

PART A: BACKGROUND TO CLINICAL GUIDELINES PROJECT

1. Introduction

The *Guidance for Integrated Paediatric Palliative Care Services in New Zealand* was published in September 2012 by the Ministry of Health. The *Guidance* document identified an urgent need for development of clinical guidelines and care pathways for paediatric palliative care service delivery, and recommended that they be developed as a key priority. An *Evidence and Options* report, commissioned by the Ministry of Health from the Paediatric Society of New Zealand in 2013, indicated a need for robust development processes. This project responds to those recommendations.

It is intended that the guidelines are to be placed on the website of Starship Children's Health, so that they may be accessed by clinicians throughout the country. Hence, instead of using a conventional layout, this report is presented as a series of discrete documents, each of which is individually referenced. The documents are organised in two sections.

Part A, the **Background to the Clinical Guidelines Project**, introduces the components of the final report and briefly summarises key principles of palliative care delivery and the policy context. A **care framework**, which was developed as a component of this project, sets out a three-stage care palliative care continuum. For each stage, criteria for entry, key features and key interventions are provided. The second stage embeds the clinical guidelines for end-of-life care.

Part B comprises a suite of discrete documents: (1) the **Introduction**; (2) the **Guidelines**; and (3) a **Development Methodology** document. So that they may be easily accessed by tertiary and secondary-level providers throughout the country, it is intended that these be placed on the Starship Children's Health website. Hence, each document is presented as simply as possible, without page numbers or other embellishment and with references attached to each. When the documents are translated as web pages, hyperlinks will be inserted in the places identified in the documents.

The first Part B document, the **Introduction**, summarises the clinical guideline rationale, purpose, target audience and target population. This introduction meets the quality requirements of the ADAPTE development instrument (see Development Methodology).

There follows a suite of **clinical guidelines** which are presented as stand-alone documents. The guidelines address: sharing significant news; planning care; pain assessment and management; symptom management; and psychosocial, spiritual and cultural care. Clinical guidelines address the following:

- Sharing significant news
- Planning care
- Introduction to children's pain at end of life
- Pain assessment
- Pain management
- Emergencies management
- Massive bleeding management
- Bowel obstruction management
- Seizure management
- Spinal cord compression management
- Superior vena cava obstruction management
- Agitation management
- Breathlessness management

- Nausea and vomiting management
- Nutrition and hydration management
- Upper airways secretion management
- Psychosocial care
- Spiritual care
- Cultural care

The final Part B document is a **Development Methodology**, which describes the clinical guideline development methodology and process. The ADAPTE (ADAPTE Collaboration, 2009) and AGREE II (Brouwers et al., 2010) instruments, which guided development, and the development process, are described.

2. Background

The care framework and clinical guidelines for end-of-life care were developed in response to recommendations made by the *Guidance for Integrated Paediatric Palliative Care Services in New Zealand* (MOH, 2012). The *Guidance* document provides a comprehensive rationale for a New Zealand model of paediatric palliative care delivery. This section captures that information relevant to end-of-life clinical guideline development.

2.1 Life-limiting illness trajectories

A widely-acknowledged typology of life-limiting diseases influences children’s palliative care service development and delivery. The typology comprises four groups, each of which presents a different illness trajectory:

Disease groups and trajectories

Disease group	Examples	Illness trajectory
1. Diseases for which curative treatment may be feasible but may fail	Cancer Irreversible organ failure of heart, liver, kidney	Temporary deterioration following treatment, with subsequent recovery/remission and relapse. Partially successful further treatment followed by deterioration and death.
2. Diseases for which premature death is anticipated but intensive treatment may prolong good quality life and participation in normal childhood activities	Cystic fibrosis Muscular dystrophy HIV/AIDS	Condition maintained with treatment over some years. Severe exacerbations characterise deterioration.
3. Progressive diseases without treatment options, for which treatment is exclusively palliative and may extend over many years	Batten’s disease Mucopolysaccharidoses	Some improvement following initial treatment, followed by steady deterioration.
4. Diseases with severe neurological disability which may cause weakness and susceptibility to complications. While not progressive, these diseases	Disabilities that follow brain or spinal cord injuries, such as severe cerebral palsy	Step-wise deterioration with plateaux.

lead to vulnerability and complications likely to cause premature death		
---	--	--

Sources: ACT & RCPCH, 2007; Goldman, 1998:49; Field and Behrman, 2003; Hynson, 2006)

2.2 Palliative care needs of children and their families/whanau

The delivery of palliative care services to children and their families/whanau acknowledges particular needs. These are:

- A small number of children die, compared with the number of adults
- Life-limiting conditions are diverse and, often, rare and unpredictable
- The child may survive into early adulthood
- Many conditions are long-term, extending over months and years and requiring decision-making at different points of the disease trajectory
- Some illnesses are familial in nature and affect more than one child in a family
- Care must include siblings and parents, who carry the burden of care
- Consideration must be given to the developmental nature of childhood, and should address the impact of physical, emotional and cognitive development on understanding of illness and death
- Educational needs require complex multidisciplinary interactions (ACT & RCPCH, 1997; Himelstein et al., 2004; McConnell et al., 2004)

2.3 Principles of paediatric palliative care

Palliative care and its principles are defined by the World Health Organization as:

- Palliative care for children is the total active care of the child's body, mind and spirit, and also involves giving support to the family
- It begins when the illness is diagnosed and continues regardless of whether or not a child receives treatment directed at the disease
- Health providers must evaluate and alleviate a child's physical, psychological and social distress
- Effective palliative care requires a broad multidisciplinary approach that includes the family and makes use of available community resources; it can be successfully implemented even if resources are limited
- It can be provided in tertiary care facilities, in community health centres and even in children's homes (WHO, 2002)

New Zealand has developed the WHO definition to acknowledge: the Treaty of Waitangi and Maori models of health; the evolution and development of palliative care; integrated service delivery by generalist and specialist providers; acknowledgment of diverse cultural beliefs and practices in New Zealand; and continuity of care by generalist providers. The New Zealand working definition of palliative care is:

- Care for people of all ages with a life-limiting illness which aims to:
- Optimise an individual's quality of life until death by addressing the person's physical, psychosocial, spiritual and cultural needs
- Support the individual's family, whanau, and other caregivers where needed, through the illness and after death
- Palliative care should be provided according to an individual's need, and may be suitable whether death is days, weeks, or, occasionally, even years away. It may be suitable sometimes when treatments are being given aimed at improving quality of life.

- It should be available wherever the person may be.
- It should be provided by all health care professionals, supported where necessary, by specialist palliative care services.
- Palliative care should be provided in such a way as to meet the unique needs of individuals from particular communities or groups. These include Maori, children and young people, immigrants, refugees, and those in isolated communities (NZ Cancer Treatment Working Party, 2007)

2.4 Principles of service delivery

There is wide international agreement that children, young people and their families with palliative care needs require service responses that provide:

- Access to competent care
- Continuity of care that links local and specialist services and intersectoral services
- Care delivered over long illness courses that addresses transitions of illness
- Respite care and caregiver support
- Care at the time of death
- Bereavement care
- Relief from pain and other distressing symptoms
- Care that meets developmental needs
- Emotional and spiritual support
- Help to live as normal a life as possible
- Help with decision-making
- Timely and truthful information
- Family support that respects family choices, values and cultural traditions, and that addresses family structure and support, location and access to health services
- Help with financial and practical concerns
- Transition for adolescents to adult services
- Care that meets the needs of ethnic minority communities
- Continued research and education (ACT & RCPCH, 1997; American Academy of Pediatrics, 2000; Field and Behrman, 2003; Hynson, 2006)

The *Guidance* document (MOH, 2012) sets out the vision for paediatric palliative care in New Zealand as:

All children with palliative care needs and their families/whanau have timely access to high-quality palliative care services



Six key principles frame service delivery. These are:

- A service framework utilises a paediatric palliative care network and links levels of care
- Services are coordinated and flexible
- Services are equitable and needs-based
- Services are of high quality
- Services are culturally-anchored
- Services are fiscally responsible and appropriately resourced (MOH, 2012)

3. Care Framework

The framework, which was developed as a component of this project, locates end-of-life care as the second stage of the palliative care “journey”. Drawing from the UK-focused *Core Care Pathway*

(Together for Short Lives, 2012), the framework identifies three stages. Within each, it identifies conditions of entry, features and key palliative care interventions. Because of the uncertainty of illness trajectories in life-limiting conditions, progress is not linear: a child may, sometimes repeatedly, move between Stage 1 and Stage 2.

STAGE 1: LIVING WITH A LIFE-LIMITING CONDITION	
<p>Entry Recognition of a life-threatening illness, based on the clinical questions: would it be surprising if this child died: (1) in the next year; (2) during this episode of care; or (3) prematurely because of a life-limiting illness?</p>	<p>Key palliative care interventions</p> <ul style="list-style-type: none"> • Share significant information with child and family • Support complex needs via cycle of assessment, planning, intervention and reassessment • Manage pain and other symptoms • Anticipate, and prepare for, deterioration • Complete advance care planning • Provide psychological, emotional, spiritual, social and cultural support to child • Provide psychological, emotional, spiritual, social and cultural support to family/whanau
<p>Features Concurrent delivery of life-sustaining interventions and quality of life measures Continuity of care in hospital and community settings Coordination of medical care and social support by palliative care or other paediatric clinician Honest and timely information Long-term care that addresses illness transitions Care that meets developmental needs Support to live life as normally as possible Interdisciplinary teams</p>	
	
STAGE 2: END OF LIFE	
<p>Entry Life-sustaining interventions no longer offer benefit The child may die soon</p>	<p>Key palliative care interventions</p> <ul style="list-style-type: none"> • Share significant information with child and family • Complete advance care planning • Support complex needs via cycle of assessment, planning, intervention and reassessment • Manage pain and other symptoms • Provide psychological, emotional, spiritual, social and cultural support to child and to the family/whanau • Provide knowledge and skills about care of the child to family/whanau • Provide information about after-death and funeral arrangements to family/whanau • Anticipate, and plan for, death of child
<p>Features Condition-dependent Focus: comfort and quality of life Coordination of medical care and social support by palliative care lead clinician 24/7 support Minimisation of suffering Maintenance of comfort Honest and timely information Care that meets developmental needs Care of the child at the time of death Support of the family/whanau at the time of death</p>	
	
STAGE 3: AFTER DEATH	
<p>Features Care of the body of the child Grief and bereavement of family/whanau</p>	<p>Key palliative care interventions</p> <ul style="list-style-type: none"> • Provide psychological, emotional, spiritual and cultural support to family/whanau • Assess bereavement needs and make appropriate referral (if required)

4. References: Part A

American Academy of Pediatrics Committee on Bioethics and Committee on Hospital Care. 2000. Palliative care for children. *Pediatrics* 106(2):351-357 . Cited in: Ministry of Health. 2012. *Guidance for Integrated Paediatric Palliative Care Services in New Zealand*. Wellington: New Zealand Ministry of Health

Association for Children with Life-threatening or Terminal Conditions and their Families (ACT) and Royal College of Paediatrics and Child Health. 2003. *A Guide to the Development of Children's Paediatric Care Services*. Bristol: ACT. Cited in: Ministry of Health. 2012. *Guidance for Integrated Paediatric Palliative Care Services in New Zealand*. Wellington: New Zealand Ministry of Health

ADAPTE Collaboration. 2009. *The ADAPTE Process: Resource Toolkit for Guideline Adaptation*. Version 2.0. Available at: <http://www.g-i-n.net>

Brouwers M, Kho ME, Browman GP, Burgers JS, Cluzeau F et al, for the AGREE II Consortium. 2010. Advancing guideline development, reporting and evaluation in healthcare. *Canadian Medical Association Journal* 182E:839-42 doi.10.1503/090449

Field Marilyn J and Behrman Richard F (eds). 2003. *When Children Die: Improving Palliative and End-of-Life Care for Children and their Families*. Washington DC: National Academies Press. Cited in: Ministry of Health. 2012. *Guidance for Integrated Paediatric Palliative Care Services in New Zealand*. Wellington: New Zealand Ministry of Health

Goldman Ann.1998. ABC of palliative care: Special problems of children. *British Medical Journal* 316:49-52. Cited in: Ministry of Health. 2012. *Guidance for Integrated Paediatric Palliative Care Services in New Zealand*. Wellington: New Zealand Ministry of Health

Himmelstein Bruce, Hilden Joanne M, Boldt Ann M and Weissman David. 2004. Pediatric palliative care. *New England Journal of Medicine* 350(17):1752-1762. Cited in: Ministry of Health. 2012. *Guidance for Integrated Paediatric Palliative Care Services in New Zealand*. Wellington: New Zealand Ministry of Health

Hynson Jenny L. 2006. The child's journey: transition from health to ill-health. In: Goldman Anne, Hain Richard and Liben Stephen (eds). *Oxford Textbook of Palliative Care for Children*. Oxford and New York: Oxford University Press . Cited in: Ministry of Health. 2012. *Guidance for Integrated Paediatric Palliative Care Services in New Zealand*. Wellington: New Zealand Ministry of Health

McConnell Yarrow, Frager Gerri and Levetown Marcia. Decision making in pediatric palliative care. In Carter Brian S and Levetown Marcia (eds). 2004. *Palliative Care for Infants, Children and Adolescents: A Practical Handbook*. Baltimore and London: John Hopkins University Press. Cited in: Ministry of Health. 2012. *Guidance for Integrated Paediatric Palliative Care Services in New Zealand*. Wellington: New Zealand Ministry of Health

Ministry of Health. 2012. *Guidance for Integrated Paediatric Palliative Care Services in New Zealand*. Wellington: New Zealand Ministry of Health

Ministry of Health. 2013. *Paediatric Palliative Care Clinical Guidelines and Care Pathways: Evidence and Options*. Unpublished document of Ministry of Health

New Zealand Cancer Treatment Working Party. 2007. *New Zealand Palliative Care: A Working Definition*. Unpublished document of the NZ Cancer Treatment Working Party. Cited in: Ministry of Health. 2012. *Guidance for Integrated Paediatric Palliative Care Services in New Zealand*. Wellington: New Zealand Ministry of Health

Together for Short Lives. 2012. *A Guide to End of Life Care*. Bristol: Together for Short Lives. Cited in: Ministry of Health. 2012. *Guidance for Integrated Paediatric Palliative Care Services in New Zealand*. Wellington: New Zealand Ministry of Health

World Health Organization. 2002. *WHO Definition of Palliative Care*. Available at: <http://www.who.int/cancer/palliative/definition/en/> Accessed 6.9.14