breath, fatigue, constipation, nausea, loss of appetite, and difficulty sleeping.
- Helps with communication and coordination of care between the medical team and the parents.
- Helps the child and family have the physical, emotional, and mental strength to carry on with daily life and provide the best quality of life for the affected child and the family.

Adapted from Get Palliative Care. What Is Pediatric Palliative Care? getpalliativecare.org/whatis/pediatric/. Accessed September 24, 2013.¹⁵

mucositis, nutrition through a central line in her chest, chemotherapy-induced skin burns, disfigurement from steroids used for weight maintenance, and severe neuropathy impairing her ability to walk. Over 13 months, cancer and its treatment ravaged Alexandra's beautiful and tiny body. When finally faced with the last chemotherapy cycle, my instinct was strongly against it. I believed her body had been pushed too far. Yet I did not trust this personal intuition enough to speak up, and I now reflect on that period and wonder if the integration of palliative medicine would have emboldened me to voice my concerns. And although such a change may not have affected the eventual outcome, it may certainly have affected our daughter's quality of life. Only in those final weeks of her life, with hospice helping us prepare for her death in our home, was highly specialized pediatric palliative care embedded into Alexandra's daily cancer treatment, with the specific purpose of helping to support and guide us in our decision-making to ensure quality of life as a critical goal in the management of her disease. Alexandra's autopsy would ultimately show that proton radiation-induced necrosis, rather than cancer, caused her death, a striking revelation that underscores how much we have still to learn about the behavior of childhood cancers and their response to treatments.

There is no reliable road map for maneuvering this journey, but there is ready help to make sure parents, siblings, and children facing serious illnesses such as cancer feel they are seen, heard, and understood, and thus perceive themselves as truly respected and having a voice on the journey. Pediatric palliative care is that critical piece of assistance that focuses on the quality of life for the child and family. It is provided by a team of doctors, nurses, and other specialists who work together with a child's oncologists and other clinicians to provide relief from the symptoms, pain, and stress of cancer. Pediatric palliative care is appropriate at any stage, and can be provided along with treatments meant to cure. This last point is critical, as palliative care has historically been misunderstood as being tantamount to "giving up hope." On the contrary, palliative care can be concurrent with treatments and is about providing families with hope: hope for comfort, hope for well-being, hope for as good an

TABLE 2. What Clinicians Can Do

- Understand that any cancer diagnosis is a loss; parents will be in mourning, grieving the loss of a "healthy child" and perfectly envisioned future.
- Recognize that "whole-family care" is needed, not simply medical care of the affected child. For example, be prepared to offer advice about how to support siblings.
- Identify sources of support for the family, including other affected parents.
- Talk about palliative care as an "extra layer of support helpful at every point in the child's care." Help families understand it is life-affirming, not "giving up" or only for end-of-life care.
- Ask families what they use as their touchstones for decision-making and support that approach, serving as a nonjudgmental and nondirective guide and facilitator, recognizing that your own emotions, values, and framework may differ.

Adapted from National Tay-Sachs & Allied Diseases Association. Parenting a Child with Life-Limiting Illness: Caring, Coping, Surviving. A Resource Guide for Parents and Professionals. ntsad-ny.org/film.html. Accessed September 24, 2013.¹⁷

outcome as the medical prognosis will allow (including comfort and peace at the time of death).

When caring for pediatric patients with cancer, doctors and parents are often faced with difficult decisions concerning the benefits versus burdens of medical technology and impacts on the child's quality of life. With the close communication that palliative care provides, families are better able to make choices that are in line with their values, traditions, and culture. This improves the well-being of the entire family and promotes emotional and spiritual healing even if the disease persists. Ideally, palliative care begins at the time of diagnosis, so that families have this extra layer of support all the way along their child's continuum of care, which in turn enables emotional and psychological healing beyond the disease.^{13,14} Table 1 shows some of the benefits of pediatric palliative care.¹⁵

Caring, Coping, Surviving: Community Call to Action

When a child is diagnosed with cancer, the family, the family's community, and the broader childhood cancer community engage with an outpouring of concern, support, and a desire to take action. Advocacy in the childhood cancer community has been a remarkable force to date, led by those touched deeply and personally by the specter of a suffering child. Indeed, in the wake of Alexandra's death, my husband and I thought to start a new 501c3 nonprofit organization in her honor. In the months that followed, however, as we surveyed the cancer advocacy landscape, we observed how replete it was already with organizations, each tasked with their own worthy efforts to cure or eradicate childhood cancer, their own piece of the childhood cancer pie. Collectively, these organizations are passionate in their purpose but, as both a parent and cancer advocate, I worry that they are inadvertently engaged in splintered

messaging and competition to be heard, costly activities when what we really need is a unified message that gets at the common goals shared by all: continued research for childhood cancer cures with a concurrent focus on quality of life for the child and the family. After all, for children and their families, treating the pain, symptoms, and distress that accompany a cancer diagnosis is as important as treating the cancer itself.

This focus on quality of life requires 2 things. First, it requires palliative care training for all pediatric specialists, not just those who seek it out. Just as all physicians caring for children receive training in therapeutic and preventive medicine, they should also be trained in the essential elements of palliative medicine, which are, at their core, the essential elements of all medicine: person- and family-centered care. Every pediatric clinician, in partnership with other health care providers and supporters, is capable of addressing the needs of children with cancer and their families, through personal knowledge and skill or by providing proper referral, treatment, and follow-up, including referral to specialized dedicated palliative care teams. Every childhood cancer journey should start with a plan for specialized medical care, including palliative care, tending to the important differences in how children and their families experience life-threatening illness. Unfortunately, however, pediatric palliative care teams are not yet available in all settings in which children and families receive their care. And even when these specialist teams are available, significant variation exists in the services offered.¹⁶ Second, we need more research about quality of life for pediatric patients with cancer along the entire cancer care continuum. Using the data presented by Ward et al⁴ as our benchmark for prioritizing action and evaluating our progress going forward is the next step toward this goal.

When I reflect on our daughter’s journey, including her enormous physical suffering, I ponder how an early emphasis on palliative care, alongside the excellent disease-directed medical care she received, might have alleviated some of our suffering and prevented regrets about decisions we made regarding her treatment, regrets that were all the more painful in the wake of the loss of our beautiful and courageous little girl. Anything that can be done to help the children and families forced to make decisions with their doctors but without a crystal ball must be done. Table 2 lists some things clinicians can do to aid families at this time.¹⁷ ■

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