Improving aspects of palliative care for children
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Chapter 1

General Introduction
Introduction

Despite drastic improvements in medical care, there will always be children who die of their disease before reaching adulthood age. In the developed world, about 20-25% of pediatric oncology patients cannot be cured and will die as a result of the disease. In the Netherlands, this leads to about 110-140 patients per year\textsuperscript{1,2}. Also, there are numerous metabolic, neurodegenerative, chromosomal and other diseases which cannot be treated, and will result in an early death, which accounts in the Netherlands for about 1000 deaths per year\textsuperscript{3,4}. Besides, medical improvements have led to longer survival of these patients resulting in an increased prevalence of children with life-shortening disease\textsuperscript{5,6}. These patients, about 4000-6700 each year in the Netherlands, are entitled to receive palliative care\textsuperscript{7,8}.

Over the last 20 years, there has been an increasing awareness of pediatric palliative care which focusses on alleviation of suffering of children\textsuperscript{9-12}. Unfortunately, there is no consensus on what pediatric palliative care should comprise\textsuperscript{9}. In literature, different definitions of palliative care are used, among which the WHO definition\textsuperscript{10,13-16}. In this study we have used the following definition of palliative care for children, which is based on the palliative care guidelines of the WHO, AAP and ACT\textsuperscript{10,13,14}.

“Palliative care for children (0-18) is the active, total approach of care for children with life threatening or life shortening disease. The care for the child and/or the family includes physical, psychological, social, pedagogical, cultural and spiritual aspects. The care is aimed for the highest possible quality of life of the child and the family. There has to be attention for palliative care from the moment of diagnosis. The palliative care should be continuous, regardless of transitions in location, caregiver and/or perspectives of care. Effective palliative care demands a coordinated, broad approach by a multidisciplinary team, which includes the family.”

“Palliative terminal care for children is part of palliative care for children. An essential aspect is learning to handle the approaching death and to conclude the life of the child as positive as possible; this focus has to be present in the contacts between the child/family and the involved caretakers. Also bereavement support is an essential part of this care.”

In this thesis we present the results of several studies performed within the PANDA study (Palliative care, ANticipated Dedicated and Adapted) a collaborative research initiative between groups of the Emma Children’s Hospital of the Academic Medical Center and the Julius Center for Health Sciences and Primary Care of the University Medical Center. We focus on children with life-shortening disease, either malignant disease (MD) or non-malignant disease (NMD), whom are not expected to reach the age of independent adulthood and who are entitled to receive palliative care. Palliative care of neonates is beyond the scope of this thesis. This thesis comprises three main topics that all contribute
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to improving the quality of pediatric palliative care; 1) Insight in the palliative course of disease of children with malignant disease, 2) Insight in the intensity and content of care provided by a pediatric palliative care team (PPCT), and 3) Compliance of the PPCT to the national clinical guideline pediatric palliative care.

**Insight in the palliative course of disease of children with incurable malignant disease**

Pediatric palliative care (PPC) should be anticipating, establishing goals of care and discussing end-of-life issues\(^{11,17,18}\). To be able to anticipate, caregivers need information on what course of disease can be expected. Questions such as, how long will the palliative phase last, will the patient be able to stay home, is the patient likely to be able to die at home and with which supportive management, what symptoms will the patient have to endure, need to be answered. Information is needed on the diversity and aggravations in symptoms that can be expected and what the likelihood is that these aggravations will occur. “Effective symptom management for all patient groups requires a deeper understanding of all possible symptoms that might occur in addition to a comprehensive analysis of the various factors influencing the well-being of the child and the family, be it in a clinical or home setting”\(^{19-21}\). Little data is available on pediatric palliative care and symptom management, particularly in home care settings\(^{22,23}\). Most published studies depict symptoms of patients with MD in a hospital setting\(^{19,24-26}\). Studies describing occurring symptoms, often do not differentiate between malignant- and non-malignant disease\(^{27-29}\). Moreover, most reports on malignant disease fail to differentiate between different types of tumor\(^{26,30-36}\). Only few studies differentiate between different tumor groups\(^{5,25,37-40}\), or present tumor specific symptoms\(^{41,42}\). The Center to Advance Palliative care, recommends the collection and analysis of various domains of metrics to provide the necessary data\(^{19}\).

Insight in the course of disease is also needed to inform parents sufficiently. Different studies showed that parents of children with advanced cancer rated the quality of care provided by oncologists more highly when they felt they had received clear information about what to expect during the end-of-life period, and when parents felt prepared for the circumstances surrounding their child’s death\(^{43,44}\). Similarly, Kassam et al found that a majority of parents who cared for a child with advanced cancer highly valued receiving anticipatory guidance about death and dying by their healthcare team\(^{45}\). In yet another study, parents clearly voiced the value of understanding the medical course and options, having time to reflect on the decisions, being able to more clearly communicate and control the process of care with multiple providers, and simply being relieved of the stress and anxiety that can come when the future is unclear\(^{46}\).

Insight in the palliative course of disease of children with MD may help to improve the quality of PPC by supporting care to discuss and anticipate on specific symptoms. Therefore, it is important to assess the trajectory of the palliative phase of children with incurable MD.
Insight in the intensity and content of care provided by the pediatric palliative case management team

Although there are several definitions and guidelines of what pediatric palliative care should look like13,18,47, there is still no uniformity on how this palliative care should be set up. Recent inventory studies have shown that the organization and coordination of palliative care is troublesome48. Parents need to arrange many things on their own initiative, which is at the expense of the time and energy they can and want to spend with their child. They also point out that caregivers lack expertise in palliative care and that cooperation between the first- second- and third- line care is insufficient8,49. It appears that parents feel the need to take care of an essential part of the daily care of the child, including complex nursing care8,38. There is evidence of the risk for parents to get a burn-out within a short period of time following the transition to becoming the child’s primary care giver, with the potential for physical or mental ill health50,51. Other research shows that palliative care givers are merely focused on physical care for the child, but pay limited attention to the psychological functioning of the child and parents38 and siblings29,44,45.

A central coordinating person is needed to relieve parents from the urge to coordinate the care themselves and allow parents to spend time with the child10,14,52. This coordinating person should be available for all problems regarding the palliative care of the child, should guide parents through the whole trajectory of disease, from diagnosis to death53. Internationally, several pediatric palliative care (PPC) teams have been set up aiming to fulfill this need, and provide a full palliative care service54-65. There is quite extensive research pointing out the advantages of these palliative care teams. When PPC teams were involved, parents of children with cancer reported less distress from pain, dyspnea, and anxiety at end-of-life (EOL)58. Other results are an improved quality of life56,59,61, that children were more likely to have fun and to experience events that added meaning to life66, and that communication between parents and professionals was improved44,67 with discussions on do-not-resuscitate decisions and end-of-life care performed earlier in the course of disease58. In addition, patients receiving PPC spend fewer days in the hospital before death due to both less and shorter hospital admissions61,63,68, were less likely to die in an intensive care setting68 and more likely to die in the preferred place of death55,57,58, accrued fewer average daily charges and underwent fewer diagnostic/monitoring procedures68, reducing health care costs and utilization61,69,70.

The pediatric palliative care team

To improve the quality of PPC in the Netherlands, in June 2012 The Emma Children’s university hospital initiated the first Dutch hospital-based multidisciplinary PPCT. The PPCT is located in a University children’s hospital in the capital of the Netherlands, and is a tertiary referral center for patients below the age of 19 with any type of life shortening disease. The PPCT comprises specialized nurses experienced and trained in PPC, child life specialists, a psychologist, a chaplain and a social worker71. Two pediatricians and two
pediatric oncologists are connected to the team, and join the weekly multidisciplinary conferences. The specialized nurses act as liaison case managers to organize PPC for the patient and provide emotional support. The case managers coordinate the logistics of care delivered from all involved professionals. Furthermore, the PPCT supports the first line professionals. The hospital based PPCT bridges the gap between hospital and home. Contacts can be undertaken by telephone, mail as well as personal visits at home or during hospitalizations.

The primary physician remains responsible for the patient’s medical treatment and will introduce the PPCT to all patients with a life shortening disease, following any category as set by the WHO\textsuperscript{13}, early in the palliative phase of disease. The support is continuous during the course of illness and also provides bereavement support after the patient’s death. The PPCT’s support does not replace care, but is offered to navigate the patient through the complex care.

Although the research on the benefits of PPC is quite extensive, literature on the contents of PPC is scarce. The formation and organization of care provided by a PPCT is generally based on expert opinions\textsuperscript{72}. There is no consensus on items such as, which disciplines should be represented in a PPCT, whether or not 24- hour coverage is needed, and where care should be provided. However, information on these topics is needed to create or adapt teams aligned to the patients and family’s needs. Therefore, research is needed on the different aspects of the PPCT, including intensity, timing and location of care provided by the PPCT, and disciplines needed in PPC.

**Compliance of the PPCT to the national clinical guideline pediatric palliative care**

“To ensure that childhood patients receive optimal care, clinical practice guidelines (CPGs) are essential\textsuperscript{73,74}. CPGs are defined as “statements that include recommendations intended to optimize patient care that are informed by a systematic review of evidence and an assessment of the benefits and harms of alternative care options”\textsuperscript{75}. CPGs aim to bridge the gap between research and clinical practice and are regarded as powerful tools to improve the quality of care\textsuperscript{73,76-80}. In addition, CPGs can contribute to a reduced variability in daily practice and costs\textsuperscript{73,74,81,82}.

Unfortunately, within pediatric palliative care, uniform guidelines are scarce. The ACT, AAP and WHO have set general guidelines which should provide guidance in how to deliver palliative care\textsuperscript{10,13,14}. However, these guidelines provide general directions instead of practical guidelines which can be implemented in daily clinical practice. A recent systematic review of published clinical practice guidelines on pediatric palliative care reveals that many clinical practice guidelines are from single institutions, often focusing on coordination of care, continuity of care, and early integration of palliative care\textsuperscript{83}. However, very limited evidence for symptom treatment, of symptom treatment specific guidelines is available. Knops et al, have performed a systematic review on treatment options for symptoms within the palliative phase for children\textsuperscript{84}. They describe that as a result of the
scarce evidence available for treating children within the palliative phase, most evidence was retrieved from adult research. This systematic review was used to develop the Dutch guideline for pediatric palliative care, finalized in 201385.

Although CPGs are regarded as powerful tools to improve the quality of care, this effect is highly dependent on the compliance of the healthcare professionals to the guideline. Therefore, in our aim to seek improved quality of PPC, it is important to assess the compliance of our PPCT to the Dutch CPG for pediatric palliative care.

**Goals and aims of the thesis**

This thesis adds to the current literature by focusing on three aspects that are expected to contribute to the quality of palliative care for children with cancer.

**Goal 1.** Insight in the palliative phase of disease of children with incurable malignant disease (chapter 2 and 3)

**Aims**

- To obtain detailed insight in the palliative phase of children with an incurable brain tumor
- To obtain detailed insight in the palliative phase of children with an incurable non-cns malignancy

**Goal 2.** Insight in the intensity and content of care provided by the pediatric palliative care team (chapter 4 and 5)

**Aims**

- To obtain insight in the intensity and location of care provided by the PPCT
- To get insight in the contents and timing of care and the need for different disciplines in the PPCT.

**Goal 3.** Compliance of the PPCT to the Dutch guideline for pediatric palliative care (chapter 6)

**Aim**

- To assess compliance to the guideline with the use of quality indicators
- To assess whether an e-learning module can enhance the compliance to the guideline
Outline of the thesis.

The general introduction of this thesis is covered in Chapter 1. The chapters include information on the palliative phase of children with MD, insight in the contents of care provided by a pediatric palliative care team, and information on the introduction and usage of the first national guideline of pediatric palliative care.

Part one of this thesis, covers information on the palliative phase of children with cancer, is presented in chapter 2 and 3. In chapter 2 we aimed to provide insight in the palliative phase of children with an incurable brain tumor. We performed a retrospective chart research to find out what symptoms occurred, when they occurred and how they were managed. Moreover we retrieved information on the duration of the palliative phase, whether and when DNR discussions were held on whether the child was included in this discussion. Also, we assessed the planned and actual location of death. In chapter 3 we performed a comparable study for children with an incurable NMD. In this study we also compared the palliative phase and the occurring symptoms between children with CNS- and non-CNS tumors.

The second part of this thesis covers information on the pediatric palliative care team and is presented in chapter 4 and 5. In chapter 4 the pilot phase of our palliative team is described. In this retrospective study we looked at the intensity of care provided by the PPCT, the location where care was provided, and on what other case management activities time was spent. We compared these data for children with MD versus NMD. In chapter 5 we present a prospective study investigating the contents of case management consultation as performed by the PPCT. We asked the team-members to complete a questionnaire right after every contact the team had with parents or the patients. The study focused on subject discussed during the contact, for the timing of the contact, the disciplines contacted and who initiated the contact. Again the information was compared between patients with MD versus NMD.

In the third part of the thesis the compliance of the national pediatric palliative care guideline was tested. In chapter 6 we describe the development of quality indicators in order to test the compliance of the case managers of the PPCT to the guideline. We prospectively looked at the compliance of the case managers of the PPCT around one year after the start of the PPCT by reviewing the charts for clinical situations in which the guideline should have been used. To enhance compliance, we developed an e-learning module which was completed by the team members after the first assessment. After a subsequent interactive educational meeting for the PPCT members a second assessment period was performed, investigating whether the e-learning sessions indeed enhanced the compliance. This thesis ends with chapter 7; a general discussion including a summary of the results, main findings, limitations, future perspectives and the key messages.
Reference list


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