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Palliative care and pediatric surgical oncology

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ABSTRACT

Survival rate for childhood cancer has increased in recent years, reaching as high as 70% in developed countries compared with 54% for all cancers diagnosed in the 1980s. In the remaining 30%, progression or metastatic disease leads to death and in this framework palliative care has an outstanding role though not well settled in all its facets. In this landscape, surgery has a supportive actor role integrated with other welfare aspects from which are not severable. The definition of surgical palliation has moved from the ancient definition of noncurative surgery to a group of practices performed not to cure but to alleviate an organ dysfunction offering the best quality of life possible in all the aspects of life (pain, dysfunctions, caregivers, psychosocial, etc.). To emphasize this aspect a more modern definition has been introduced: *palliative therapy* in whose context is comprised not only the care assistance but also the plans of care since the onset of illness, teaching the matter to surgeons in training and share paths. Literature is very poor regarding surgical aspects specifically dedicated and all researches (PubMed, Google Scholar, and Cochrane) with various meshing terms result in a more oncologic and psychosocial effort.

Introduction

According to WHO definition, palliative care is *“an approach that improves the quality of life of patients and their families facing the problem associated with life-threatening illness, through the prevention and relief of suffering by means of early identification and impeccable assessment and treatment of pain and other problems, physical, psychosocial, and spiritual.”*[1] As a matter of fact, despite improving rate of success in diagnosis and treatment of cancer,[2] many patients experience progression of disease or recurrence and finally die of tumor, and the management of this phase of the disease is the field of palliative care.[3] The term “palliative” is derived from Latin verb “palliāre” (meaning “to cover”) and from substantive “pallium,” which was a particular type of dress worn by ancient romans on the tunic, as a coat.

For pediatric patients, the first and most important concept is that palliative care is unique and specific and requires skills, organization, and resources, which are different from those for adults. In recent years, the developed world has seen an increase in the prevalence of incurable disease and disability.[4,5] Medical and technological advances have reduced infant and child mortality rates and, at the same time, have improved the survival rates of children with severe and potentially lethal pathologies, who need complex palliative care.[6] For many years, palliative care was not offered to pediatric patients and even today in Europe, despite the development of pediatric palliative care programs in several countries, only a minority of children with incurable illness benefit from palliative care services. Many of these children will die in inadequate conditions[7]; without relief from distressing symptoms, usually in a hospital setting and rarely with care support in their own home where many would prefer to spend as much time as possible and eventually die. The reasons for these shortcomings in the provision of pediatric palliative care can be attributed to organizational, cultural, and economic barriers. The unique aspect of childhood illness further complicates the situation and small patient numbers, together with the multiplicity and broad geographical distribution of cases, and determines the organization of palliative care services very different to those provided for adults. Children with life-limiting and life-threatening illnesses deserve a thorough cultural and organizational reappraisal of how we care for them when treatment is not aimed at recovery but at offering the best quality of life possible.[8] In this respect, children with life-limiting and life-threatening conditions and their families have diverse and multiple needs (clinical, psychological, social, and spiritual).

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The needs of children

Physical needs:

- *Symptoms control*: the timely pharmacological and integrative non-pharmacological management of pain and other symptoms by appropriately skilled professionals (consequentially resulting in a reduction in the number of unnecessary hospital admissions);
- *The attainment of their full potential of growth and development*: these aspects must be considered and included when considering any form of treatment; and
- Advanced care planning is essential.

Psychological needs:

- Open and clear communication appropriate to the developmental stage of the child is necessary;
- Continual emotional support to help the child cope with emotive issues: understanding, acceptance, anger, self-confidence, trust, and love; and
- Access to resources and tools that promote the development of the child's personality, self-awareness (and that of others), the enhancement of individual characteristics and talents, and, where possible, the continuation of daily routines, incentives, targets, and future projects.

Social needs:

- Recreational opportunities, tools, techniques, and activities appropriate for individual needs;
- Schooling for as long and often as possible, even at home with individual tuition; and
- Social activities offering appropriate opportunities for interaction with peer groups, possibly through voluntary groups and organizations.

Spiritual needs:

- Access to appropriate spiritual care and support respecting the family's cultural and religious background.

All these individual needs are in continuous evolution, both in prevalence and intensity, directly linked to the child's psycho-physical-emotional development, the phase of the illness, and its effect on growth and maturity.[9]

One purpose is extremely clear. As stated from the American Academy of Pediatrics, the design for pediatric palliative care services must be carefully considered, because children differ in a number of ways from adults.[10] Focusing on children affected by cancer, nearly 2000 children die of a neoplasm in the United States of America annually, while about 16,000 children are expected to receive a new cancer diagnosis in the same period.[2] The same figure in EU talks about 25,071 new diagnoses and 3133 expected deaths,[11] while in Italy about 1500 children develop cancer every year, and about 320 of them die every year[12] (population data for 2014: USA, 321 million; EU, 503 million; and Italy, 61 million).

The major categories of pediatric cancer are as follows:

Leukemia	30%
Central nervous system (CNS)	26%
Neuroblastoma	6%
NHL	5%
HL	3%
RMS	3%
Osteosarcoma	2%
Ewing sarcoma	1%

Survival for all invasive childhood cancers combined has improved markedly over the past 30 years due to new and improved treatments. The 5-year relative survival rate increased from 58% in the mid-1970s to 83% in the most recent period (2004–2010). However, rates vary considerably depending on cancer type, patient age, and other characteristics. The 5-year survival rate among children of age 0–14 years with retinoblastoma is 97%, Hodgkin lymphoma is 97%, Wilms tumor is 90%, non-Hodgkin lymphoma is 88%, leukemia is 85% (89% for lymphoid leukemia and 64% for acute myeloid leukemia), neuroblastoma is 79%, Ewing sarcoma is 75%, brain and other central nervous system tumors is 72%; osteosarcoma is 71%, and rhabdomyosarcoma is 68%. Mortality rates for childhood cancer have declined by 67% over the past four decades, from 6.3 (per 100,000) in 1970 to 2.1 in 2011 and the substantial progress in reducing childhood cancer mortality is largely attributable to improvements in treatment and high rates of participation in clinical trials.[2]

The majority of deaths are caused by progression of initial tumor or metastatic disease and quite all patients receive palliative treatment during the last months of life, including different and/or multiple surgical procedures. In this respect, the role of surgery in palliative care is to reach if possible relief of symptoms respecting or restoring the organ function improving quality of life. Next it is possible to include in the same course prolongation of quality-adjusted and progression-free survival.

Palliation is among the most challenging tasks pediatric oncologists have to accomplish. End-of-life measures usually come after an articulated process of aggressive treatments and require a significant shift in clinical management. In this scenario, quality of life becomes the highest goal to be accomplished.[13]

The decision to perform a palliative surgical oncology procedure in a symptomatic patient is a frequent challenge for surgical oncologists and it requires a high level of surgical judgment and surgical expertise, and must consider the following:

(1) indications for the operation;
(2) prognosis of the disease;
(3) whether the benefits of the procedure outweigh the risks;
(4) anesthesiological risk; and
(5) do-not-resuscitate (DNR) status.

Surgical palliation should be discussed within a comprehensive multidisciplinary palliative care plan to achieve the best outcomes and maximize patient satisfaction. Surgical palliative care education should be an integral part of the surgical residency curricula and requires the development of specific competency assessment tools. Education of surgeons in training about palliative care is an emerging field. There is consensus that a competent surgeon will have a basic level of skill in meeting the needs of all of those patients with life-threatening illnesses, including the dying patient. As a matter of fact, early integration of palliative care as a standard approach for children and adolescents diagnosed with cancer carries meaningful opportunities to improve symptom control and quality of life for patients and their families. Children and adolescents living with cancer and their families should receive early, integrated access to family-centered palliative care concepts (symptom assessment and intervention; direct patient report; effective communication; and shared decision-making) to minimize symptom burden, ease suffering, effectively manage pain, and provide preventative bereavement care. The essence of all of these palliative care efforts seems to be “preservation of dignity.”[14] The goal is to make palliative care “usual care” in advanced illness, also for children.