

A practical guide to
PALLIATIVE CARE
in paediatrics

FOREWORD

The progress in Paediatric Palliative Care in Australia since the first publication of *A Practical Guide to Paediatric Oncology Palliative Care* in 1999, has been substantial with the continued development of clinical services throughout Australia, and the on-going review of the quality of those services through the National Standards Assessment Program, [NSAP (<http://www.palliativecare.org.au/Standards/NSAP.aspx>)] which provides a method for each service to benchmark. Further, the development has been towards delivering care at home, or as close to home as possible, as novel communication technologies and education initiatives evolve.

This edition, incorporating care for both children with cancer and non-cancer conditions, is testimony to its value as an education resource, empowering clinicians caring for children at home or as close to home as possible. Coordinated by the Paediatric Palliative Care Service at the Royal Children's Hospital, Brisbane, it also represents the collaborative efforts of paediatric palliative care clinicians throughout Australia and New Zealand. As such, it is a national resource and a major contribution towards supporting dying children and their families to have the best quality of life.

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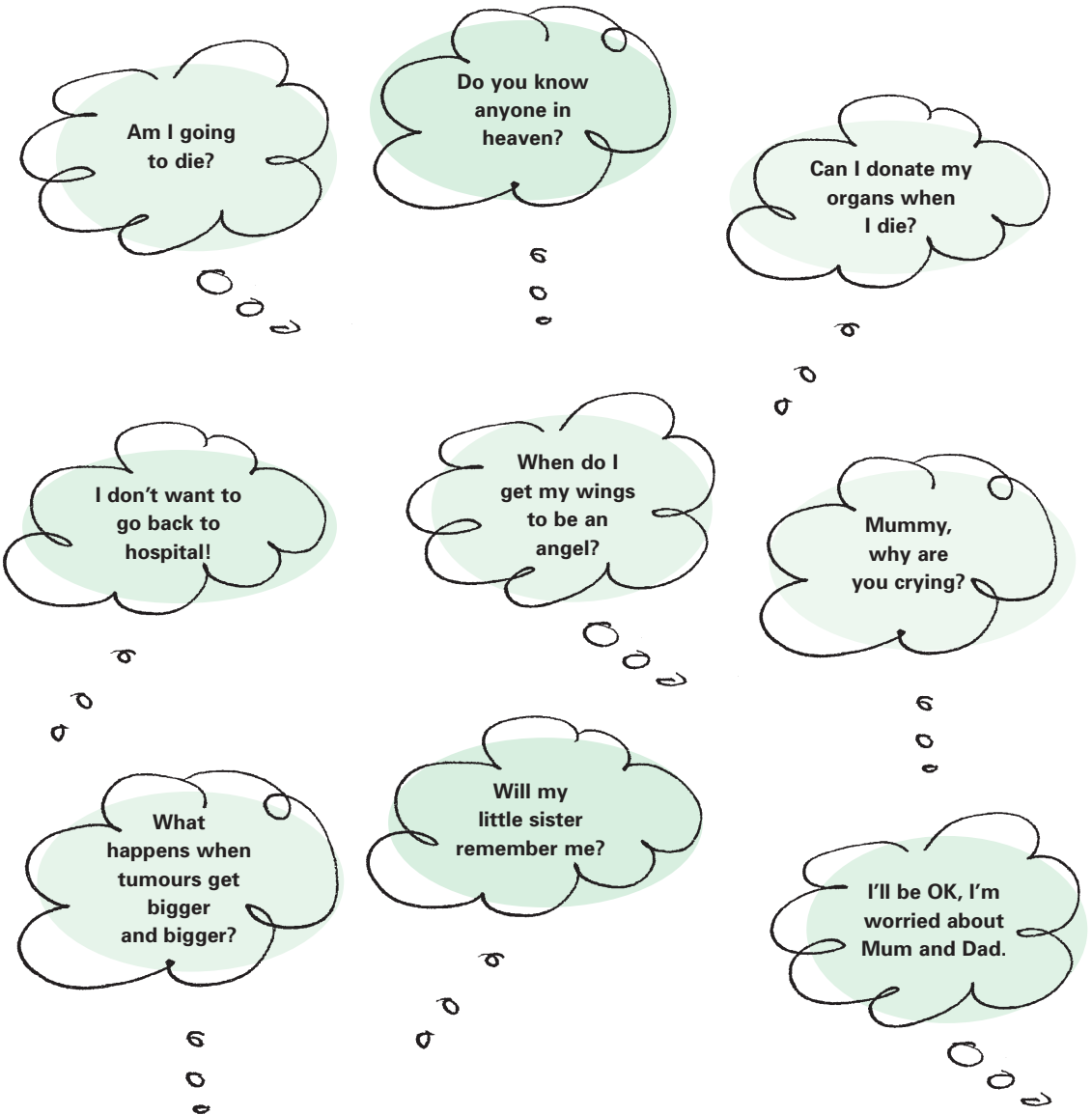
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INTRODUCTION

Children are not expected to die. When faced with the news that a child has a progressive incurable disease it is natural for the child, family and health professionals to have numerous fears, concerns and questions about what will happen. The aim of this book is to highlight and address many of these issues.

DYING CHILDREN AND TEENAGERS



PARENTS AND SIBLINGS

How long have we got left?

How do we tell him and our other children he is dying?

How do we know she has died?

He hasn't eaten for days, shouldn't we do something?

Isn't morphine addictive?

What do we do if he has a fit?

Will she have pain as death gets closer?

Do we phone the police if he dies at home?

How long will we grieve?

INTRODUCTION

HEALTH PROFESSIONALS AND SCHOOL TEACHERS

How do we tell his classmates he is dying?

How do you talk to a child about dying?

What do you do when parents choose not to tell their child they are dying?

What should we do with his school desk now he has died?

How do staff cope with children dying?

How will I know he has pain?

When should you stop giving antibiotics and blood?

Do parents ever get over this?

INTRODUCTION

Palliative care as a speciality is gaining a reputation and recognition for the positive outcomes that this approach to care can deliver to patients and their families.^{1,2}

Paediatric palliative care from its outset has shared the same philosophy of care as the adult specialty, (e.g. multi-disciplinary care, maximizing supports at home and collaborative practice) while at the same time developing its own distinctive model.

For example, more than half of the patients a paediatric palliative care service care for will have non-cancer life-limiting conditions which are not seen in adult practice. Paediatric palliative care services will usually remain consultative, including the end of life phase of care, and collaborate closely with other health care teams involved in the child's care.³ Furthermore, parental desire for resuscitation does not preclude involvement of a paediatric palliative care service in the care of a child with a life-limiting condition.⁴

The types of diseases encountered, the symptoms experienced, and the way children understand and communicate about their illnesses, are all different. These differences need to be acknowledged and inform the care provided to dying children and their families.

It is our experience that many children and families express a wish for their child to die at home in an environment that enhances their sense of security and normality. Dying at home can also increase the opportunity for parents, siblings, friends and family to assist with care, and thereby optimise the child's quality of life.⁵ However, some children and families will prefer not to die at home and flexible options of care should be available such as, tertiary hospital, hospital closest to home, and if available, hospice.⁶ It is important that children and families receive care appropriate to their needs at the right

time in the place of their choice ("right care, right place, right time"). Such choice in planning for location of care at end of life will reduce regrets that families may experience after their child dies.⁷

The geographical vastness of Australia often means that a child who is dying at home may be hundreds of kilometres from the tertiary paediatric hospital where they received their diagnosis, management and/or treatment. In these situations, health professionals from the child's local community are required to meet the palliative care needs of the child and family. This can present a challenge to the health professional because of the relatively small number of children who die each year which limits the opportunity for regional and rural health professionals to develop experience in paediatric palliative care.

It is for this reason that "A Practical Guide to Palliative Care in Paediatrics" has been developed.

This guide addresses the many and varied aspects of caring for dying children and their families. This includes pain and symptom management, practical supports, psychosocial issues, communication and available resources. It is intended to be user-friendly and complementary to the existing knowledge and resources of the health professionals who use it.

Due to the success of the first and second editions of "A Practical Guide to Palliative Care in Paediatric Oncology", the current edition has been produced to incorporate aspects of palliative care for both cancer and non-cancer life-limiting and life-threatening conditions. Contributions have been received from all states of Australia and New Zealand. It is hoped that this national approach will benefit children and families, and those caring for them, anywhere in the country – irrespective of diagnosis.

For the purpose of this book, a child has been defined as a young person up to their 19th birthday. The specific upper age range of children managed by a paediatric palliative care service can vary between jurisdictions. There may be some flexibility depending on whether the child is currently being managed by a paediatric team or is still attending school.⁸

Children's palliative care has been defined as:

"Palliative care for children and young people with life-limiting conditions is an active and total approach to care, from the point of diagnosis or recognition, throughout the child's life, death and beyond. It embraces physical, emotional, social and spiritual elements, and focuses on the enhancement of quality of life for the child/young person and support for the family. It includes management of distressing symptoms, provision of short breaks and care through death and bereavement." (Together for Short Lives, UK).⁸

The World Health Organisation offers a similar definition and makes the following additional points:

- Palliative care for children is the active total care of the child's body, mind and spirit, and also involves giving support to the family.
- It begins when 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 in children's homes.⁹

A key concept, and one that is not always well understood, is that palliative care can be integrated with on-going efforts to cure or modify disease. The child and their family are supported to live as well as they can for as long as they can, within the limits imposed by the illness. In this context palliative care is about both living and dying.

Categories of life-limiting and life-threatening conditions

Life-limiting conditions are those for which there is no reasonable hope of cure and from which children or young people will die. Some of these conditions cause progressive deterioration over months to years, rendering the child increasingly dependent on parents and carers.^{8, 10}

Life-threatening conditions are those for which curative treatment may be feasible but can fail (e.g. cancer). Children in long-term remission or following successful curative treatment are not included.^{8, 10}

There are four broad groups of life-threatening and life-limiting conditions (see Table 1). Categorisation is not always easy and the examples used are not exhaustive. Some patients can be classified by more than one group. Diagnosis is only part of the process – the spectrum and severity of the disease, subsequent complications, and the needs of, and impact on the child and family need to be taken into account.

CONDITIONS

TABLE 1 Categories of life threatening and life limiting conditions¹⁰

Group	Definition	Example
1	Life-threatening conditions for which curative treatment may be feasible but can fail.	Children with cancer when treatment fails (e.g. stage 4 neuroblastoma). Irreversible organ failure (not amenable to transplantation or if transplantation is unsuccessful).
2	Life-limiting conditions where premature death is inevitable. However there may be long periods of intensive treatment aimed at prolonging life and allowing participation in normal activities.	HIV, complex cardiac disease and Duchenne muscular dystrophy (DMD). Ongoing research and medical improvements have meant that some patients with Cystic Fibrosis are surviving into their 40's and beyond. A similar trend is seen with DMD.
3	Life-limiting, progressive conditions without curative treatment options, where treatment is exclusively palliative and may commonly extend over many years.	Neurodegenerative conditions (e.g. Batten disease), metabolic conditions (e.g. mucopolysaccharidoses) and neuromuscular conditions (e.g. spinal muscular atrophy type 1).
4	Irreversible but non-progressive life-limiting conditions causing severe disability, leading to susceptibility of health complications and likelihood of premature death. Complications that may cause death include severe recurrent pneumonias or intractable seizures.	Severe cerebral palsy or multiple disabilities (such as following brain or spinal cord injury).

Fortunately, death rates reduced by more than half for infants and children between the years of 1996 and 2010 in Australia. In 2010, there were 1229 deaths of infants aged less than one year, a mortality rate of 4.1 per 1000 live births. Infant deaths comprised almost three-quarters of deaths among children aged 0–14 years.

There were 507 deaths of children aged 1 to 14 years; a rate of 13 per 100 000 children. The death rate for children aged 1 to 4 years (19 per 100 000 children) is twice the rate for children aged 5 to 14 years. Among children aged 1 to 14 years in 2008–2010, the leading causes of death were injuries, cancer, and diseases of the nervous system.

Between 2004 and 2008, there were approximately 583 new cases of cancer diagnosed in children aged 0 to 14 years, each year representing a rate of 14 per 100 000 children. From 2008–2010, there were 274 deaths from cancer in children aged 0 to 14 years; a rate of 2.2 per 100 000 children, accounting for approximately 5% of all childhood deaths. The most common cancers causing death among children during this period were malignant neoplasm of the brain (33%), acute myeloid leukaemia (10%) and acute lymphoblastic leukaemia (9%).¹¹

Figure 3 shows the diagnostic categories based on organ system of children referred to the Queensland state-wide paediatric palliative care service over a two year period (2009–2011).

It can be difficult for treating teams to know when to refer a patient to a palliative care service. “Would you be surprised if this patient died within the next 6–12 months?” is a question which can act as a useful indicator of when palliative care would be appropriate. If the answer is ‘no’, a referral should be made.

FIGURE 1 Leading causes of infant mortality, 2008–2010¹¹

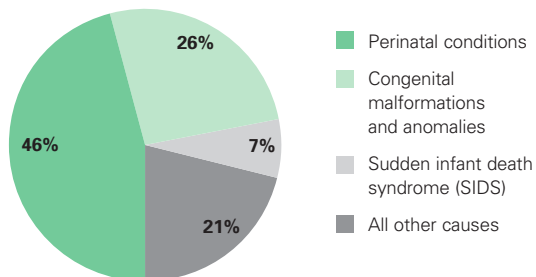


FIGURE 2 Leading causes of child mortality (1–14 years), 2008–2010¹¹

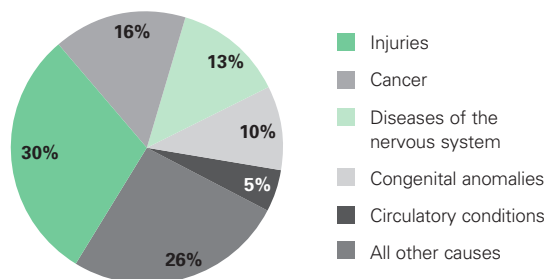
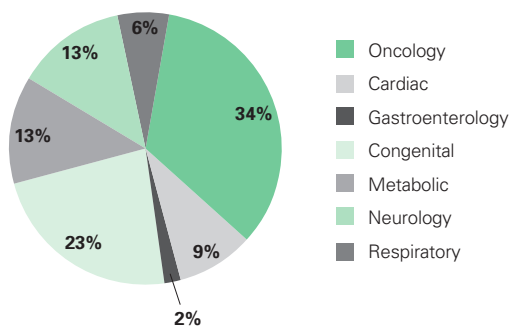


FIGURE 3 Diagnostic category of children referred to Queensland paediatric palliative care service 2009–2011¹²



EPIDEMIOLOGY

Table 2 shows examples of conditions for each of these diagnostic categories. It is not an exhaustive list as there are many other conditions which may be referred to a palliative care service.

TABLE 2 Examples of conditions within diagnostic categories which may be referred for palliative care

<i>Diagnostic category</i>	<i>Examples</i>
Oncology	Brain Tumours <ul style="list-style-type: none"> • Glioblastoma Multiforme • Diffuse Intrinsic Pontine Glioma Relapsed Leukaemia (especially after bone marrow/stem cell transplantation) <ul style="list-style-type: none"> • Acute Myeloid Leukaemia, • Acute Lymphoblastic Leukaemia • Congenital Leukaemia Stage 4 Neuroblastoma Progressive solid tumours e.g. alveolar rhabdomyosarcoma.
Congenital/Genetic	Trisomy 18 Trisomy 13 Asphyxiating thoracic dystrophy Potter Syndrome Some patients with Epidermolysis Bullosa Some patients with Rett's Syndrome Other rare chromosomal anomalies with known poor neurologic prognosis
Neurology	Progressive neuromuscular conditions <ul style="list-style-type: none"> • Muscular Dystrophy • Spinal Muscular Atrophy Type 1 and 2 Neuro-degenerative conditions <ul style="list-style-type: none"> • Batten Disease Leukodystrophy <ul style="list-style-type: none"> • Metachromatic • X-linked Adrenoleukodystrophy Severe Traumatic Brain injury Hypoxic Ischaemic Encephalopathy (moderate – severe) including neonatal onset Severe Cerebral Palsy (Gross Motor Function Classification System Level 4 or 5 with complex medical needs) Severe Encephalopathy or Encephalitis Intractable Seizure Disorders <ul style="list-style-type: none"> • Alpers' Syndrome Brain reduction Syndromes: <ul style="list-style-type: none"> • Anencephaly • Hydranencephaly Neuronal Migration Disorders <ul style="list-style-type: none"> • Lissencephaly • Schizencephaly

Continues...

TABLE 2 continued

Diagnostic Category	Examples
Metabolic (Stem cell transplantation is a therapeutic option for some but not all metabolic conditions)	Mucopolysaccharidoses <ul style="list-style-type: none"> • Hunter's/Hurler's disease • Sanfilippo syndrome Krabbe's disease Niemann-Pick disease Menke's disease Pompe Disease Tay Sachs disease Fabry's disease Sandoff's disease Mitochondrial disorders Methylmalonic Acidaemia (often require dialysis for renal failure and to remove toxic metabolites)
Cardiac	Hypoplastic left heart syndrome Cardiomyopathy (not amenable to transplantation) Severe pulmonary hypertension Ebstein's anomaly Pulmonary atresia (especially if associated with hypoplastic pulmonary arteries) Combination of cardiac diagnosis with underlying neurologic/chromosomal diagnosis
Gastrointestinal	Liver failure – not amenable to transplantation or post-transplantation. Short gut syndrome and TPN dependence – limited central venous access, recurrent sepsis or liver failure
Respiratory	Cystic Fibrosis – being considered for lung transplantation or symptomatic (e.g. pain or dyspnoea not responsive to disease-modifying therapy)
Dermatology	Epidermolysis Bullosa Restrictive dermopathy
Renal	Some children with End Stage Renal Disease (ESRD). Causes of ESRD include renal dysplasia, congenital nephrotic syndrome and autosomal polycystic kidney disease in younger children and glomerulonephritis, nephropathy, nephrotoxic injury and haemolytic uraemic syndrome in older children. Progressive chronic renal failure despite RRT (renal replacement therapy) or surgery. Bilateral renal agenesis.

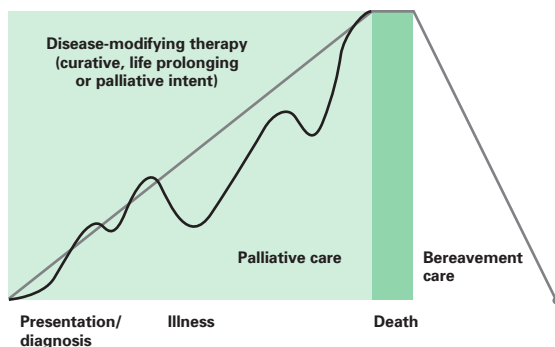
TRANSITION TO PALLIATIVE CARE

For many childhood life-limiting conditions, there is no definitive point at which a curative, or treatment orientated focus, changes to a palliative one. Palliative care is most effective for patients and their families when it is seamlessly integrated across the health care continuum. The transition is often more 'fluid' in nature and is directed by the child's illness. This transition varies widely from a child who has had a long curative approach to their illness with a relatively short palliative care period, to a child who, from diagnosis, is treated with a palliative focus over several months or even years.

Figure 4 illustrates the continuum of care for a child with a progressive condition. Treatment intended to modify the disease decreases as illness progresses, while palliative care increases as the child or young person reaches the end of life. Various components of the palliative approach can be provided to support the child and family during this entire period by the health professionals who provide frontline care for these children and their families. Such components include pain and symptom management, multi-disciplinary care, psychosocial care, spiritual support and various practical supports (e.g. assistance with finances and equipment). The curved line suggests that this is not a linear process. Further, highly technical invasive treatments may be used in conjunction with palliative care, to prolong life and improve quality with each becoming dominant at different stages of the disease.

After the child dies, bereavement support for parents, siblings, grandparents and friends becomes important. Parents experience grief and loss at the time their child is diagnosed with a life limiting condition and as the illness unfolds. As such, they can require anticipatory bereavement support before their child dies.

FIGURE 4 Continuum of care associated with disease modifying therapy and palliative care for a child with a progressive condition ^{10, 13, 14}



The care of a dying child requires the combined effort of the primary treating team, a palliative care service and health professionals in the child's local area. Early referral, good communication, and meticulous care planning enables health professionals to adopt the palliative approach with more confidence and competence.¹⁵ It also means those who will be providing care to the child at home have a chance to develop a trusting relationship with the child and family. Joint home visits and telemedicine consultations are extremely valuable in facilitating the sharing of knowledge and expertise.¹⁶⁻¹⁸

While integration of the principles of palliative care with disease-modifying therapies is the best approach for much of a child's illness, there generally comes a time when palliation becomes the dominant mode of care. Through gentle guidance, compassionate discussion, and often after several days of contemplation, a formal transition from a curative to a palliative focus begins. The child's medical team discusses the facts with the family and the child, if appropriate.

TRANSITION TO PALLIATIVE CARE

This would normally include a discussion as to what palliative care means for that particular child and family. This is generally a shared decision and occurs when all realistic curative avenues have been exhausted or if symptom burden is high.

Every child's situation will be different. For some, a rapid transition to exclusively palliative goals will occur (e.g. transfer from an intensive care unit to home for end-of-life care). For others, decisions regarding such interventions will be made in a step-wise fashion (e.g. initial withdrawal of invasive ventilation, progressing to withdrawal of non-invasive ventilation and other measures such as intravenous antibiotics).

An approach of "hoping for the best, while preparing for the worst" can be helpful. At this time the family's sense of hope may begin to evolve. The hope for a cure evolves into a hope for a pain-free and dignified death. In the perinatal context, parents can modify their hopes from having a healthy infant to other hopes, such as having time with, and holding their infant after delivery.¹⁹

It is important to be aware that while the child and family may appear to intellectually comprehend that death is now inevitable, they may have greater difficulty in emotionally accepting this.²⁰ Parents will experience feelings of shock and disbelief when their child dies, even when this has been anticipated for some time.

The move from a curative focus to palliative care is disease-directed but family driven. It is one of the most difficult decisions a family will ever have to make. Each family will respond to this transition in their own way. Some families are accepting of a palliative focus, while in others the desire for a cure remains dominant and all therapeutic options are explored and insisted upon. In many cases there is a combination of acceptance with hope for a 'miracle'. Each individual family's approach needs to be respected by staff caring for the child and family. Specialists in palliative care bring considerable experience and skill in talking to families about these difficult decisions. Health professionals who have known the child for some time, for example their General Practitioner (GP), also have an important role to play as trusted family confidantes. Many GPs will have experience in palliative care which can be of benefit to the child and family.

Some children can survive against all odds and patients do sometimes 'graduate' from the palliative care service. This model of care can allow children to receive the supportive benefits of palliative care when prognostic uncertainty exists. Evolving technologies coupled with the excellent care parents provide, mean that a number of adolescents with life limiting conditions are now transitioning to adult care services.



Understanding 'quality of life' involves making some kind of judgement about the satisfaction that people feel about their lives. This sense of satisfaction generally includes many dimensions such as quality of relationships, access to necessary resources (such as housing), physical health and comfort, access to activities which are intellectually, emotionally and physically fulfilling, the pursuit of important life goals, and issues of spirituality.

When quality of life is discussed in the setting of palliative care it has to be approached within the limitations imposed by the disease or condition. Even this is a relative concept. It is important to identify with the child and their family what the goals of care are, especially as the disease progresses. Discussions about quality of life should occur throughout the entire illness and take into consideration the wishes of both the parents and the child. This often allows the family and child to get on with living in the present and to be reassured that there are plans in place for future care.

It is important for the health professional to create and recognise such opportunities. It is also important to recognise that the goals of care for individuals within a family can be very different, and individuals within a family may protect each

other from discussions that involve considering the possibility of the death of the child.

Enabling quality of life is therefore a goal that the health professional should consider, but which the child and family should define for themselves. It is important not to superficially confuse quality of life with a concept of being busy, or with the achievement of particular activity. Quality of life is often more deeply connected with the quality of relationships and of time spent together, than it is with activity or outward achievement. The challenge however, is to enable families to achieve their sense of quality of life, and to be able to do this in the changing circumstances of progressive disease. In these circumstances, it is quite normal for the defining issues for families to change over time. At first, pursuing quality of life may focus on doing things the family thought they would have had more years to achieve (such as having special holidays or visiting theme parks). Later it may focus on achieving physical comfort and peaceful time together, away from the interruptions of visitors.

Whatever the case, the role of the health professional is to assist families to identify what quality of life means for them and to assist them to overcome blocks to achieving it. Importantly, as the disease progresses and the limitations on quality of life that are increasingly imposed, there will be elements of grief that the health professional can help the family to address.

In the early stage of a life-limiting condition, children can feel quite well and may have very few symptoms. They are likely to be at home with their parents, attending school and participating in family activities. Most children express the wish to have fewer trips to hospital. As the child's disease progresses and symptoms develop, closer follow up will be necessary. Depending upon where the family live this may be via telephone review, telehealth, home visits, or hospital visits. At times the child may need admission to a hospital (or hospice if available) for assessment and management of difficult symptoms, and for respite care.

It is important to discuss with the family, and if appropriate the child, where they would prefer the child to die. Some families presume their child will have to die in hospital. Families need to be informed of their choices and what supports are available to them.

Hospice

Children's hospices exist in some states of Australia. Hospices offer an additional choice for place of care beyond the home or acute hospital setting. These facilities can be a source of practical support, providing a home-like environment, with the back-up of health professionals on-site. They also offer psychosocial support for all members of the family. Examples of this type of facility are Bear Cottage in New South Wales, and the Very Special Kids House in Victoria.

Care at home

It is important that families know they can change their mind during the terminal phase of their child's illness about where they would like their child to die. This is particularly important for families who

choose home care. It can be very reassuring for families to know that there is always a hospital bed available if, for any reason, they feel hospital care is more appropriate. When developing a plan of care, it is important to ensure the family has access to appropriate contact details should they wish to move place of care urgently, or after-hours. For example, some families may decide to return home, or to the hospital, in the last few days or hours of life. It should be recognised that initially, some families may be fearful of having 'bad' memories if their child dies at home. However our experience demonstrates that this fear often decreases over time, and these families often choose to be at home despite their earlier concerns.

Health care professionals in this setting should be aware of a potentially difficult situation which can arise for some parents with a dying child. The parent may have made a very strong commitment to support the child dying at home, and may really struggle with this commitment if home care demands become too difficult either physically or emotionally. This possibility should be anticipated and openly discussed with the family to ensure that such a transition of care can occur, without inducing a sense of failure or guilt in the parents, or health professionals. The advantages of home care include:

- The opportunity for siblings and extended family to feel more involved in care.
- Parents have more control over the home situation and feel less judged by others.
- The child can be nursed in a familiar environment surrounded by the possessions, people and pets they love.
- Privacy.
- Spiritual and cultural needs can be more easily addressed.

The disadvantages include:

- The physical and emotional burden of care (parents may be more free to be parents when they are in a hospital or hospice).
- Some symptoms can be difficult (though not impossible) to manage at home (e.g. airway obstruction, severe haemorrhage).
- Health professionals are not immediately available to attend. Some parents may feel more secure with the round-the-clock availability of health professionals in the hospital or hospice rather than “on call” access to local community health professionals.

It is also important that there is ongoing contact between the hospital and home setting, as some families have described a sense of abandonment and insecurity when hospital visits become less frequent. This should be negotiated with the community palliative care service so as not to undermine their vital role.

Death in hospital

If death is to occur in hospital, it is important that where possible the child is nursed in a single room, and is able to personalise this space by bringing in their own doona, pillows, favourite toys, photos and other special items from home. The hospital option may include the hospital closest to the child’s home, or alternatively a ward environment where the child has received much of their care, and where the family trusts the care provided by the staff.²¹ Some families may prefer to be in a tertiary children’s hospital, with access to their specialists. Transfer of clinical information and handover of care is a critical step for children who are receiving palliative care when they are transferred from a tertiary children’s hospital to their local hospital.

Death in a paediatric intensive care unit

For some children, the “right place” to die will be the paediatric intensive care unit (PICU). This can be due to varying factors such as the acute onset of illness (e.g. trauma) or deterioration (e.g. sepsis or seizures), ongoing disease-modifying therapies, and the use of ventilatory support. Excellent symptom management, care co-ordination, multi-disciplinary support and bereavement care can, and should, be provided within the intensive care environment.²²

Within the PICU there can be a need for compassionate extubation.^{10, 23} This involves the discontinuation of mechanical ventilatory (and other critical care) support for a child with a poor prognosis. Where possible, and after consultation with PICU staff, families should be offered a choice as to where they would like their child extubated. Options could include the PICU, the home ward, quiet suite, hospice or home. It is important to note that some patients may be too unstable to move from the PICU setting despite their family’s hope for them to die in an alternative setting.

If the option is available, extubation may be possible at home. Members of the intensive care team can accompany the child in an ambulance to the home; this is sometimes referred to as a “reverse retrieval”. To support this process, there should always be an extended care plan to provide for a short-to-moderate duration of survival.

The quiet suite or room is a designated area within most children’s hospitals, often located near the PICU, which is home like. Patients can be extubated in such a room with privacy and dignity.

Further, families can stay with their child for an extended period after their child dies. If the body is to remain in the room for an extended time, (more than 12 hours) it may be possible to lower the temperature of the room to delay deterioration of the body.

Good symptom management and parallel planning are required at the time of extubation. While most patients will die shortly after extubation and cessation of ventilation, there are a small number of patients who may breathe spontaneously and live for a longer period of time. It is important families are aware of this possibility, and that the treating team ensure ongoing optimal symptom management.

Death in a neonatal intensive care unit

Likewise, in neonatology, the “right location” for end of life care for some neonates will be the Neonatal Intensive Care Unit (NICU). This may be due to prognostic uncertainty and the very gradual realisation by both parents and health professionals that a child is not responding to intensive care interventions.

However, it is important to recognise that when the goals of care transition from curative to palliative, many life-limiting and life-threatening conditions can be managed outside of the intensive care environment – either on post-natal wards, a hospital closer to home, at home (or, in a paediatric hospice, and some adult hospices).^{24, 25} Many families will opt for these settings of care if the appropriate support systems are put in place.

Wherever the place of care is, parents need access to 24-hour advice and support from health professionals from a variety of disciplines with experience in paediatric palliative care. Hospital and community based health professionals need to work together to assist the family care for their dying child. This requires meticulous planning.

Professional staff who may be involved in delivering paediatric palliative care include doctors, nurses, social workers, occupational and music therapists, physiotherapists, teachers and pastoral carers (Figure 5).²⁶

It is important to acknowledge that no one individual, or health discipline, can meet all the family's needs. However, by working together and sharing skills and expertise, a coordinated approach to care can be achieved.

To ensure that the family is not overwhelmed with services, it is important that a member of the team takes on the role of coordinator. The coordinator needs to be someone who will have ongoing involvement with the family during the palliative care phase. The specialist liaison nurse often takes on this role. However, in regional and rural areas it may be more appropriate for a local member of the team, such as the GP, paediatrician, ward nurse, or domiciliary nurse to assume this role. For families moving between care settings, it may be helpful to have a coordinator in each setting to ensure continuity of care and information transfer. This should also include a plan for sharing information and communication between visits.

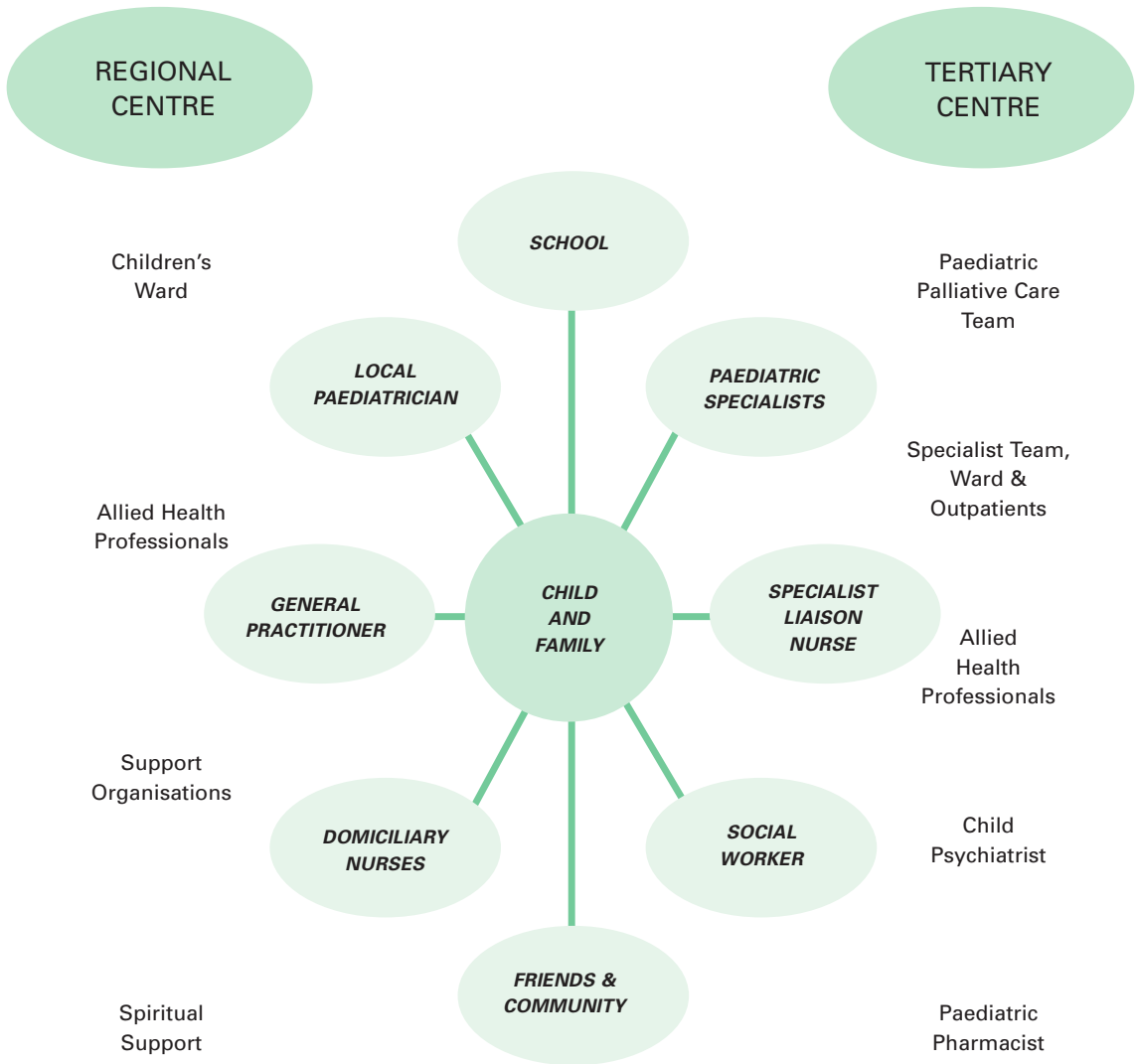
The coordinator needs to be easily accessible to the family as a first point of contact as new problems or concerns arise. A key role of the coordinator is to empower parents with the skills and knowledge needed to care for their dying child. The importance of this is discussed in further detail in the following section. The coordinator requires good communication skills, compassion and the ability to listen to the child, family and other health professionals caring for the child.

Knowledge of the child and family's support networks is important, as parents may ask for advice on how to involve their community, such as the school.

It is important that all members of the team are kept informed of the child's progress. Hospital paediatric ward and emergency department staff need to be kept up to date about the child's symptoms and know when a child may be admitted for acute symptom relief, or potentially terminal care. A palliative care plan may be a useful communication tool for this purpose. The coordinator can document the child's current symptoms and management in the plan, which should be made accessible to nursing staff working all shifts (Appendix 1). Domiciliary nurses and the child's GP need to know in advance when the child is returning home, and the type of care and support the family may require. They also need a primary contact identified for further advice or peer support. It is helpful to have a clear plan worked out with community teams for after the child dies. This includes who is responsible for paperwork, such as life extinct forms or death certificates, both during and after hours. This coordinated, flexible approach ensures that the child and family are well supported to receive care either at home or in hospital.

COORDINATION OF CARE

FIGURE 5 Model of family centred paediatric palliative care (adapted from Thompson A, et al²⁶)



The aim of this section is to provide some clear and manageable guidelines for health professionals that will enable the provision of good psychosocial support. There are certain underlying principles that are essential to good psychosocial support and form the foundation upon which all other aspects of palliative care are built. It is essential that this foundation is established from the beginning of involvement with a family.

These foundations enable the creation of an environment in which families can feel safe enough to address the strong, and often unfamiliar, emotions and issues that they are likely to experience during palliative care.

The essential aspects of this foundation are:

- Respect for, and understanding of, the uniqueness of each family.
- Empowerment.
- Excellent communication; including developing a relationship that is open, honest and fosters trust.
- Community perspectives.
- Emotional safety.

The strain for a family of caring for one (or more) of their children in the palliative phase of an illness is certain to challenge a family's coping capacity enormously.

The diagnosis of a life-threatening or life-limiting illness and the transition to palliative care causes fundamental changes and challenges to family life. It disrupts family routines, increases the demands on practical, financial, emotional and spiritual resources, and impacts on everyone in the family – as well as extended family and friendship or collegial networks.

It is essential that health professionals openly acknowledge this and that appropriate support is provided in all of these areas. All members of the team will have an effect upon the psychosocial well-being of the family. Consequently, the best psychosocial support is facilitated if all team members are equipped to work with families in ways that intentionally provide support and enhance the family's well-being and autonomy.

RESPECT FOR THE UNIQUENESS OF EACH FAMILY

Every individual and family has unique life experiences, perceptions, strengths and challenges. Failure to respect this makes it impossible to provide the best possible individualised care.

In exploring the uniqueness of a family it is useful to have an understanding of some key issues. These issues fall into five main areas:

1. **Social** (e.g. the degree of social dislocation they have experienced, the responsiveness of their support network, their financial situation).
2. **Family** (e.g. the make-up of their family, their communication patterns, roles and relationships, stress management style, problem solving skills).
3. **Individual** (e.g. personality, developmental stage, past experiences of illness and grief, level of exhaustion, personal methods/means of managing experiences).
4. **Disease** (e.g. duration, physiological impact, disease course, disfigurement, distressing symptoms).
5. **Grief history** (e.g. exposure to death, dying, trauma and/or previous losses and coping strategies).

The death of a child is beyond a parent's control, but how the care is provided is something they can control. Parents know their child the best. It is important to always respect this principle.

Health professionals do not 'own' the children and families that they work with. They are a resource for families to use in finding their place in the very broad spectrum of adaptive coping and grieving.

An empowering culture is one in which the family is enabled to have maximum control over the resources, information, decision making, and relationships that affect them. Such a culture respects the reality that each family has their own resources and information, and is capable of making decisions, and of maintaining and contributing to relationships (including their relationships with palliative care providers). Within this culture, families are trusted to manage their own lives. They are not perceived as passive recipients of professional services but as competent and leading partners in their child's palliative care.

It is well documented that families facing a terminal illness feel an enormous loss of power and control in their lives. The psychosocial value of assisting them to regain or increase their sense of power and control whenever possible cannot be over emphasised.

Empowerment can be facilitated by the health care team listening to the family's needs, offering encouragement and advice, being advocates for them and their child, and by ensuring they have access to skilled health professionals and practical assistance, including home care equipment.

Essentially, communication is the exchange of meaningful information from one person to the other. However, communication is a complex and multi-leveled process. The physical environment, the availability and attentiveness of participants, their emotional responsiveness and cultural biases are all part of communication, as are numerous other verbal and non-verbal components.

The goals of communication in palliative care are to:

- Establish a positive working relationship between the family and health professionals involved.
- Develop an accurate understanding of the family and of the messages they are attempting to communicate.
- Enable the family to understand the information communicated by the health professionals.
- Understand the communication styles and processes that occur naturally within the family, and use this to facilitate manageable communication for the child and family and all involved.

Listening

Interventions with families often commence with professionals telling families what can be done for them, or what their opinion is about the situation. It is always better to start by listening carefully to families. This involves recognition of both verbal and non-verbal cues.

Health professionals should listen to the family's perception of a situation, especially their views on the problems, needs, solutions and resources inherent in it. It is important not only to listen to the obvious content of what families are saying, but also to the emotional and other less obvious

content – including assessing what is not said overtly. By doing this, health professionals join the family on their journey, rather than telling them where they should go. The family feels respected and empowered, and concerns, skills and resources are identified from their own perspective. From this foundation, health professionals are well positioned to offer their expertise to the family in the most needed and useful ways.

Good listening is a dynamic process requiring active participation from the listener. Health professionals must demonstrate that they are listening to and valuing what families are communicating.

For example, in the context of discussing palliative care issues with families it is not rare for a parent to say *"I can't cope with this"*. Often the response given is a superficial reassurance that while the situation is very hard, *"You will cope"*, or *"Everyone feels that way"*. Such statements may be accurate but their immediate effect can block the opportunity to listen to:

- how the individual is really feeling
- what their perceptions are
- what other influences are affecting their perception of the situation.

Additionally such comments do not communicate an acceptance of what the individual is feeling or a sense of partnership.

A better response would seek clearer understanding of what the parent was actually meaning and would invite further information from them. This could be achieved through giving careful attention to the parent by using open questions and reflective feedback such as *"As I understand it, what you were saying is...."*

This is more likely to lead to an understanding of what it really means for this person to say *"I can't cope with this"*.

In this example it may have been assumed that the parent was expressing fears about caring for the child at home when they were actually expressing a more acute physical stress reaction at that very moment, such as the need to get some fresh air or to be physically sick.

Alternatively the parent may have helped care for a dying grandmother in their own childhood and have stressful memories which need processing.

Giving information

The giving of open and honest information is of paramount importance in palliative care. This requires intentional effort because in dealing with issues of strong emotional pain there is a cultural propensity (based in the desire to protect people from pain) to communicate in ways which minimise or avoid the issues.

Take for example a situation where there is the possibility that a child may experience seizures. If this risk is not mentioned to the family, or if superficial statements are made about seizures being controllable and the child experiences a seizure which is difficult to control, the family will have been ill prepared for the situation, and the culture of trust may also have been damaged.

In giving information it is important to be aware of both sides of the communication equation. Choosing a mutually acceptable time and place which is without distractions is important when holding a discussion with a family. When information is being given to the family, health professionals need to check the family understands the information. This can be as simple as asking a family (in non-patronising

language) what they understand has been said. Just asking if they understand is not enough because it can lead to a simple "yes" answer, which doesn't give the opportunity to listen to what the family actually understood. This checking process will help to identify misconceptions, and enables further clarification. Extra care needs to be taken with families from culturally and linguistically diverse (CALD) backgrounds. For example, some languages do not even have a word for palliative care. Other considerations need to include the literacy and cognitive skills of the caregivers involved.

It is through open and honest communication about all issues, especially the most difficult issues, that the health professional can assist families to prepare for the challenges of palliative care and establish and maintain trusting relationships with families.

Communicating with children



For many health professionals, being involved with children receiving palliative care is as difficult an issue to deal with as death and grief itself. Among other reasons, this is often because of their uncertainty about the correct thing to say or do with children.

While the fact the children require palliative care and that their families face bereavement is undeniably sad, competent and caring adults can be of enormous assistance to these children.

The biggest issue raised by most people regarding children in palliative care is “*What do we tell the children?*” This question in its own right reveals one of the problems; **Adults are more inclined to tell children things rather than listen to them!**

The starting point with children is to listen to them. By listening we:

- Show that we respect and regard them as important in their own and unique right.
- Gain an understanding of their world (i.e. their understandings and perceptions about their lives and the life of their family).
- Build the kind of trusting relationship that children need to have with an adult in order to get support from them.

Children’s understanding of death

Trying to divide children’s understandings of death into clear developmental stages is difficult. However, there is general agreement that a child, who is cognitively intact, from the age of seven years onwards will have a reasonably full understanding of death. They understand that it is universal, irreversible, has a number of causes and relates to non-functionality.²⁷

Life experience (such as life threatening illness, the death of a grandparent or a pet, or other exposure to death or information about death) has a significant influence upon children’s understanding, and can lead individual children to have a greater or lesser understanding of death than their peers. Children known to hospital teams, often will have had a greater level of contact with illness, may be aware of the death

of other children and therefore can have a greater understanding and awareness of these issues. Generalisations about children’s understanding of death are never as important as individualised knowledge about a specific child.

Professionals, who work in the area of communication with children about grief, death and palliative care, indicate that many young children (in our experience, certainly as young as five years) have well-developed understandings of death. This understanding will not be an “adult” understanding; for example the issue of permanence may not be fully developed. However, within their understanding, these children are able to process issues associated with death to a greater degree than many adults give them credit.²⁸

Effect of illness on children

Children with serious illness and their families experience major disruptions to all areas of family life. Consequently these children will have lost, to a greater or lesser degree, the relatively safe and predictable routine of the life that they had before diagnosis.

Sick children will have had experiences of illness, painful and distressing treatments, geographical relocation, loss or reduction of contact with key family members, peers and friends. They may also have experienced disability and loss of function or changes in their personal appearance.

Siblings will also have experienced loss of parental availability because of the parent’s physical absence while at treatment centres, their involvement in increased care for the sick sibling, and because of the strong likelihood that the parents will have been to some degree emotionally unavailable.

They will have experienced multiple disruptions to their lives, significant changes in their relationship to their sibling, loss of routine, concern for their sibling, and displacement from their usual role and position in their family. They will also have witnessed treatments and the physical effects of treatment and disease. To a greater or lesser degree all of these experiences are inevitable.

For some families there is the additional burden of more than one child being affected with the same condition e.g. neuromuscular and inherited metabolic diseases. In these circumstances the often younger and less debilitated child witnesses the worsening condition of their sibling and possibly death while considering their own illness and future. Consequently the family centred model of care ensures consideration of all within the family network.

On a positive note, many siblings and extended family will experience a greater bond or connection with the sick child and other family members as a consequence of this experience. Some children will exhibit extraordinary tolerance, resilience, and wisdom. Most bereaved children will not need counselling or intervention in the long term, however it should always be considered if learning or development is not progressing as expected.

Children's perceptions of illness

Children strive to make sense of their world, to gain a sense of mastery over it and to understand how they fit into it. They gather information to do this from multiple sources, of which direct verbal communication is only one small part.

There can be no doubt that from their experiences and observations of both subtle and unsubtle clues, children in palliative care situations will recognise that something serious is happening in

their family (e.g. parents crying after a meeting but being told 'nothing is wrong'). As a result of that recognition, they will have anxieties or concerns and will attempt to make sense of the situation and to work out where they fit into it. Often their imagination creates more fear and anxiety than the actual facts delivered in a compassionate, truthful and developmentally appropriate manner.

Useful strategies for parents and practitioners might include using a range of activities such as reading, drawing or writing, to help families communicate with their child/ children. This is equally important for the ill child and any siblings or close friends/relatives.

Adults instinctively want to protect children from the distress of any situation and often convince themselves that children do not understand what is going on, or can be protected by giving them as little information as possible or by keeping it secret. This is understandable, especially in the light of the stress that the children's parents are under.

Children often engage in a similar strategy of protecting their parents. They do this by not talking about topics which they recognise as being stressful for their parents. This mutual "protection" can lead to children and parents having similar concerns yet being isolated from one another's support.

Not assisting children to develop an accurate understanding of what is going on and where they fit into the situation leaves them ill informed, unprepared, and at the mercy of their limited life experience, imagination and fears. Two prime examples of this are the common conclusions that many children reach, i.e. that they are in some sense to blame for what is going on, or that they are no longer loved by the family.

Importantly these feelings of exclusion from the family arise at the very time when children most need to feel the security of inclusion and trust.

Not communicating with children leaves them with no trusted source of information about what is happening. As health professionals it is important to create opportunities to assist families with these challenging situations whilst also respecting the parents' own expectations of communication with their child. This can be challenging when a parent does not wish to inform their child of the situation at all and does not wish to have a child involved in their own decision making or planning for end-of-life care.

Ideally, children need a sense of safety, nurturing relationships with their caregivers, assistance to make sense of their world and their feelings and a sense of self-worth.

What should we communicate?

It is important to realise that it is impossible **not** to communicate to children. Every contact with them communicates something and every exclusion from contact communicates something. In the light of this, appropriate communication with children starts with the establishment of a relationship in which the child is included, respected and actively listened to.

From that basis, ongoing communication should take place in a supportive atmosphere in which children are invited to explore and express their feelings, their needs, and their understanding of what is happening. Children should be invited to ask questions and in response to their feelings, perceptions and questions, they should be given honest information, in language that is accurate and appropriate to their age.

Some very practical strategies to guide communication with children are:

- Answer the questions they have but do not overwhelm them with extra details.
- Observe their play and behaviour and other non-verbal signs for cues regarding what may be issues for them and explore these.
- Be willing to use media that they are comfortable with (e.g. books, art, toys, and storytelling).
- Give information gradually rather than giving it in one large session. This is especially important with progressive disease. It is better to keep the child informed of gradual developments than to give them information late in the process when the situation is more serious.
- Keep language as simple as possible. Language needs to be accurate and compassionate.
- Give the information repeatedly. Children generally need repeated explanations.
- Involve children in decision making as much as possible.
- Beware of issues such as blame and guilt.
- Reassure children that the situation is not their fault.
- Do not be afraid to show feelings to children. It can be an important model for them.
- Do not take over a parent's role. Parents are generally the most important people in their children's lives and a safe open relationship with them is invaluable for the long term wellbeing of the child. The role of the health professional is to help parents talk with their children and spend quality time with them. In doing this, the relationship between parent and child is reinforced and not undermined.

When children ask their own questions, explore with them the meaning of the question. Often as health professionals and adults we assume the expected answer. For many children their 'real' question is not what is asked. For example, if a child asks "Am I going to die?" they may not want a yes/no answer but to understand what will happen to Mum or Dad, a sibling or the pets.

It is important to identify a trusted person to be part of the conversations with children. This may be a parent but may also be a play therapist or social worker.

This style of communication gives children the clear message that they will be kept informed of what is going on, and it reassures them that they are a valued part of the family and that they will continue to be cared for. With this support children can manage the major emotional challenges of palliative care and grief.

It is important to meet with parents and discuss the types of conversations that may arise with children. As a professional you will need to negotiate with the parent whether or not you will:

- Always tell the truth.
- Always answer the child's questions honestly (this is especially important when parents do not want the child fully informed of the current situation).
- Tell the parent(s) news before the child.
- Be available when needed to help them talk with their child/children.

Communicating with adolescents

Adolescence is a time of many changing emotional needs, where peer relationships are very significant and where the relationship with parents is changing as the young person moves between dependence and independence. The adolescent with palliative care needs may be dealing with feelings of frustration, of loss of physical independence, and isolation from normal social interaction with peers. They may be more perceptive about the impact of their illness on their family than younger children. It is important to establish patterns of communication that recognise their unique needs, in particular, respect for their privacy and an honest open discussion regarding decisions about their care. At times a young person may wish to hear information independent of their parents, or have significant non family members (such as boyfriend/girlfriend) included in discussions. These arrangements will need to be discussed with both the young person and parents.

Adolescents' understanding of death, its meaning and what comes after may also be different from that of their parents. Adolescents and young adults living with a life-threatening illness want to be able to choose and record the kind of medical treatment they want and do not want, how they would like to be cared for, information for their family and friends to know, and how their life is remembered.²⁹

They are likely to want to contribute to discussions about their pain and symptom management, resuscitation planning, organ donation and funeral planning. It is important to provide opportunity for them to express their views, their hopes and fears. Creative ways of communication such as art, music, poetry, writing and peer support groups may all be helpful.

The individuals with whom we work do not exist in isolation from their families, and families with whom we work do not exist in isolation from their communities. People are social beings and have most of their needs met through their families and communities.

In light of this it should be remembered that families also do not make decisions in isolation, and the health professional should identify the 'significant' people who are involved in the decision-making process.

These people may need to be part of the discussion when difficult decisions are being made e.g. religious/spiritual/cultural leaders or extended family members. Communities include formal organisations such as schools, churches, charitable organisations, hospitals and other health services, employers and government agencies (such as Centrelink). They also include informal support networks such as family, friends, neighbours and hobby or sporting groups.

These community relationships can play an important part in supporting a family in the palliative phase of their child's illness. Consequently, it is important to understand the role they play for an individual family, and that they *could* play if they were appropriately resourced and linked to the family. For some families it may be helpful to have a 'family/friendship network meeting' that can provide an opportunity to inform the network of the current situation and how they can best assist, especially if a family is struggling to articulate their needs and wishes. This can be organised by the social worker or other key worker. Facilitating this active support is clearly an important role for health professionals.

A major consideration in using these networks is the issue of empowerment for the family.

Families experience major challenges to their sense of power and control when a child is dying. Consequently, every caution should be taken to ensure that families do not experience additional loss of power and control. Before engaging in work with any part of a family's social network, it is essential to consult with, and gain permission from the family. This respects their right to be in control of information about themselves, and protects their relationship with those social supports and their role in the decision making process. Ideally families' direct involvement with their communities should be maximised. The health professional's role should be facilitation of the process rather than representing the family.

SPIRITUAL, RELIGIOUS AND CULTURAL ISSUES

There is a wide spectrum of belief systems and cultures within our community. Death, dying, grieving and funerals often highlight religious, spiritual and cultural differences, and may give rise to new spiritual awareness as people search for meaning in response to life changing events. Culturally sensitive palliative care requires the health professional to develop an understanding of a family's cultural needs and a willingness to ensure these needs are met. These needs and experiences may be very individual and fall outside formal cultural or religious expressions. They may also be unique for each member of the family, and in particular, the spiritual experience of children may be quite different to that of adults. It is important to provide opportunities for everyone to explore and express their spiritual and cultural needs and experiences. Resources such as books, videos, poems and prayers may help (including those found on the Internet) but many of these conversations will be spontaneous.

An example of the importance of being aware of religious and cultural needs would be when health professionals encourage families to take time in planning and holding the funeral after a child has died. In some cultures, such as the Muslim community, the body has to be buried within 24 hours. In this situation, encouraging the family to “take their time” would demonstrate a lack of understanding of the family needs.

Effective communication is the key to gaining a better understanding of religious and cultural needs. This communication can be either directly with the family or with representatives from the same cultural or religious group. Being able to use a professional interpreter service has benefits such as translation of complex medical information and protection of privacy. In all cases, the communication principles discussed in this guide should be followed. In some texts related to death and dying there are chapters that give specific information about the various religious and cultural rituals required when someone is dying. This information may also be useful in some cases.^{3, 30-33}

There may be cultural barriers to providing the best care to a patient and their family. This can be related to communication styles, language, a lack of specific support/resources or other concerns or fears that the family may have. Discussing these worries respectfully and with humility where possible is important.

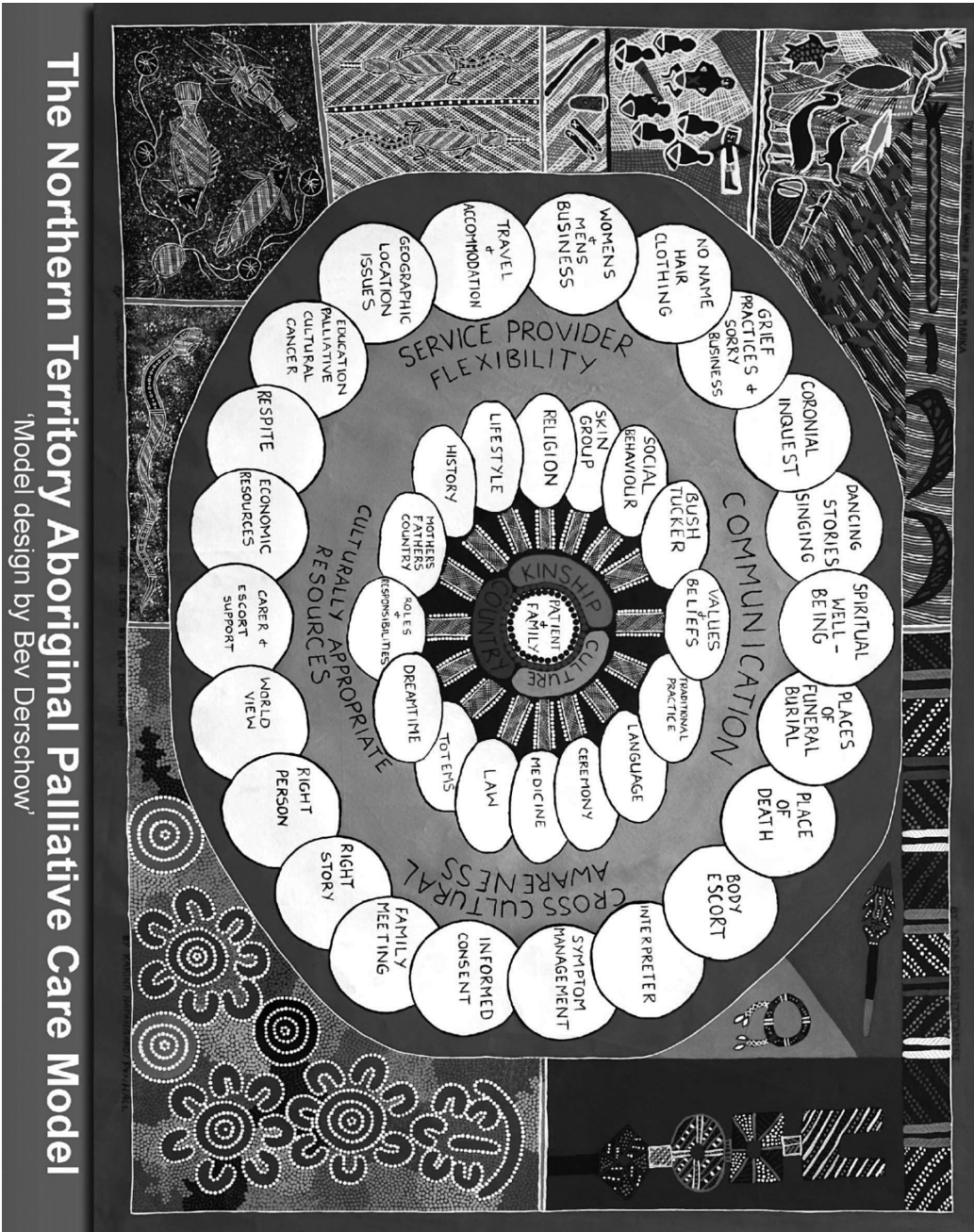
ABORIGINAL AND TORRES STRAIT ISLANDER CONSIDERATIONS

Cultural sensitivity and respect is imperative when caring for indigenous and Torres Strait Islander patients and their extended family and kinship structures. Cultural beliefs may affect how a family think about illness and pain, attitudes to medications and nutrition, their understanding of dying and death, customs and rituals surrounding death, spirituality, burial, cremation and bereavement. This may also be relevant for any specific culture.

Other things to consider:

- Avoid the use of the word death and dying. It may be more appropriate to use terms such as “sad news”, “sorry business”, or “very serious illness”.
- Care will need to be taken with use of pictures and photos post death.
- Care will also need to be taken with saying the name of the deceased child amongst the community.
- Specific tribal (or mob) ritual and networks will need to be respected.
- For some communities the role of the ‘Kurdaitcha’ man may be significant.

The use of an Aboriginal Liaison Officer (within the hospital) and/or Aboriginal Health Worker (who may be based in the local community), or an identified tribal elder may be of great benefit in the communication and coordination of care. Various jurisdictions have helpful resources such as models of care (Northern Territory) and guidelines (Queensland) which can be of benefit in increasing health practitioner’s awareness of Aboriginal cultural and spiritual values.^{117, 118}



Used with permission. Courtesy of Territory Palliative Care, Department of Health, Northern Territory. "This model was developed in conjunction with local indigenous artists to illustrate the palliative care system in a holistic approach."

REFUGEES AND ASYLUM SEEKERS

Some patients from a culturally and linguistically diverse (CALD) background may be refugees and/or asylum seekers.

A refugee is defined by the 1951 United Nations Convention as a person who:

“owing to a well-founded fear of being persecuted for reasons of race, religion, nationality, membership of a particular social group or political opinion is outside of the country of his nationality and is unable or owing to such fear is unwilling to avail himself of the protection of that country.”³⁴

Caring for the child of a refugee family who has a life-limiting illness requires an additional level of cultural sensitivity and understanding of the associated physical and emotional trauma that families may present with.

Refugees often arrive in Australia following many years in refugee camps or detention. Often, they have had little access to healthcare, sometimes resulting in negative health outcomes. Refugees may feel extremely isolated having lost their extended family, housing, income and position in society, employment, social support systems, cultural norms, religious customs and language. These issues may lead to a disconnect with cultural and religious supports. However families often seek comfort in specific rituals in the terminal phase.

Associated risk factors for refugees may include psychological trauma following the separation, loss or death of family and friends, leading to increased risk of mental illness, post-traumatic stress disorder (PTSD), anxiety and depression. Refugee parents and siblings of a child with a life limiting illness are at particular risk of complicated grief.

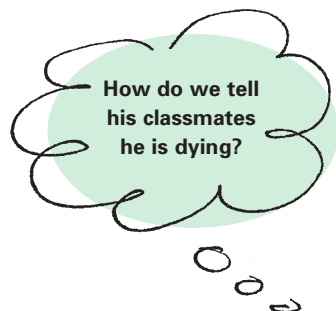
Culturally sensitive psycho-social care should be introduced early in the child's palliative care trajectory, to ensure families are linked to appropriate mental health care and ongoing bereavement care following the death of a child.

Refugees and Asylum Seekers usually lack knowledge of the Australian health care system.³⁵ They can also have unrealistic expectations of the health system based on their previous experience. There may also be issues of trust with people in authority, including health professionals, as they may have been involved in the administration of torture in their country. Consideration should also be given to literacy skills and the accuracy of using interpreters (with potential associated social and linguistic complexities).

Caring for refugee families in a holistic way involves acknowledgement of cultural, linguistic and health related issues specific to each family. Collaborating with the family's refugee support workers, with the family's permission, can be beneficial when plans are being made on how best to manage a child's progressive illness.

For further information contact the Refugee Health Network in your state.³⁶⁻⁴¹

SCHOOLS



Apart from the family, preschool, kindergarten, special schools and regular schools, are generally the most significant part of a child's community. Schools are extremely significant learning environments, not just in academic terms but also in terms of life skills. More importantly in the context of palliative care, schools are nurturers of children's social and emotional well-being, and are places where many extremely important peer relationships occur. They are also the places where the most significant extra-familial child-adult relationships tend to occur. For many families, the relationships associated with school are also a major part of the parents' social network.

Interventions in this setting generally take at least three forms:

1. **Renegotiating:** assisting the school and family to renegotiate the nature of their relationship. In particular to recognise the need for adjustment of the nature of, and time spent, at school and for the increased significance of emotional care and support.
2. **Care of the child and siblings:** assisting the school to develop skills to care for the child and siblings in the palliative phase of an illness, and to support the siblings in their bereavement after a child has died.
3. **Care of staff and students:** assisting the school to develop skills to care for all the staff and students through the grief and anticipatory grief associated with the palliative care and death of an important member of its community.

Renegotiating the relationship with school

As with all areas of psychosocial support in palliative care, renegotiating the relationship between the family and the school occurs most effectively if it is done intentionally. Consequently, it is best if the school and the family have a shared understanding of what the family needs and what the school can offer.

For schools where a student has had a long-term illness, a well thought out plan is often already in place and the child's anticipated deterioration will be part of this plan. At this point however, many schools request a meeting which includes the child's family, relevant school staff and the palliative care service to ensure best care, as well as maximising the time the child can spend at school.

Renegotiation will vary from family to family and school to school. Some schools and families have wonderfully open and supportive relationships established long before a child commences palliative care. Others may have given the situation surprisingly little consideration and have minimal communication, coordination and intentional support in place.

Most schools are within the positive part of the spectrum and those that aren't are generally willing to make the necessary changes once they become aware of a family's need. The health professional's role may entail little more than affirming the great job being done by the school

and the family. However, assistance to identify the family's needs and encouragement to enable the school to respond to them may be required. This may require advocacy with or on behalf of the family at times.

In looking at these issues with a family, it is worth reviewing how the school is contributing to the family's quality of life and psychosocial support by considering at least the following:

- Maintenance of important relationships (e.g. friendships with the dying child where appropriate).
- Maintenance of normality and routine for the child and siblings as much as possible.
- Specific support regarding particular needs.

The relationships with the child's close friends may continue at home once the child is no longer able to attend school, with guidance and support from school staff as well as negotiation with the family.

Examples of specific things that schools can do include:

- Enable sick children to attend school even if it is only for their favourite classes or for lunch breaks.
- Ensure that students keep contact with sick children who can no longer attend school through home visits, letters, videos, the internet (e.g. Skype® calls, Facebook® and e-mail) etc.
- Have a clear plan for getting messages to siblings quickly so they feel confident that they can be contacted when needed.
- Keep any school or artwork completed by the child and make this available to the family.
- Incorporate memory making into daily school activities.
- Invite siblings into the child's class for activities.

For children with longer term palliation needs (who are cognitively able) participation in classroom activities via the Internet may support learning and maintenance of classroom peer relationships.

Many families will be able to directly liaise with the school, to set goals and to problem solve around the issues. At other times it may be appropriate for the health professional to be involved in these processes.

Care of the child and siblings

School staff have high levels of skill in communicating with children; however they often find it intimidating to address the issues surrounding palliative care. The role of health professionals is:

- To affirm the skills that the school has and its importance to the family.
- To assist the school in communicating with the wider school community.
- To assist school staff to understand palliative care issues and grief processes, including how to use this knowledge in their interactions with the child and siblings.
- To work with school social worker, Chaplain or guidance officer to enhance sibling support.

The principles provided in the sections **“Communication”** and **“Grief and Anticipatory Grief”** form the primary content of information that schools need in order to achieve this.

Care of staff and students

How a school manages the palliative care and death of one of its students will be an important model for the student population regarding death and loss throughout their lives.

School staff who are aware of this generally take the responsibility of making the experience as healthy as possible seriously.

Schools have the multiple responsibility of providing appropriate care to the child who is dying and their siblings, and to the other students and staff at the school. This can amount to 1000 or more people. It is a daunting task and is a scope of care in which the school can have a great deal of experience; however, most schools greatly appreciate the support and guidance provided by experienced paediatric palliative care professionals.

It is important to acknowledge this expertise and to recognise that the health professional's role is to provide specialised input that can be used in planning care for staff and students. Much of the input is the same or similar to that provided for the care of the child and siblings.

Two of the most important issues are:

1. Planning for how the school will communicate to staff, students and parents.
2. Identifying people who are likely to require increased care.

For example, in deciding how to communicate to students, the school must decide what information would be best communicated to the whole school on assembly and to the greater school community, and what should be communicated in the smaller setting of the class room or even on an individual basis.

Generally, information which may be distressing should be communicated on as personal a scale as possible. This allows a more individualised response to the students. In the classroom, a teacher is more likely to observe which students are upset compared to in an assembly, and it is easier to give children the opportunity to ask questions and express feelings.

On a class level there are many things that teachers can do to facilitate communication of feelings and to enable children to understand what has occurred. For example, in the class of the child who has died, issues such as what to do with the child's desk can be discussed by the class and decisions made which meet the needs of the students. Similarly students can be given opportunities to write farewell letters or tributes, to have discussions, and to do drawings or other art work to express thoughts and feelings.

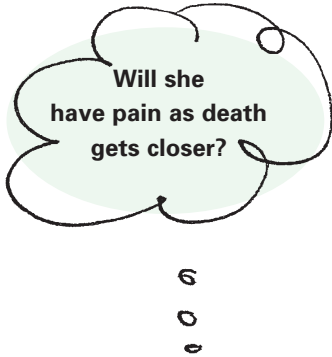
On the whole-school level, decisions such as whether or not to have a memorial service need to be made. Most schools find that memorial services and involvement in the funeral (if appropriate and acceptable to the family) are helpful events for the school community. The school could consider long term memorials for the child (e.g. a tree planting, bench seat with associated plaque and dedications). They provide a focus for a shared acknowledgment of the school's grief and of the importance of the life and death of the child to the school community. The school should always consult with the family of the child who has died about the wish for the timing and the nature of the service.

In a school there will be a spectrum of impact regarding the illness and death of a student. Some students will be greatly affected. Others will be minimally affected. With this recognition, it is important for schools to identify staff and students who may be at risk of stronger reactions to the situation, and therefore may require special care or at least a closer level of caring observation and support. Their needs should also be taken into account when planning how communication takes place in the school.

People (staff or students): who may be at increased risk are:

- Those who have already experienced significant loss in their lives (for example the death of a loved one, divorce, trauma, or refugee status).
- Those who have a close relationship with the child who has died or the siblings.
- Those who have similar health problems themselves or in their family.

It is not possible to predict who will ultimately struggle most with a death. Consequently teachers must be aware of the students they have responsibility for and allow opportunities for them to express their feelings and concerns so that difficulties can be identified and responded to.⁴² Staff should always be responsive to signs of distress in their colleagues as well.



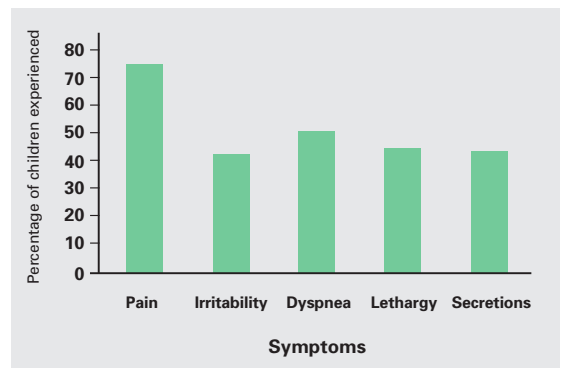
To achieve optimal quality of life, an understanding of the underlying disease process and anticipated symptoms is required, as well as an understanding of the family dynamics and likely needs of the child and family. How we approach the care of a dying child and the family will greatly influence the quality of the child's remaining life and the ability of the parents, siblings and friends to cope after the child's death.

Symptom Management is the care given to improve the quality of life of patients who have a serious or life-threatening illness. The goal of symptom management is to prevent or treat as early as possible the symptoms of an illness, side effects caused by treatment of an illness, and psychological, social, and spiritual problems related to an illness or its treatment.⁴³

The approach must be individualised, taking into account an often complex set of circumstances unique to each child's clinical situation. Hunt et al.⁴⁴ analysed the most common symptoms experienced by children dying from numerous conditions within a children's hospice. More than 80% of patients had pain recorded as a symptom in the last month of life. Over one third of children also experienced constipation⁴⁴.

A more recent study found lethargy and drowsiness were the most common symptom, and were more commonly experienced on the hospital ward compared to the intensive care unit. Other symptoms dying children experience include dyspnoea, nausea and vomiting, excess airway secretions, dysphagia, anorexia, agitation and irritability, psychological distress, skin changes, seizures and peripheral oedema.^{44, 45} Figure 6 shows the presence of the five most common symptoms based on a review of 170 child deaths in Queensland.

FIGURE 6 Symptoms experienced by dying Queensland children (n=170)



Pain is a commonly experienced symptom in children with life-limiting illnesses. Opioids are the cornerstone of pain management in this context, particularly during end-of-life care. Many children require opioid analgesia throughout the palliative phase of their illness.³ One single institution found that of 105 children who died in the hospital over a 12 month period, 90% received analgesics in the 72 hours before death.⁴⁶ Of 44 patients with Cystic Fibrosis at another institution, 38 patients (86%) received opioids for severe dyspnoea and pain at the end of life.⁴⁷ The duration of treatment, in this study, varied from one month to one hour before death.

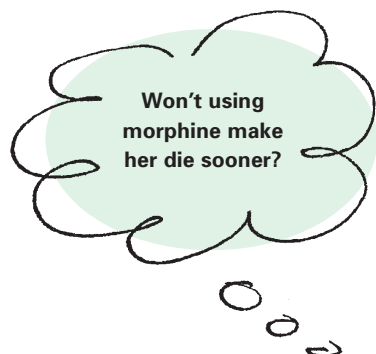
SYMPTOM MANAGEMENT

The fear of uncontrolled pain is also recognised as a source of anxiety for both children and their families.⁴⁸ Anxiety and other psychosocial factors will contribute to the total pain that is experienced by the child and family and this must be acknowledged and managed. The approach to such management of the child and family involves knowledge of the underlying disease, an understanding of the prior treatment or management, and of the child and family's responses to treatment and their transition from a curative to palliative focus.

This section is aimed at providing an overview of the more common symptoms experienced by children with life-limiting illnesses, and a guide to the possible approaches to management.

Symptom management can be broadly divided into five stages:

1. Obtaining of an accurate history and assessment
2. Identification of the cause (if possible)
3. Ongoing communication with the child and family
 - Explanation of symptoms and treatment options
 - Establishment of goals of therapy
4. Implementation of therapy
 - Treatment of underlying cause
 - Pharmacological, physical, psychological and complementary
5. Regular review and modification of treatment/management.



Dispelling the myths

Myth: *Young children and infants, experience less pain than adults.*

Truth: Similar myths suggest children tolerate pain better and there is rarely a requirement for opioid analgesia. Many clinical studies have now challenged these “beliefs” and have demonstrated that neonates, infants and children experience similar degrees of pain to adults.

Myth: *The child will become addicted to opioids.*

Truth: One of the major concerns of both family and health professionals is that of addiction to opioids. It is important to dispel this fear early in the treatment course. Addiction is predominantly a psychological dependence and patients with a life-threatening or life-limiting illness (such as cancer or severe cerebral palsy) who require titrated doses of morphine do not become addicted to opioids.³² Tolerance to the analgesic effect of morphine is not a problem and psychological dependence in palliative patients is extremely rare. Parents should be reassured in this regard.

Myth: *Only the medical team can control the child's pain medications.*

Truth: If pain is poorly controlled or escalating, patients will ask for further doses of morphine to bring about relief from pain. The opioid doses can safely be increased by parents at home after discussion with the clinical team. This parental ownership of pain control is important. Adolescent patients also benefit when given some control and choice with how their pain is managed.

Myth: *It is not appropriate to use opioids with non-cancer conditions.*

Truth: Many patients with non-cancer life limiting conditions benefit from a trial of carefully titrated opioid analgesia. However, patients with a non-cancer diagnosis often do not require the rapid dose escalation that a child with a rapidly progressive solid tumour requires.

Myth: *Sedation will affect the quality of the child's life.*

Truth: Sedation from opioids will usually improve within a few days of commencement. This is because the child will develop tolerance to the central nervous system (CNS) depressant effects of morphine over time.

Myth: *Using morphine will result in respiratory depression.*

Truth: Respiratory depression can be avoided by gradual and steady increases in the dose. Like the CNS depressant effects, children will develop tolerance to the respiratory depressant effects of morphine. The use of morphine in adult patients with advanced chronic obstructive pulmonary disease (COPD) was found to relieve dyspnoea and improve sleep. It did not worsen respiratory failure.⁴⁹

Myth: *Using morphine will shorten the child's life.*

Truth: Pain control does not shorten a child's life.

Rather it improves the child's quality of life and brings comfort to a child's death. It can even extend a child's life because they are not exhausted from fighting pain. The dosage can also be reduced or increased depending upon how the child and their disease responds to treatment. A study of adult patients found the use of opioids and sedatives did not have an association with shortened survival.⁵⁰

Assessment of pain

The evaluation of pain in the child is significantly different from that in the adult, and is dependent upon the age, developmental stage, cognition and previous life experiences. As the child's vocabulary and past life experiences are often limited, it is difficult to obtain qualitative and quantitative descriptions of their pain. The use of a number of different parameters is often helpful in determining position, nature and severity of pain. Simple observation of the child's level of activity and behaviour is useful.

Any change in behaviour, such as irritability, fractiousness or withdrawal may indicate discomfort. The FLACC scale allows observation of such parameters and has been validated as a means of measuring pain in infants and children younger than four years of age.⁵¹ Assessment in older children can be enhanced with the use of visual analogue tools, including the Faces pain scale-revised (Figure 7).⁵² These scales share a common metric (generally 0–10). Pain scores fall into three ranges:

- mild (0–3)
- moderate (4–6)
- severe (7 or more).

The Non Communicating Children's Pain Checklist can be used for children with cognitive impairment, and the CRIES pain rating scale can be used for neonates.^{53, 54}

The use of a body outline completed with the child and parent may also aid in determining the position of the pain and intensity can be highlighted by different shades of colours. Importantly, remember *"pain is where the hurt is"*.

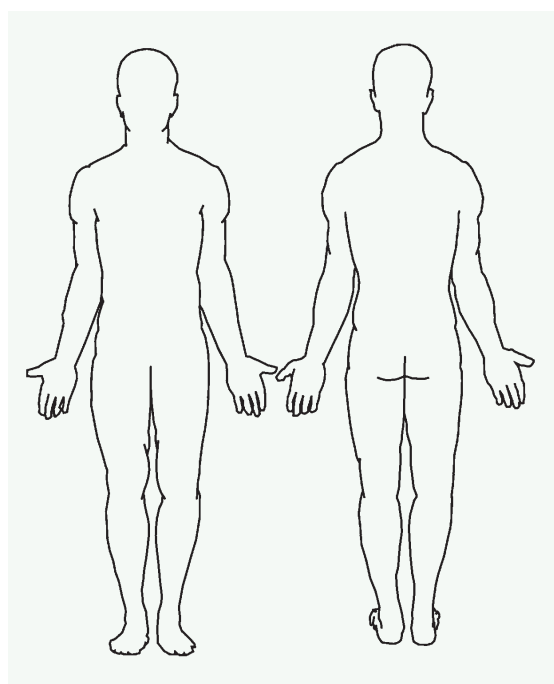
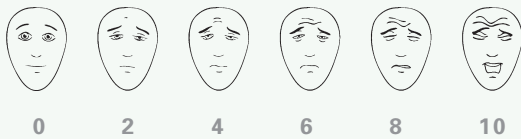


FIGURE 7 Faces Pain Scale

Faces Pain Scale – Revised (FPS-R)

The full-size version of the Faces Pain Scale (FPS-R), together with instructions for administration, are freely available for non-commercial clinical and research use from www.painsourcebook.ca.



Note: Numbers are not shown to the child

Instructions to the child are: These faces show how much something can hurt. This face (point to the left-most face) shows no pain or hurt. The faces show more and more pain (point to each from left to right) up to this one (point to the right-most face) – it shows very much pain. Point to the face that shows how much you hurt right now.

Do not use words like ‘happy’ or ‘sad’. This scale is intended to measure how children feel inside, not how their face looks. Numbers are not shown to children; they are only shown here for reference. The instructions for administration are currently available in 12 languages from www.painsourcebook.ca.

FIGURE 8 Visual Analogue Scales



No pain

Worst pain imaginable

Aetiology of pain

Invasion of bone and bone marrow is the most common cause of pain in the child with cancer and is typical of somatic nociceptive pain.⁵⁵ Other causes of pain in the child with cancer are shown in Table 3.

In children with non-cancer diagnoses, pain is often multi-factorial. Musculoskeletal causes are common in this group of patients (e.g. scoliosis, dislocated hips, osteoporosis, and fractures). Pain due to dystonia/muscle spasm or gastrointestinal dysmotility is also common. Pain may also be associated with orthotic devices and can increase around the time of orthopaedic and other surgeries. Differentiating between cerebral irritability and pain can be challenging in the child with developmental disability.

TABLE 3 Aetiology of cancer pain

Tumour involvement

- Direct tissue/nerve damage
- Bone marrow infiltration
- Infiltration of tissues
- Compression of tissues
- Nerve compression
- Raised intracranial pressure

Treatment related

- Infection due to immunosuppression
- Mucositis related to chemotherapy and radiotherapy
- Inflammation after surgery

Procedure related

- Venepuncture
- Surgical interventions
- Investigations (e.g. lumbar puncture, bone marrow aspirate)

Pain can be classified by its origins and pathway of transmission to the brain into two broad categories, nociceptive (somatic and visceral) and neuropathic.⁵⁵ Details of these categories and the pathways are beyond the scope of this guide, but different mechanisms and pain characteristics occur.

Principles of pain management

The aims of pain management are to relieve pain at rest and during activity, and to ensure comfort during sleep with minimal side effects. To achieve this, the same general principles of symptom management are applied.

The World Health Organisation (WHO) published a guide, "Cancer Pain Relief and Palliative Care in Children" in 1998. Drug therapy is the mainstay of treatment and given the correct drug, dose and interval, pain relief is possible for most patients.⁵⁶ More recently, the WHO has published guidelines on the pharmacological treatment of persisting pain in children with medical illnesses.⁵⁷

The general strategy for treatment is to keep it simple and to administer the drug(s):

- **By the mouth** – the oral route is convenient, non-invasive and cost effective.
- **By the clock** – regular scheduling ensures a steady blood level, reducing the peaks and troughs associated with "as required" or "pro re nata" (prn) dosing.
- **By the ladder** – enabling a stepwise approach to treatment commencing at an appropriate symptom level with non-opioid analgesia and progressing to opioids (Figure 9).

Treatment should be individualised according to the child's pain, response to treatment, and frequently reassessed and modified as required.

A multimodal approach to pain management has recently been advocated.⁵⁸ This approach advocates the early introduction of adjuvant therapies when appropriate.

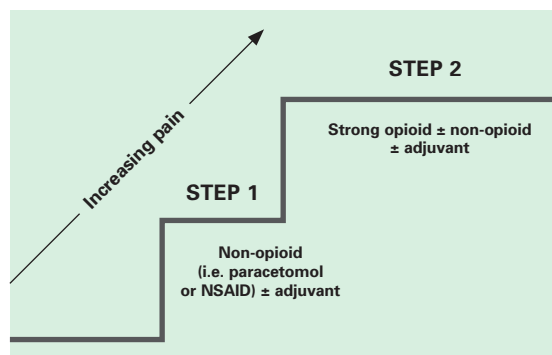
ANALGESIC AGENTS

The analgesics can be broadly classified into two groups:

1. **Primary analgesics.**
 - Non-opioid and non-steroidal anti-inflammatory drugs.
 - Weak opioids.
 - Strong opioids.
2. **Secondary analgesics/Adjuvant drugs.**
 - Antidepressants.
 - Anticonvulsants.
 - Corticosteroids.

Which drug to prescribe is dependent upon the nature and severity of pain. A step-wise approach to drug administration is recommended as shown with the WHO ladder commencing with simple non-opioid drugs and progressing to opioids at appropriate doses to control increasing pain (Figure 9).

FIGURE 9 WHO 2 Step analgesic ladder



PRIMARY ANALGESICS

Non-opioids

The non-steroidal anti-inflammatory drugs (NSAID) are weak primary analgesic agents whose main action is to suppress inflammation by their anti-prostaglandin activity. Examples include naproxen, ibuprofen and diclofenac. They are effective at reducing fever and bone pain. They have significant side effects, however, including gastric irritation and ulceration. They also interfere with platelet function and should be used cautiously in children who are thrombocytopenic, as risk of bleeding may be increased. Caution should also be used when prescribing these medications to children who are dehydrated, have renal impairment or gastric irritation. Aspirin is not prescribed to children because of the association with Reye's Syndrome.

Sensory input of pain can also be reduced by the administration of regular paracetamol. This drug has a mild anti-inflammatory effect and is very useful for musculo-skeletal pain. It is also an effective antipyretic. Paracetamol is generally well tolerated by children and is available in oral (tablet/capsule/ syrup – various concentrations available), rectal and parenteral formulations, and is the preferred non-opioid analgesic for children (15mg/kg/dose q4h orally, maximum 4g/day). There are some families that are already familiar with preparations combining paracetamol with codeine e.g. Painstop®. Careful explanation regarding the use of regular paracetamol with combined agents is required to ensure recommended daily limits are adhered to. It is preferred that the combined agents are avoided. It may be preferable to use low dose morphine syrup and regular paracetamol instead.

Paracetamol used in conjunction with an opioid is a simple example of the multimodal approach.

Care should be taken when prescribing paracetamol for extended periods of time. Hepatic toxicity has been reported in children following chronic therapeutic dosing.

Oral opioids

The opioids are the mainstay of treatment for the majority of patients with severe pain associated with cancer and other life-limiting illnesses. If pain is not controlled with paracetamol, a weak opioid such as codeine phosphate (0.5mg/kg/dose q4h) could be commenced. However, the WHO guidelines now suggest low dose morphine is preferable in children, as codeine has a ceiling analgesic effect, causes significant constipation, and a small percentage of the population does not metabolise codeine adequately.

Morphine

Morphine remains the standard against which other opioid analgesics are compared. Morphine is available in oral (mixture/ syrup – in various concentrations, tablets, and immediate and slow release formulations), parenteral, spinal and rectal preparations. The oral route is the preferred route of administration as it is readily absorbed and tolerated by most children. Liquid / syrup morphine, in the appropriate dose, provides 4–6 hours of pain relief and it should be prescribed as a regular dose every four hours.

There is no role for *prn* dosing in the palliative patient. End of dose breakthrough pain is distressing and more difficult to control as the plasma drug level falls.

Severe pain that is not adequately controlled with the commencing dose of morphine orally (0.2mg/kg/dose q4h) is an indication to increase the dose. Incremental increases of 30–50% per dose may be required within 24 hours. Once the appropriate 24 hour dose of morphine is determined, transfer to sustained release morphine preparations is possible.

The slow release preparations, MS Contin® (available in tablets and dispersible sachets) and Kapanol® (capsule), have a slower onset of action than immediate release morphine, but have a longer duration of action.

The dose is determined by calculating the total daily amount of morphine (six x regular dose). For example 5mg of morphine mixture every four hours is a total daily dose of 30mg, which is converted to 15mg of MS Contin® twice daily. Immediate release morphine should be available for “breakthrough pain” which may occur. The “breakthrough” dose is equivalent to one-sixth of the total daily dose. Using the same example, this would equal 5mg (see Table 4). The breakthrough dose of morphine should be increased enough to alleviate any breakthrough pain. Breakthrough pain is covered in more detail in the following pages.

If repeated doses of breakthrough morphine are required, this is an indication to increase the total daily dose of sustained release morphine to the equivalent total dose of morphine required in the preceding 24 hours. For example if two doses of 5mg were required it would be appropriate to

increase the dose of MS Contin® to 20mg bd. With an increase in MS Contin® the breakthrough dose of morphine will also need adjustment. For 40mg/day the appropriate breakthrough dose of immediate release morphine is approximately 6.5mg. If greater than six breakthrough doses of opioids are required in a 24 hour period, it is an indication to increase the background opioid dose by 50%.

Oxycodone

Oxycodone is a useful alternative to morphine and is available as tablets, capsules and syrup. It has an increased potency compared to morphine in children (morphine: oxycodone = 1.5:1). It may also have kappa-receptor agonist activity, which relieves neuropathic pain. Its oral bioavailability is higher than morphine (50–60%). The oral dose is 0.1–0.2mg every 4 hours. It is available as a tablet (scored), capsules and syrup (concentration 1mg/ml only). Onset of action is 20–30 minutes making it a good option for breakthrough pain. It is a good alternative to morphine especially when morphine contributes to vomiting, pruritus or delirium. Slow release preparations are available in tablet form only and cannot be crushed.

Oxycodone-with-naloxone controlled-release (CR) tablets (Targin®) are also available and provide equivalent analgesia to that of oxycodone CR tablets of the same oxycodone dose. The addition of the naloxone component reduces, but does not eliminate, constipation. (There is no literature available on the use of Targin® in children and it

TABLE 4 Example of daily morphine equivalent dosing and conversion from immediate release morphine to sustained release morphine, for a 15kg child receiving an initial dose of ~ 0.3mg/kg.

<i>q4h dose</i>	<i>Total 24h dose</i>	<i>MS Contin dose</i>	<i>Breakthrough dose</i>
5mg	30mg	15mg q12h	5mg

can initially provoke withdrawal symptoms or diarrhoea in children who are opioid tolerant).

Tramadol

Although not a pure opioid, tramadol can be effective for moderate to severe pain in older children. It is a weak mu-receptor agonist and a serotonin and norepinephrine reuptake inhibitor. The dose is 1mg/kg every 4 hours. While not a first line agent, it can have a role in certain situations. Care should be taken to avoid drug interactions especially with selective serotonin-reuptake inhibitors and tricyclic antidepressants. It can also lower the seizure threshold.

Hydromorphone

Hydromorphone is a derivative of morphine with a higher potency. The parenteral formulation is up to five times as potent as the oral formulation. It is available as a syrup (concentration 1mg/ml and in tablet form). There is also a slow release preparation available in tablet form (Jurnista®) which is taken once daily.

Buprenorphine

Buprenorphine is a synthetic lipophilic opioid with 30–50 times the analgesic potency of morphine. This molecule has been available for parenteral and sublingual administration for more than 25 years. Currently its use in paediatrics is expanding due to the introduction of a transdermal matrix patch. Sublingual tablets (200 micrograms) have a duration of action of 6–8 hours. The lowest dose patch (5 microgram/hour) has the benefit of having a much lower morphine equivalent dose than the lowest dose fentanyl patch (12 micrograms/hour) and is applied every seven days. The patches are an easily administered option for low dose

background opioid analgesia in a stable situation e.g. in severe neurological impairment.⁵⁹

In clinical practice, buprenorphine does not have a ceiling effect. A ceiling effect has however, been demonstrated for respiratory depression resulting in an improved safety profile compared to other opioids, such as fentanyl. Its pharmacokinetic properties, such as faecal elimination, allow its use in patients with renal impairment. It has both opioid agonist and antagonist properties and may precipitate withdrawal symptoms in children dependent on other opioids.^{60, 61}

In Table 5, the available preparations and dose for oral morphine, oxycodone and hydromorphone are presented.

Methadone

Methadone is a synthetic, lipid soluble opioid and has a chemical structure very different to morphine. It acts on the μ -opioid receptor and antagonises the N-methyl D-aspartate (NMDA) receptor. Consequently, it is very effective at treating somatic, visceral and neuropathic pain. Methadone has a long half-life with large inter-individual variability in its half-life (3.8 to 62 hours). It can accumulate in the plasma causing severe somnolence which is slow to reverse. Patients need to be observed closely when commencing methadone or switching from another opioid.

Methadone also prolongs the QT interval through its effect on cardiac potassium channels. This may predispose patients to arrhythmias, particularly when the parenteral formulation is used. Consequently, it is often reserved for patients receiving palliative care where other methods of pain management have failed. The goals of care, including the risk of arrhythmia, need to be discussed with the family.

PAIN

TABLE 5 Oral Morphine, Oxycodone and Hydromorphone Preparations

Drug	Release	Form	Dose
Morphine	Immediate Q4 hr	Liquid	1, 2, 5 and 10mg per ml (Ordine®) (40mg/ml on request)
	Immediate Q4 hr	Tablet	10 and 20mg (Sevredol®)
	Controlled Q12 hr	Suspension (granules dispersed in water)	20, 30, 60, 100 and 200mg (MS Contin® Sachets)
	Controlled Q12 hr	Tablets	5, 10, 30, 60, 100 and 200mg (MS Contin®)
	Controlled Q12–24 hr	Capsule	10, 20, 50 and 100mg (Kapanol®)
Oxycodone	Immediate	Liquid	1mg/ml (Oxynorm®)
	Immediate	Tablets	5, 10mg (Endone®)
	Immediate	Capsule	5, 10, 20mg (Oxynorm®)
	Controlled	Tablets	5, 10, 20, 40, 80mg (Oxycontin®) (Targin® – Oxycodone-with-naloxone)
Hydromorphone	Immediate	Liquid	1mg/ml (Dilaudid ®)
	Immediate	Tablets	2, 4, 8mg (Dilaudid ®)
	Controlled Q24 hr	Tablets	4, 8, 16, 32, 64mg (Jurnistor ®)

Methadone is available in tablet and liquid formulation and has very good bioavailability (almost 100%). The dose should be doubled when converting from parenteral to oral methadone. The starting dose for an opioid naïve patient is similar to that of morphine (0.1mg/kg every 6–8 hours).⁶² This frequency of dose can be continued until the patient becomes pain free or drowsy. At this point the dosing frequency should be reduced to two to three times daily.

Calculating the dose of methadone in patients already established on another opioid is more complex. This is because cross-tolerance with methadone will be low. The starting dose is usually one tenth of the morphine equivalent dose. Because it is infrequently used in paediatric pain management and it has complex pharmacokinetics, methadone should only be prescribed by experienced practitioners. Its use needs further evaluation in a clinical research setting.

TABLE 6 Parenteral and Transdermal Opioid Preparations

Drug	Parenteral Preparations	Transdermal Preparations
Morphine sulphate	5mg/ml, 10mg/ml, 15mg/ml, 30mg/ml	
Morphine tartrate	120mg/1.5ml, 400mg/5ml	
Hydromorphone	2mg/ml, 10mg /ml, 50mg/5ml, 500mg/50ml	
Methadone	10mg/ml	
Fentanyl	100mcg/2ml, 500 mcg/10ml	12 mcg/hr, 25mcg/hr, 50mcg/hr, 75mcg/hr, 100 mcg/hr.
Buprenorphine	300mcg/ml	5mcg/hour, 10mcg/hour, 20mcg/hour

Parenteral opioids

If the child is unable to take drugs orally, alternative routes of drug delivery are available. MS Contin tablets can be given rectally and morphine suppositories can be obtained. In the acute situation, a subcutaneous injection of morphine (0.1–0.2mg/kg) is easily administered and has a rapid onset of action.

There is no role for intramuscular pethidine (or other drugs) in the management of pain related to progressive illness.

If the oral route remains problematic, a subcutaneous infusion of morphine is a simple and effective mode of drug delivery. Some children will still have a central venous access device (central line or port-a-cath) and morphine can then be delivered intravenously. Insertion of a central venous access device for pain management in certain situations is appropriate.

The 24 hour dose of parenteral morphine is equivalent to one third (1/3) of the total oral dose. For example, a child receiving 150mg of oral

morphine per day would require 50mg of morphine delivered as a continuous subcutaneous infusion per 24 hours.

Fentanyl, hydromorphone and methadone can also be administered parenterally (see Table 6).

Fentanyl

Fentanyl is a lipophilic synthetic opioid and is approximately 10–40 times more potent than morphine. It is less potent in neonates and smaller children. Like morphine, it has an affinity for the mu-opioid receptor and acts as a pure agonist. It is available in a transdermal formulation. Clinical studies in children have shown that transdermal fentanyl is an effective alternative to oral opioids with studies suggesting fewer side effects (particularly constipation and nausea) and improved quality of life.^{63–65} Its topical route of absorption removes the need for oral or parenteral administration. Recently developed matrix patches can be cut, allowing smaller doses to be administered.

Fentanyl is well absorbed via the oral mucosal route providing prompt pain relief. Oral transmucosal fentanyl citrate (OTFC) lozenges and buccal tablets offer promise in providing non-invasive means of treating breakthrough pain. The use of intra-nasal fentanyl (dose 1–2 mcg/kg using mucosal atomiser devices) for breakthrough pain relief has been described in emergency departments, cancer pain relief, and in neonates and infants.⁶⁶⁻⁶⁸

Refer to Appendix 2 for guidelines on commencing a subcutaneous infusion.

BREAKTHROUGH PAIN

When exacerbations of medium to severe pain occur on the background of otherwise controlled pain, it is commonly described as breakthrough pain. There is a high incidence of breakthrough pain in paediatric oncology patients with one study finding at least one episode of such pain per day in 57% of patients.⁶⁹ This pain can last from seconds to minutes and is frequently described as “sharp” or “shooting”. The pain may be incidental (related to movement, breathing, coughing or eating) or spontaneous with no obvious cause.

Management of breakthrough pain

Breakthrough pain usually requires the administration of rescue doses in addition to medications used to control baseline pain as outlined above. If the child experiences breakthrough pain, morphine can be administered as a bolus. This is usually one sixth (1/6) of the total daily dose; the equivalent of 8mg for a 50mg 24hr dose). This can be administered through the sidearm of the subcutaneous cannula.

The following pharmacological approaches can help in the management of breakthrough pain:

- Increasing the quantity of long-acting opioid.
- Increasing the frequency of a short-acting opioid.
- Replacing a short-acting opioid with a long-acting opioid.
- Adding a rapid-onset opioid to around-the-clock medication.⁷⁰

Non-pharmacological management strategies may also be appropriate and may be combined with the use of medications. This may include heat, ice and some of the complementary therapies outlined below.

Paediatric pain crisis

A pain crisis in a child is an emergency and requires treatment beyond conventional means. If a child has persistent and severe pain (and with close medical supervision and instructions) the dosing frequency of oral opioid can be reduced to 1–2 hours. At the same time a specific diagnosis of the underlying cause should be attempted to allow more effective therapies to be implemented.

The intravenous route of administration provides rapid onset of analgesia. On occasion, the clinician may need to remain by the child’s bedside to titrate incremental intravenous doses every 10–15 minutes until effective analgesia has been achieved. The analgesic effects of opioids increase in a log-linear function, with incremental opioid dosing required until either analgesia is achieved or somnolence occurs. The total amount of opioid administered to require this reduction in pain intensity is considered the opioid loading dose. A continuous infusion of opioid may need to be commenced to maintain this level of analgesia. An alternative to a continuous infusion of opioid is intermittent parenteral opioid, especially in the setting of an unpredictable pain syndrome.

Patient Controlled Analgesia (PCA)

A PCA is an infusion device which can be activated by the patient to self-administer a set bolus dose of analgesia. Children as young as seven years of age are able to use a PCA.⁷¹ Nurse controlled analgesia (NCA) is appropriate for the control of pain in infants, pre-verbal children and children with severe developmental delay who cannot use a PCA. Some hospitals allow parent controlled analgesia in younger children. Parent controlled analgesia can also be provided in the home. Two modes can be used, bolus only or bolus plus a continuous background continuous infusion.

Additional doses of pain relief can be administered for breakthrough and incident pain in addition to a

background infusion. The prompt and safe delivery of analgesia associated with this modality allows control over the wide and unexpected analgesic requirements of children. There is good evidence to support the use of PCAs in patients with mucositis undergoing bone marrow transplantation.^{72, 73} Adolescents have reduced sedation and morphine intake with similar pain relief when using PCAs compared to infusions.⁷² Both modes (PCA and NCA) are suited to provide analgesia where there is a large component of "incident pain" e.g. pain on movement or with coughing.⁶⁹ There is also evidence to support this modality of pain management in the palliative care context.⁷⁴

TABLE 7 Patient Controlled Analgesia and Nurse Controlled Analgesia

Mode	Concentration	Continuous infusion	Patient bolus	Lock out interval	Hourly dose limit
Morphine PCA	0.5–1mg/kg in 50mls (max 50mg)	0–1ml/hr	1ml	5 min	150mcg/kg/hr
Morphine NCA		0–0.5ml/hr	1ml	10–30 min	120mcg/kg/hr
Morphine Infusion		0–2.5ml/hr	N/A	N/A	N/A
Fentanyl PCA	25–50mcg/kg in 50mls (max 2.5mgs)	0–1ml/hr	0.5ml	5 min	5mcg/kg/hr
Fentanyl NCA		0–0.5ml/hr	0.5ml	10–30 min	3mcg//kg/hr
Hydromorphone PCA	150mcg/kg in 50mls (max 7mg)	0–1ml/hr	1ml	5 min	20mcg/kg/hr
Hydromorphone NCA		0–0.5ml/hr	1ml	10–30 min	15mcg//kg/hr
Adding ketamine to morphine OR	Add 1mg/kg ketamine to the 50ml opioid syringe. Use parameters as given above. (Ketamine may be added to the opioid-containing syringe)				
Run a separate ketamine infusion	Add 5mg/kg ketamine to 50mls. Run at 1ml/hr to deliver 100mcg/kg/hr				

The PCA device must be programmed to deliver an opioid dose at a predetermined frequency, with a maximum total dose allowed per hour. Table 7 outlines guidelines for the initial prescription of a PCA in an opioid naïve patient. Rescue doses are kept as a proportion of the baseline opioid infusion rate and re-calculated as between 50% and 200% of the hourly basal infusion rate. Bolus attempts and successes should be documented as part of the observations.

A PCA (or NCA) can be used to safely titrate analgesia during a pain crisis. It is possible to transition a patient back onto long acting opioid preparations after the child's pain has stabilised and the total daily opioid requirement has become clear. A PCA can be continued at home using a computerised ambulatory delivery device (CADD) pump. Parent controlled analgesia is also possible in younger children in the home setting.⁷⁵ Methadone can be administered in a PCA if the lockout interval is increased to 30–60 minutes given its long half-life.

SIDE EFFECTS AND PRECAUTIONS OF OPIOIDS

All preparations of opioids have side effects. Constipation can be a major problem and consideration should always be given to prescribing laxatives whenever opioids are used. Unlike many of the other side effects, in particular nausea and drowsiness, tolerance to constipation does not occur. Methylnaltrexone (sub-cutaneous) is a competitive antagonist for the μ -receptor. It works at the level of the gastro-intestinal tract and does not cross the blood brain barrier. Its use has been described in case studies and a dose of 0.15mg/kg administered subcutaneously has been recommended.^{76, 77} It has a role when children

cannot tolerate or respond to enteral or rectal laxatives.

Nausea and vomiting can occur upon initiation of treatment with an opioid and an antiemetic will be required. After administration of breakthrough doses of morphine it is not unusual for drowsiness to occur, but once a stable dose of morphine is achieved this becomes less problematic and children are likely to be more active and alert with good analgesia. Pruritus is not uncommon and can be relieved with the concomitant use of an antihistamine. Low dose naloxone infusions have also been used to prevent itch associated with opioid infusions (dose 0.5microgram/kg/hr).⁷⁸

Opioids will cause respiratory depression only if given in an inappropriate dose, which is usually above that required for analgesia. This is particularly the case for opioid naïve patients. Extreme caution should be used in giving naloxone at a treatment dose to patients who have been receiving chronic opioid therapy, since severe pain and symptoms of opioid withdrawal may ensue.

Patients with significant renal impairment will accumulate metabolites of morphine. In this context, drowsiness and respiratory depression can occur after 24 hours. Careful supervision of dosing is required and a lower initial dose should be prescribed and titrated according to response. Similarly, a lower initial dose should be used for children with liver failure as bioavailability of morphine is increased.

Opioid rotation

It is possible to improve pain management by changing to a different opioid medication. This process is called opioid rotation.

The usual indications for switching to an alternative opioid are:

- Excessive side effects (e.g. itch, nausea, delirium) with adequate analgesia (70%).
- Side effects with inadequate analgesia (17%).
- Tolerance (7%).⁷⁹

Alternatives to morphine include fentanyl, hydromorphone and sometimes methadone. A switch from one opioid to another is often accompanied by change in the balance between analgesia and side-effects.

Table 8 assists with conversion of morphine to fentanyl and hydromorphone. Both background and bolus doses should be taken into account when switching opioids. Switching to methadone is more complex as discussed above.

Opioid resistant pain

While opioids are the gold standard for treatment of cancer pain, not all pain is opioid sensitive (see Table 9).

Relative resistance can be overcome by increasing the dose, improving support of the child and family, or using an alternative route of drug delivery. The addition of a non-steroidal anti-inflammatory drug or paracetamol may alleviate pain related to soft tissue or bony metastases. Pelvic pain is also potentially difficult to control and consideration of nerve blocks or palliative radiotherapy may be required.

Neuropathic pain tends to be relatively resistant to the above approaches and is due to the compression or infiltration of nerves, or to neuropathy, which may be disease or treatment related. For example, medications used in cancer (e.g. vincristine) and HIV therapy (e.g. didanosine, zalcitabine) can cause neuropathy. The nature of neuropathic pain has different characteristics and tends to be either spasmodic/shooting or continuous burning/piercing. Abnormal sensation, either increased with or without tingling, may co-exist. Younger children often cannot differentiate between or precisely describe the different qualities of pain.

TABLE 9 Causes of opioid resistant pain

Relative resistance
Under-dosing
Poor absorption orally
Lack of emotional support
Semi-resistant pain
Soft tissue/muscle infiltration
Bony metastases
Raised intracranial pressure
Neuropathic pain
Resistant pain
Neuropathic pain
Muscle pain

TABLE 8 Conversion of morphine to fentanyl and hydromorphone

Drug	Morphine	Fentanyl	Hydromorphone
Relative Potency	1	40 ¹¹⁹	5–7 ¹²⁰
Example Dose Comparisons	4mg	100 micrograms (i.e. 0.1mg)	570 micrograms

SECONDARY ANALGESICS

Antidepressants in low dose are useful for neuropathic pain, particularly painful paraesthesia and peripheral neuropathy. As well as having a direct analgesic effect, they potentiate opioid analgesia via adrenergic or serotonergic mechanisms. A low dose of amitriptyline (starting dose 0.2mg/kg) at night usually has an effect within 48–72 hours.

Anticonvulsants, such as gabapentin, pregabalin, carbamazepine or sodium valproate are useful for pain related to nerve infiltration/compression, which is often periodic or spasmodic. The anticonvulsants have a stabilising effect on excitable cell membranes and prevent the spread of neuronal excitation.³²

Clonidine and ketamine can be useful for patients with refractory neuropathic pain. They are, however, rarely required in children receiving palliative care.

Corticosteroids either alone or in combination with an anticonvulsant is effective to reduce swelling associated with nerve compression/infiltration. They can also alleviate some of the symptoms related to raised intracranial pressure. Low-dose steroids act as anti-inflammatory drugs and can reduce bone pain. Dosing should be restricted, however, as significant side effects occur, including excessive weight gain, gastric irritation and susceptibility to infection. Acid suppressing agents (e.g. omeprazole or ranitidine) are usually given concurrently with corticosteroids.

Other drugs recognised as secondary analgesics include: antispasmodics, anxiolytics and bisphosphonates. Hyoscine N-Butylbromide (Buscopan®) is useful for bladder or bowel spasm and low dose diazepam is effective treatment for muscle spasms and myoclonus. Regular review,

reassessment and an individualised approach are essential for successful treatment of pain.

OTHER THERAPIES FOR PAIN

Antibiotics or antifungal agents will improve pain control when there is an underlying infection such as cellulitis or mucositis. Consideration should be given to practical, oral anti-infective therapy in these situations. Bisphosphonates such as pamidronate and zoledronate inhibit bone reabsorption and are useful for the treatment of pain secondary to hypercalcaemia, disseminated bone metastases, avascular necrosis and pathological fractures related to osteoporosis.

Both chemotherapy and radiotherapy can be used as palliative treatment, and radiotherapy particularly, can have a potent analgesic effect in patients with cancer. One or two fractions are often all that are required, and the effect can be quite rapid. Consequently the opioid requirement is likely to lessen and drowsiness may occur.⁸⁰ During radiotherapy it is often useful to revert from sustained release preparations of morphine to immediate release morphine.

Pain from bone metastases can also be relieved with injectable radiopharmaceuticals. Samarium has an affinity for skeletal tissue and concentrates in areas of bone turnover.

Meta-iodobenzylguanidine (MIBG) is taken up by neuroblastoma cells. Treatment with these agents can allow the reduction of opioid use and can provide several weeks of pain relief.⁸¹ Eventually treatment with radiotherapy and chemotherapy will not be a viable option, as the child will continue to have progressive disease, and the journey from home to treatment centre will become too exhausting for child and family.

Nerve blocks are occasionally used for children with well-defined somatic or visceral pain.⁸² Spinal opioid therapy and epidural anaesthetics are very effective for pelvic pain and often allow the reduction of sedation doses of oral or subcutaneous opioids. Blocks can be temporary, prophylactic or permanent and should be placed by anaesthetists experienced in such pain management techniques.

Sedation as a therapeutic modality for intractable pain

The use of sedation is reserved for select situations of refractory pain where conventional therapies have been unsuccessful.⁸³ All acceptable means of providing timely analgesia without compromising consciousness should be exhausted. This trade-off between sedation and inadequate pain relief requires the consideration of the wishes of the child and his or her family. Sedation should be prescribed by an experienced practitioner with the primary aim of relieving suffering. Ethical principles including the principle of double effect are important in this context. The continuation of high-dose opioid infusions in these circumstances is recommended to avoid situations in which a patient may have unrelieved pain but inadequate clarity to report pain perception. A variety of drugs have been used in this setting, including barbiturates, benzodiazepines, and phenothiazines.⁸⁴

NON-PHARMACOLOGICAL THERAPIES FOR PAIN MANAGEMENT

A range of physical therapies such as warmth, cold, touch and electrical therapy are used in the management of various types and locations of

pain. Touch and massage can produce relaxation as well as stimulating afferent pathways. Transcutaneous electrical nerve stimulation (TENS) is useful in treating musculoskeletal and neuralgic pain. It acts by inducing electrical activity in larger afferent fibres thereby reducing the nociceptive pain signals in the dorsal horn of the spinal cord, inducing paraesthesia over the painful area. Involvement of a physiotherapist may be helpful with education for the family on how to use a TENS and in the provision of massage therapy.

Acupuncture is another modality which offers promise as a potential source of assistance for children with unrelenting pain, nausea, or other unpleasant symptoms due to disease or treatment.⁸⁵ Some families explore numerous other complementary therapies in order to help their child with pain and other symptoms. It is important to encourage open conversation regarding these therapies.

Fear and anxiety will aggravate pain, and communication with the child and family about symptoms and their treatment can assist in management. Simple measures of distraction, play, and music therapy, may be helpful. Touch and massage will facilitate relaxation in many children. Parents and siblings often like to perform the massage, and particularly for siblings, this enables them to be actively involved in their brother or sister's care. Older children and adolescents are also able to learn different relaxation techniques which may be as simple as listening to music. Cognitive-behavioural techniques of pain control such as breathing exercises, guided imagery and hypnosis are also important to consider.⁸⁶ These non-invasive measures allow control to be regained and will often aid in total pain relief for the child.

ORAL PROBLEMS

Mouth care

Mouth care at the end of life is essential, as children who are debilitated have poor oral intake and/or poor oral hygiene, and are susceptible to mouth problems. A reduction in a child's immune system as a result of prior radiotherapy, chemotherapy and progressive malignancy also increases their susceptibility to mouth problems.

Regular mouth care can prevent many oral problems. Cleaning teeth twice daily with a soft tooth brush or swab, and mouth washes with chlorhexidine or chlorhexidine gel are beneficial. Gentle irrigation of the mouth with warm salt water (0.9% saline solution) will help remove debris and soothes the mouth. Lips can be kept moist with lip balm or paraffin. The mouth can be kept fresh by sucking ice chips.

Xerostomia

Xerostomia or dry mouth is a common problem. This can result from mouth breathing, dehydration, anxiety, drugs and infection. Simple measures such as sucking ice cream, ice cubes, frozen juices and drinks will moisten the mouth and relieve thirst, while chewing or sucking unsweetened pineapple pieces increases salivary flow and can also help clean the mouth as it contains ananase, an enzyme which breaks down protein.

Mucositis

Mucositis or mouth ulcers are a well-known side effect of chemotherapy and after head and neck radiotherapy. Mucositis can also arise as a consequence of poor oral hygiene, neutropenia (low white blood cell levels) and infection.

Aphthous ulcers are small, shallow, painful ulcers that can be relieved with simple analgesic mouth wash, such as benzydamine hydrochloride (Difflam®). Lignocaine (Xylocaine®) viscous (local anaesthetic gel) may also be used.

Herpetic ulcers are painful, larger ulcers and can also cause significant oesophagitis. Oral acyclovir in addition to an analgesic mouthwash should be used. The pain of severe ulceration may require oral or parenteral morphine in combination with parenteral acyclovir to ensure absorption.

Children who are immunosuppressed are at risk of fungal infection. Candidiasis may present as stomatitis (inflammation of the mouth) when the obvious white plaques may not be evident. Oral nystatin (Nilstat®) (100,000 U/ml) 1–2mls, or amphotericin (Fungilin®) lozenges, or miconazole (Daktarin® gel) every 4–6h should be used. In children where there is clear localised candida infection, or where topical treatments have been ineffective, once daily oral fluconazole (Diflucan®) may be indicated.

Mucosal bleeding can be reduced by improving mouth care, and treating or preventing infection. Thrombocytopaenia (low platelet levels) will aggravate bleeding and a platelet transfusion should be considered. This will depend on the child's stage of disease. Oral tranexamic acid (Cyklokapron®) and topical sucralfate suspension (1g tablet dissolved in 10ml) may be beneficial.⁸⁷

GASTROINTESTINAL SYMPTOMS

NAUSEA AND VOMITING

There are many reasons why a child can become nauseated or vomit. Common causes in palliative care include:

- Opioids and other drugs.
- Upper gastro-intestinal inflammation.
- Raised intracranial pressure.
- Metabolic disturbances.
- Constipation.
- Infection.

TABLE 10 Choice of antiemetic therapy

Site	Aetiology	Antiemetic
CTZ	Drugs (opioids) Metabolic (hypercalcaemia)	Ondansetron/Zofran® (0.1–0.2mg/kg q8–12h; Max 8mg/Dose)
		Metoclopramide/Maxolon®* (0.15mg/kg q6–8h; Max 10mg/Dose)
		Prochlorperazine/Stemetil®* (0.1–0.2mg/kg q8h)
		Haloperidol/Serenace®* (0.01mg–0.1mg/kg q12–24h)
Vomiting centre	Direct stimulation (pain, fear) Viscera (gut obstruction) Raised intracranial pressure	Anti-anxiety medication
		Promethazine/Phenergan® (0.2–0.5mg/kg q8h)
		Hyoscine N-Butylbromide/Buscopan® (0.3–0.5mg/kg q6–8h; Max 20mg/Dose)
		Hyoscine hydrobromide (6–10 µg/kg q6h; Max 400 µg/Dose)
		Cyclizine/Valoid® (0.5–1mg/kg q8h; Max 50mg/Dose)
Gastric outlet	Opioids Stasis /compression	Metoclopramide – see above
		Domperidone/Motilium® (0.2–0.4mg/kg q4–8h; Max 10mg/Dose)
GI Inflammation	Gastritis (secondary to NSAIDs, steroids)	Mylanta 10–20mls qid
		Omeprazole/Losec® (0.5–1mg/kg q12–24h; Max 20mg/Dose)
		Ranitidine/Zantac® (2–4mg/kg q12h; Max 150mg/Dose)
		Sucralfate (dose depends on age)

*The phenothiazines, (Maxolon® and Stemetil®) can cause dystonic reactions more commonly in children compared to in adults. These reactions are usually easily recognised and families should be informed of such a possibility. Benztropine/Cogentin® (0.02mg/kg SC) rapidly reverses this effect and should be available.

Vomiting is initiated by the vomiting centre when it is stimulated directly or through the chemoreceptor trigger zone (CTZ), autonomic afferents from the viscera (gut organs) and higher brain centres. Antiemetic drugs have different effects upon these sites and the agent chosen should depend upon the most likely reason for the nausea or vomiting (Table 10). For example, drugs and metabolic disturbances act on the CTZ and antiemetics affecting this site are indicated while disturbances of gastric (stomach) emptying can be helped with agents that increase gastric emptying such as metoclopramide (Maxolon®) and domperidone (Motilium®).⁸⁷ It is important to note that children with a possible gut obstruction should avoid stimulant agents such as metoclopramide as they can aggravate the obstruction resulting in increased pain. Octreotide, administered intravenously or subcutaneously as a bolus or infusion, may relieve vomiting associated with bowel obstruction.⁸⁸

Other causes of vomiting include severe constipation which should be relieved with adequate treatment. Raised intracranial pressure which causes vomiting is usually, but not always, associated with headache. Steroids can alleviate these symptoms, but long term steroid use is associated with excessive weight gain with a change in appearance, behavioural changes, fragile skin and ultimately resistance. Drugs acting on the vomiting centre can be beneficial and used instead of steroids (e.g. cyclizine).

If possible, the preferred route for medication is by mouth. It may be necessary to give initial doses of medication by the intravenous, subcutaneous or, very occasionally by the rectal route. Many agents which can be administered subcutaneously are compatible with morphine and therefore, can be used in combination in infusion pumps.

Further details regarding drug compatibilities are outlined in Appendix 3.

Non-pharmacological management strategies to consider include:

- Chewing ginger pieces or taking ginger tablets.
- Sipping water, juice or flat soft drinks.
- Eat foods with a lot of water in them. Try clear soups, ice blocks and jelly.
- Offering bland foods or salty foods and snacks.
- Altering feed regimes if the child is receiving enteral nutrition (e.g. giving continuous instead of bolus feeds).
- Trial of hydrolyte or similar in replacement of milk based feeds.
- Aromatherapy.

CONSTIPATION

Normal bowel function ranges from three motions a day to one motion passed every three days, or up to two weeks in a breast feed infant.

Constipation refers to a significant variation from the normal bowel habit. It refers to difficulty, discomfort or delay in passing a bowel motion.

Constipation is an extremely common symptom and can contribute to abdominal pain, anorexia, nausea and vomiting as well as overflow diarrhoea. Factors known to make constipation more likely are outlined in Table 11.

The assessment of constipation is based upon:

- The underlying condition (including any neurological problem).
- Food and fluid intake.
- Medication.
- Previous laxative use.
- Previous and current pattern of bowel habit, including frequency and consistency of stool.

GASTROINTESTINAL SYMPTOMS

Chronic constipation is common in children with an underlying neurologic impairment, usually as a result of poor tone and reduced mobility.

Constipation should be expected in all children receiving opioid analgesia. Opioid receptors in the gut increase the tone and non-propulsive motility in the ileum and colon.⁸⁹ Laxatives should **always** be prescribed with opioid medication.

The effect of laxatives is dose related and there is large variation between individuals. If diarrhoea occurs, and overflow incontinence has been excluded, the laxative treatment should be modified according to the child's needs.

TABLE 11 Exacerbating factors for constipation

Exacerbating factors
Poor dietary intake
Poor fluid intake
Immobility
Medication <ul style="list-style-type: none">• Opioids• Anticholinergics• Antidepressants• Anticonvulsants• Antiemetics
Local factors <ul style="list-style-type: none">• Anal fissures/infection
Previous constipation
Electrolyte disturbance (e.g. hypercalcaemia, hypokalaemia)

Treatment and general measures

- Predict and prevent.
- Encourage fluid and fibre intake.
- Encourage movement.
- Stop or reduce unnecessary drugs .
- Laxative treatment.

The laxative used should be based upon patient preference of formulation and the degree of constipation (see Table 12). Parachoc® is well tolerated by children. Lactulose is often used as the laxative of first choice and can be mixed with juice, cordial or milk. However, the sweet taste can deter some children, and it can cause bloating and cramping – particularly for children with slowed bowel activity from medications such as opioids. Coloxyl®, as tablets, is favoured in older children and adolescents.

Treatment for opioid induced constipation should be directed at the large bowel to stimulate and soften the stool. For example the combination of coloxyl with senna promotes secretion of fluid thereby softening the stool through the coloxyl component while senna stimulates peristalsis. Care must be taken when using senna as it may cause excessive bowel spasm. Macrogol 3350 (e.g. Movicol ® sachets given up to three or four times a day) can also be very effective. An alternative to Movicol® is OsmoLax® (polyethyleneglycol laxative) which does not have the salty background taste of Movicol®.

If the stool volume or frequency remains inadequate, and constipation is well established, suppositories or a small enema will be required to clear the lower bowel before a normal pattern can be established. Enemas or suppositories are best avoided in neutropenic patients. DuroLax® or glycerin suppositories are generally effective.

Small volume enemas such as Microlax® are easy to use and usually not too distressing for the child. Larger volume enemas can lead to fluid and electrolyte disturbances particularly in the debilitated or dehydrated child. For refractory constipation an oil retention enema can be useful. Once the constipation is relieved, prophylaxis should continue.

GASTROINTESTINAL SYMPTOMS

During the terminal phase, significant discomfort related to constipation is uncommon. As fluid and oral intake are generally decreased, oral laxatives can be discontinued. A suppository or Microlax® enema may help reduce rectal discomfort arising from the urge to defecate or if there is overflow incontinence from faecal impaction.

TABLE 12 Action of laxatives

Lubricants and stool softeners
Parachoc® (Paraffin liquid) Coloxyl ® (Docusate Sodium) – softener and promotes secretion of fluid
Stimulants/contact laxatives – promote secretion of fluid into bowel to stimulate peristalsis
Senokot® (Senna), cascara Durolox® (Bisacodyl) – primary effect on colon within 24h
Osmotic – draws fluid into bowel to soften the stool & stimulates peristalsis
Duphalac® (Lactulose) Sorbitol Movicol ® (Macrogol 3350 with electrolytes) ClearLax ® (Macrogol 3350) OsmoLax ® (Macrogol 3350) Golytely® – effective within 6 hours but may cause significant fluid shift; should be used with caution
Suppositories and enemas – often a combination of softener and stimulant (rectal administration)
Glycerin suppository – softener Durolox® (Bisacodyl) – contact/stimulant

Non-pharmacological management strategies to consider include:

- Pear or prune juice.
- Liquorice.
- Adding fibre to feeds.
- Sugar free gum or lollies in generous amounts have a laxative effect.

DIARRHOEA

Diarrhoea is characterised by an increase in frequency and wateriness of the stool. The cause is usually evident from the history and the underlying condition. Simple measures such as discontinuing laxatives, high fibre foods and enteric supplements will often aid management. Consideration should be given to possibility of infectious causes of diarrhoea (e.g. rotavirus).

Medication is frequently required and loperamide (Imodium®) is generally well tolerated and effective. If severe watery/osmotic diarrhoea is suspected, such as can occur with severe graft versus host disease of the gut and in children with HIV infection, then octreotide can be helpful. Octreotide may also reduce diarrhoea refractory to other treatments. It can be administered intravenously or subcutaneously, and given as a bolus or infusion.¹⁰³

In the terminal stage, loperamide (a potent μ -receptor antagonist at 0.2mg/kg qid, increasing to 2mg/kg/day if required) will usually be sufficient. Loperamide capsules can be dispersed in water to make a 0.2–1mg/ml solution allowing for easier administration to children who prefer liquid formulations. Likewise, morphine either orally or subcutaneously can also alleviate diarrhoea.

GASTROINTESTINAL SYMPTOMS

Non-pharmacological management strategies may include provision of oral rehydration solutions (e.g. gastrolyte or hydrolyte) instead of feeds for 24–48 hours. Sometimes changing feed regimes from bolus to continuous feeds, or to an elemental feed may reduce the volume of diarrhoea. Collaboration with the paediatric team and dietician is often required in this context.

Extra care and attention is recommended with hygiene and skin care to the peri-anal area in the presence of ongoing diarrhoea. There is also an increased risk of electrolyte disturbance.

ANOREXIA AND CACHEXIA



Anorexia or loss of appetite is common in the later stage of the child's illness and can be associated with cachexia (loss of muscle and fat). There are often multiple causes in addition to the underlying disease including pain, nausea, constipation, drugs, anxiety, depression, oesophagitis and gastritis; addressing these is an important step in treatment.

A pre-emptive explanation of what can occur is essential and providing simple advice in regards to dietary habits such as offering the child small, simple meals can be very effective.

Medical devices for feeding

A large number of children, particularly those with non-cancer life-limiting diseases, receive part or all their nutrition by medical devices. Such devices include NG (nasogastric), NJ (nasojejunal), PEG (percutaneous enteral gastrostomy) tubes, and intravenous fluids including total parenteral nutrition (TPN). Providing food and fluids to a child through a medical device may be the only way a child can receive nutrition, or may be a comfort measure, but in some situations, particularly at the end of a child's life, can increase or prolong suffering.

In a child with a relatively prolonged palliative period who is having swallowing difficulties or difficulty maintaining their weight because of insufficient caloric intake, it may be considered beneficial.

Use of devices to provide food and fluids at the end of life should be considered on the basis of whether it will have more benefits than burdens for the child to allow the child's interests to be advanced.

FEEDING INTOLERANCE

A small group of children who receive nutrition through medical devices can develop a progressive intolerance often seen as worsening abdominal symptoms such as reflux, vomiting, abdominal bloating, irritability, and pain, which can be severe. If feed intolerance does not respond to the usual dietary modifications, (e.g. changing the formula type or route, decrease in feed volume, or usual anti-reflux medications) then visceral hyperalgesia should be considered.

Visceral hyperalgesia seems to result from an alteration in response to normal bowel sensations which are perceived as pain. Successful treatment often requires analgesic medication including strong opioids or agents such as tricyclic antidepressants (amitriptyline) or anticonvulsants (gabapentin).

FORGOING NUTRITION AND HYDRATION

It is ethically permissible to stop, or not start, medically provided food and fluids when the hope of recovery for the child is low or non-existent. However, families often find this thought distressing as it raises fears of their child suffering and “starving to death”.

Clinical experience would suggest this is not the case. Children with progressive disease can survive for significant periods of time with little oral intake and have minimal complaints during their last days or weeks of life. Comfort medications, such as morphine or benzodiazepines, are infrequently needed because of limited nutritional intake.

A reduced fluid intake can lessen distressing symptoms such as nausea, vomiting, cerebral oedema, excessive secretions and urinary incontinence while maintaining artificial hydration can aggravate such symptoms.^{31, 90}

The family require a careful and sensitive discussion on the reasons for considering limiting or withdrawing food and fluids, with the focus being the goal of improving distressing symptoms. It can be helpful to work with a dietician who has experience with paediatric palliative care to help the family understand the changing goals. If the family remain resistant to the idea then a three-day trial reduction in feed volume of 25 percent to 50 percent may provide objective evidence to gain their confidence and improve rapport.

Consideration should be given to cultural influences where provision of nutrition and food has significant meaning and where limiting nutritional intake can be a challenging concept to understand and adhere to.

Goals of care

In patients with non-cancer life-limiting illnesses, the occurrence of episodes of severe respiratory symptoms is often the point at which issues around resuscitation come sharply into focus. However, it is important to recognise that for many children, there may have been numerous previous episodes of severe respiratory symptoms from which the child has recovered well despite predictions otherwise from treating health care professionals. Severe respiratory symptoms are also relatively common in children dying from cancer.

Whether severe respiratory symptoms occur at home or in the hospital, a common question to be addressed is whether the patient is to be 'resuscitated'. These discussions will require the parents or care-givers to indicate whether the child is to be intubated, ventilated (via tube or via non-invasive ventilation – see below). There will also need to be discussion about location of care at these times and whether care in an Intensive Care Unit or retrieval to another hospital is to be considered.

Another aspect of these discussions relates to the risks and benefits of investigations, interventions and symptom management as part of the treatment plan for the respiratory symptoms. There is often discussion around the extent of monitoring and what is to be done if the child's vital observations fall outside normal parameters. Many parents will also wish to monitor their child's oxygen saturations and other vital signs at home (especially at night). Monitoring equipment is not routinely provided for this and the discussions in relation to these requests can be quite challenging. The key is to validate parental concerns and re-affirm their skills at detecting or observing deterioration in their child without the presence of technology.

DYSPNOEA

Dyspnoea (breathlessness or shortness of breath) is relatively common in children with life limiting illnesses. Dyspnoea may reflect deterioration or may be intermittent and/or reversible. At times dyspnoea may be so severe as to constitute a palliative care emergency. It frequently occurs in conjunction with other symptoms and is almost always associated with anxiety, for both child and family. Causes of breathlessness include respiratory and non-respiratory system related conditions. The types of conditions which affect the respiratory system include infection, inflammation, fluid accumulation and problems with the muscles for breathing or the structure of the chest wall (see Table 13).

Emergency management of severe dyspnoea

- Determine resuscitation status and underlying disease status.
- Correct easily reversible causes.
- Position patient sitting up if possible.
- Oxygen if tolerated.
- Suction secretions if indicated (and used with care).
- Morphine 0.05mg/kg parenterally, repeat in 15minutes (if given iv) or 30 minutes (if given subcutaneously) if dyspnoea not settling.
- Midazolam 0.05mg/kg parenterally or clonazepam drops (2.5mg/mL) 0.01mg/kg buccally for anxiety/distress – repeat if necessary.

RESPIRATORY SYMPTOMS

Specific treatment

Some causes are reversible with relatively simple measures. For example, a short course of antibiotics may be appropriate for the management of infection, and analgesia will alleviate dyspnoea related to pain. Packed cell transfusion may be considered in individual situations when anaemia may be contributing to dyspnoea.

A course of intravenous antibiotics may be appropriate for pneumonia but this will depend

upon a number of factors including parental wishes, prior performance status and disease trajectory, and ease of obtaining intravenous access. Parents will often opt for enteral antibiotics for pneumonia if they have adopted a more palliative approach for their child.

Pleural effusions in children with non-cancer conditions are less common than in the setting of a cancer diagnosis. Management should be discussed with a respiratory paediatrician.

TABLE 13 Causes of breathlessness

Respiratory	Non-Respiratory
Airway obstruction – lower or upper	Mediastinal disease
Chest wall deformity such as severe scoliosis	SVC obstruction
Rib fractures (with or without pneumothorax)	Cardiac disease/congenital heart disease
Respiratory muscle dysfunction including generalised muscle weakness (e.g. Spinal Muscular Atrophy) or phrenic nerve palsy	Metabolic causes including metabolic acidosis
Pleural effusion – inflammatory, cardiac, hypoalbuminaemia, malignant	Cerebral causes including raised intracranial pressure
Atelectasis and mucous plugging	Elevated diaphragm:
Asthma	- Ascites
Bronchiectasis	- Abdominal distension
Infection (viral pneumonitis/pneumonia)	Anaemia
Pneumothorax	Anxiety
Pulmonary oedema	Pain
Interstitial lung disease (idiopathic, chemotherapy, radiation)	
Pulmonary embolism	
Pulmonary hypertension	

In a child with a known cardiac condition, respiratory symptoms can sometimes be eased with titration of medications including diuretics. This should be done in consultation with the child's cardiologist.

Supportive measures

Simple measures are often helpful at reducing the sensation of breathlessness. The child and family should be managed in calm and reassuring manner, as anxiety will contribute to the degree of dyspnoea. Breathing exercises and relaxation techniques may be beneficial to the older child. Increasing air movement with a portable fan and improving ventilation in the room by opening windows are simple, often helpful steps. Positioning the child in a comfortable and upright position in bed or chair will also aid breathlessness. Breathlessness is often accompanied by tachypnoea and mouth breathing which can lead to a dry and painful mouth. Simple measures directed toward keeping the mouth and lips moist will help. Humidification of room air may also be beneficial.

Drug therapy

Bronchodilators

If bronchospasm is present, or if there is a history of asthma, a trial of bronchodilators is indicated. Salbutamol (Ventolin®) via a spacer or nebuliser is simple to deliver and will aid in reversible airways disease.

Corticosteroids

Corticosteroids (either inhaled or systemic) are also effective and can be used in addition to bronchodilators in children who have a component of bronchial hyper-reactivity.

Prolonged therapy with steroids is not generally indicated, as their effect is not sustained and the side effects can be significant.

Opioids

Opioids moderate the reflexive drive to breathe and decrease patient awareness of dyspnoea. They may also improve the efficiency of breathing and exercise endurance.³² Opioids, commenced at low dose and increased as required, may be given as required to children with intermittent dyspnoea or regularly for those with persistent breathlessness.

The best evidence is for morphine for the management of breathlessness. It is unclear whether other opioids are as effective as morphine and therefore morphine should be used as first line agent in this setting unless there is a compelling medical reason to the contrary. Oral/enteral, subcutaneous or nebulised morphine can be used. There is no proven advantage in administration of nebulised morphine over other routes of administration. However, some children do find this route of delivery beneficial.

The dose of morphine required for relief of dyspnoea is usually lower than that required for analgesia. In opioid naïve patients, a suggested starting dose would be 0.1mg/kg/dose orally/via PEG or 0.05mg/kg/dose subcutaneously or intravenously. Dosing intervals need to be determined on the basis of severity of dyspnoea, renal function and other clinical considerations. Morphine can be combined with midazolam (see below) in a subcutaneous infusion to relieve respiratory symptoms, particularly in the terminal phase.

Anxiolytics

The sensation of breathlessness can be very frightening and a small dose of oral diazepam (0.04–0.2mg/kg q8h) is often helpful to reduce associated anxiety. Optimising general symptom control, particularly pain, will also reduce the level of anxiety. Midazolam (0.1mg/kg intravenously or sub-cutaneously OR 0.3mg/kg buccally) decreases anxiety, agitation and distress in the child with dyspnoea.

Oxygen therapy

Many children with non-cancer diagnoses will have long-term respiratory issues where supplemental oxygen may be required either continuously or at times of respiratory illness. Many have chronically low oxygen saturations which they tolerate with no respiratory distress. The decision to administer oxygen on increased oxygen flow rates should be made in consultation with the child's parents.

Some children have chronic Type 2 respiratory failure and oxygen flow rates will have to be carefully considered to avoid hypercarbia. The decision to initiate and continue oxygen therapy in hospital should not be made based only on oxygen saturation readings but with consideration of level of dyspnoea, tolerability of oxygen mask or nasal cannula and child and parental wishes.

Home oxygen

Administration of supplemental oxygen in the home setting has additional complexities. The first issue is to determine whether supplemental oxygen will be required for a short period only (usually for end-of-life care) or whether it is likely provision will need to be long term.

Different funding pathways and equipment provision entities will be required for each child. Oxygen should be discontinued if there is no definite benefit noted.

Oxygen concentrators are useful for home use and are connected to a domestic power supply (rebates may be available from electricity suppliers for these). It is advisable to notify the electricity provider that the patient is receiving home oxygen to ensure their power is prioritised in the event of a power outage.

The use of oxygen prior to walking to the toilet or bathing may be all that is required. The maximum flow is 5L/min via mask or nasal prongs. This flow is sufficient for the majority of children but if a greater flow is needed an oxymiser nasal cannula can be obtained for use and allow a greater concentration of oxygen to be delivered. Portable oxygen concentrators are available but usually must be privately purchased.

Portable oxygen cylinders are also available and allow the child and family to leave the house. Care is needed when travelling with cylinders in the car as they need to be securely transported. A "C" size cylinder lasts for 2–3 hours at 2L/min. Reservoir cannulas or pendants conserve oxygen use such that oxygen is only used on inspiration however these are not routinely used for children as the child's inspiratory effort may be insufficient to trigger oxygen flow.

It is important to advise parents to ensure that no-one smokes close to the patient, near the source of the oxygen or anywhere inside as there is a significant explosion risk associated with oxygen concentrators and cylinders within the home.

COUGH

Many of the conditions causing dyspnoea will also produce cough. Cough results from irritation to the receptors in the upper or lower respiratory tract, the pleura, pericardium or diaphragm. Avoidance of irritants, the use of antihistamine or anticholinergic agents for post-nasal drip and antibiotics may provide symptomatic relief. Simple linctus will soothe the throat and reduce dry cough. For children with persistent dry cough, suppression is indicated. Opioids, due to their central action, are the treatment of choice. If a child is already receiving morphine for pain relief, increasing the total dose may be effective.

Bronchospasm can also contribute to cough and should be treated with nebulised or inhaled Salbutamol (Ventolin®). Nebulised saline is an effective mucolytic agent while nebulised local anaesthetic agents may be of benefit for intractable cough. Lignocaine 2% or bupivacaine 0.25%, 5ml via nebuliser every 4–6 hours can be used. **It is important to recognise that the gag reflex will also be impaired after this treatment. It is advisable not to eat or drink for 1–2 hours after.**

Non-pharmacological management strategies include humidification or use of a vaporiser in the child's bedroom. Cough related to pulmonary congestion can be eased with more upright positioning. Cold drinks should be avoided as these can trigger a coughing spasm.

Heart failure and pulmonary oedema

It is rare for a child who does not have known congenital heart disease to develop left sided heart failure or congestive cardiac failure.

If this is clinically thought to be occurring then frusemide (Lasix®) 0.5mg/kg sub-cutaneously can be given and repeated 4–6th hourly if necessary.

Acute pulmonary oedema is an emergency and should be treated with frusemide, morphine, oxygen, and positioning. Non-invasive ventilation may be helpful if available and indicated depending upon the stage of the child's illness.

In the case of a child with known congenital heart disease and deteriorating cardiac function, advice of the treating cardiologist should be obtained.

Home ventilation and tracheostomies

It is exceptionally rare for children to have home invasive ventilation and this topic is not covered further here. It is increasingly common that children with tracheostomies are referred to Palliative Care Services. Tracheostomy issues are best guided by Ear Nose and Throat (ENT) Surgeons and nursing staff, and are beyond the scope of this document.

Children with non-cancer life-threatening illnesses may have been commenced on home non-invasive ventilation (Continuous positive airway pressure [CPAP]/Bi-Level Positive Airway Pressure [BiPAP]/ Variable positive airway pressure [vPAP]) for sleep-disordered breathing or other indications, which may be used continuously, nocturnally or only when unwell.

Common issues arising from this intervention include:

- Navigating goals of care and escalation of intervention issues.
- Tolerability of non-invasive ventilation.
- Risk of aspiration.
- Equipment related issues including ill-fitting mask with leaks and/or pressure areas.
- Expert advice should be sought from a Respiratory Paediatrician for issues related to home ventilation.

Careful discussion with the respiratory team and family is required if decreasing the settings or cessation of ventilatory support seems appropriate due to disease progression.

EXCESS SECRETIONS

Gentle suction and physiotherapy can also have a role in managing secretions for patients at all stages of palliative care. Other non-pharmacological management strategies include regular positioning to allow secretions to drain, in combination with meticulous mouth care.

Hyoscine hydrobromide can be administered subcutaneously as a bolus (0.2–0.4mg every 4 hours) or by infusion (0.6–1.2mg over 24 hours). Glycopyrrolate (Robinul®) (4–10µg/kg q6h; max 0.4mg) also has anticholinergic properties and has a selective and prolonged effect on salivary and sweat gland secretions. Glycopyrrolate can be administered subcutaneously or intravenously, and is also compatible with morphine and midazolam. Consideration of the use of glycopyrrolate should be given if there is an inadequate response from hyoscine. Atropine (1% or 10mg/ml) drops are also an option as they are readily available in community pharmacies and can be administered sublingually (commencing with one drop every four hours). Atropine can cause bradycardia with repeated dosing.

ANAEMIA AND BLEEDING

ANAEMIA

Children with incurable leukaemia almost always develop significant anaemia. One of the most common sites of metastatic disease in children with cancer is the bone marrow. Consequently, anaemia is frequently seen as a complication in children receiving palliative care for cancer diagnoses. Children with non-cancer conditions can have anaemia due to chronic disease, nutritional deficiency, or blood loss due to various causes.

Decisions regarding red cell transfusion should be made on an individual basis and will depend upon the stage, life expectancy and symptoms of the child. If anaemia is interfering with the child's activity level causing tiredness, headache or irritability, whilst they otherwise are experiencing a reasonable quality of life, transfusion of packed cells may be appropriate. Packed cell transfusions can also be beneficial if the child has a special planned outing or activity arranged. However, as the disease progresses and the general activity level of the child is reduced, anaemia will be less symptomatic and any potential benefit of transfusion should be re-evaluated, especially with regards to the travel required to the hospital facility for transfusions. Discussion with the family regarding the value of ongoing transfusion should take place as it becomes evident that further transfusions are not appropriate.

BLEEDING

Thrombocytopenia can be due to primary bone marrow infiltration or failure, hypersplenism or medications. Significant thrombocytopenia may lead to spontaneous or easy bleeding. Bleeding can also occur due to coagulopathy secondary to liver disease, nutritional deficiencies,

disseminated intravascular coagulation or drugs – especially NSAIDs and steroids.

During curative treatment platelet transfusions are given to children when the platelet count drops below a defined level. As disease progresses, platelet transfusions are reserved for cases of significant bleeding such as epistaxis, bleeding gums or gastrointestinal bleeding. The decision to transfuse with platelets should be based on each individual patient, discussed with the family and reviewed regularly.

Generally children and parents are prepared to come to hospital for platelet transfusions. However, as the child's disease progresses trips to hospital can be a major ordeal, both physically and psychologically. It is in these circumstances that the feasibility of delivering platelets at home could be considered. There are blood and blood product standards developed by the Australian Commission on Safety and Quality in Health Care. These standards must be adhered to within the palliative care context. Therefore, there may be some limitations on availability of blood and blood products for transfusion in regional, rural and home settings. These practical issues need exploring prior to incorporating transfusions into the child's management plan.

However, it is possible to administer platelets at home, if suitable experienced support is available. Transfusion reactions including anaphylaxis are the main side effects of platelet transfusions.

To minimise transfusion reactions at home, all children receiving platelets should receive pre-medications (e.g. promethazine and hydrocortisone) and have platelets infused via a platelet leucocyte filter. Medications should also be available to treat anaphylaxis.

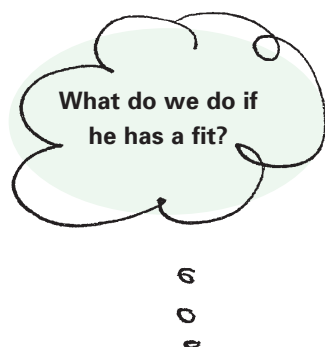
ANAEMIA AND BLEEDING

Bruising and petechiae are common with thrombocytopenia and coagulopathy. Catastrophic bleeding is unusual, but active bleeding is a very distressing event for the child and family, and prevention of major bleeding episodes should be attempted. Subconjunctival haemorrhages are obvious and frightening, but the child and family should be reassured that these bleeds are not life or vision threatening and do not require treatment. In the case of epistaxis, application of gentle pressure to the bridge of the nose, or icepacks on the back of the neck, will stem most episodes. Oozing from mucosal surfaces can lead to bleeding gums, dark stools, haematuria and rectal bleeding. Tranexamic acid (Cyklocapron® 20mg/kg q8h) may be given to help stabilise clots that form over the bleeding area. This can be given orally or as a mouth wash for mouth bleeds (500mg tablet dissolved in 5ml). Bleeding from ulcerated areas on the skin or peri-anally can usually be settled with topical 1:1000 adrenaline. Sucralfate dispersed in water-soluble gel can also be used topically to control local bleeding or oozing.

For major bleeding when death is imminent, treatment should be directed at calming the family and simple supportive measures. Bleeding generally lessens as the blood pressure and cardiac output drops. The use of dark coloured bedding and towels to disguise the extent of bleeding and the use of disposable pads and nappies can be helpful. If the child is aware, appropriate analgesia and sedation should be administered to relieve distress. It is helpful to have medications such as morphine, midazolam and adrenaline soaked dressings available in the home. If major bleeding is anticipated, it can be helpful to have a sensitive conversation with the family about how this distressing symptom can best be managed.

ANXIETY

All children will experience anxiety during the treatment of their life-threatening or life-limiting condition and when deterioration occurs this may be significantly exacerbated. Fear of the unknown, or of potential symptoms and suffering, will cause agitation in both the child and family. Anxiety is a normal response to these issues. Communicating with the child and family will help allay some fears, but occasionally anxiolytics may be of benefit. The use of a hypnotic such as temazepam (Normison®/Temaze®) at night will be helpful for some children and adolescents. Lorazepam (Ativan®) (0.02–0.06mg/kg q8–24h) is an intermediate acting benzodiazepine, can reduce anxiety and may be helpful in addition to being a hypnotic. Antidepressant medications can be helpful and are usually prescribed after liaison with the psychiatry team. Relaxation techniques, distraction, music and meditation for older children will reduce levels of anxiety. Members of the allied health team can assist with the provision of some of these strategies.



SEIZURES

Even brief generalised seizures can be very distressing for parents to witness, and the family should be prepared for such a possibility.

As with other symptoms, knowledge of the patient's past history and an understanding of the natural history of the underlying disease will suggest which children may be at risk (Table 14).

TABLE 14 Causes of seizures

Brain tumours – primary, metastatic or meningeal lesions

- Raised intracranial pressure
- Intracranial haemorrhage
- Metabolic disturbances
 - hypoglycaemia
 - hyponatraemia
 - hypocalcaemia
 - hepatic encephalopathy
- Infection and fever
- Pre-existing epilepsy

Treatment

Children who have a history of epilepsy or previous seizures will usually be on anticonvulsants and these should be continued. However, control of seizures may be lost if the child becomes unable to tolerate medication. Phenytoin (Dilantin®), however, has a relatively long half-life and levels may not fall until several doses have been missed (Table 15).⁵⁹

Buccal or intranasal Midazolam is a good first line agent for breakthrough seizure management where seizures are not controlled by prescribed anticonvulsants. It has been shown to be at least as effective as rectal diazepam in the acute treatment of seizures.^{59, 91} Administration via the mouth is more acceptable and convenient and may become the preferred treatment for prolonged seizures that occur outside hospital.⁹²

NEUROLOGICAL SYMPTOMS

TABLE 15 Treatment of seizures

Emergency treatment
Diazepam 0.2–0.4mg/kg IV or PR or Clonazepam PO drops 0.5mg <10y 1mg >10y IV, SC or PR or Midazolam 0.1–0.2mg/kg IV or SC < 20 kg: 0.3–0.5mg/kg buccal or intranasal > 20 kg: 5–10mg/dose buccal or intranasal
Maintenance treatment – oral
Phenytoin 2mg/kg q6–12h or Phenobarbitone 2.5mg/kg q12h Carbamazepine 2mg/kg q8h
Continuous treatment when oral route not possible
Diazepam PR (5mg 1–5yrs, 10mg >5yrs) as required Midazolam 50–150micrograms/kg/h (1mg/kg–4mg/ kg 24h). A starting dose is often 10–30mg/24 hours by SC infusion, increasing as required

The buccal or intranasal dose is the same as the oral dose of midazolam used for sedation (0.3–0.5mg/kg per dose, maximum dose 10mg). It is helpful to have a supply of oral midazolam or rectal diazepam in the home for emergency use for fitting or agitation, as they are safe and easily administered by carers. A seizure management plan with clear guidelines of when to call for further assistance or medication advice is necessary. When prescribing midazolam, specify plastic vials if available, as these will be easier to manage in this situation. Subcutaneous or intramuscular diazepam should be avoided because of local irritation and poor drug availability.

Oral clonazepam drops can be administered sublingually and may be useful for diazepam resistant seizures. Clonazepam may increase oral secretions and thus this needs to be taken into account especially when given regularly. If further seizures are likely, regular oral or subcutaneous anticonvulsants should be commenced. Oral phenytoin (Dilantin®) or phenobarbitone can be given as maintenance therapy.

If the child is unable to tolerate oral drugs, alternative routes and drugs will be required if seizures are a possibility. Midazolam is easily prepared and can be administered subcutaneously. It is compatible with morphine and also has an anxiolytic and sedating effect. If seizures occur despite a continuous infusion, boluses of midazolam can be given followed by an increase in the dose of infused midazolam. Increasing doses will generally obtain control over seizures. Phenobarbitone and clonazepam are also effective anticonvulsants, and can be administered subcutaneously if necessary. It is recommended these medications are titrated by a specialist team.

MUSCLE SPASM AND MYOCLONUS

Muscle spasm can occur as a result of immobility, pain, neuropathic spasm or cramp. Appropriate analgesia will reduce the protective muscle spasm effect. Low dose diazepam (Valium®) (0.1mg/kg/dose) can also be considered if muscle spasm is causing pain. Encouraging mobility or changing position regularly in children with little energy will also reduce spasm and the development of painful contractures.

For long term treatment of spasm, baclofen can be considered. The main concerns with baclofen are a possible reduction in seizure threshold and

potential adverse effects on swallow and airway protection. Involvement of a paediatric rehabilitation specialist or neurologist is helpful in this context. Localised spasticity can sometimes be managed with Botulinum toxin (Botox®) injections. For patients with severe spasticity, an intrathecal baclofen pump can be considered.

Myoclonus is involuntary twitching involving single muscles or groups of muscles. It is a recognised toxic effect of opioids, occurring more frequently if pethidine is used.⁹⁰ It is more common in the terminal phase of illnesses and in those with underlying renal impairment especially with morphine use due to the build-up of secondary toxic metabolites. Treatment includes reduction of the dose of opioid, if possible, or change to an alternative drug. Midazolam as a bolus or infusion is usually effective in controlling myoclonus.

IRRITABILITY AND AGITATION

There are many causes of irritability and agitation, many of which may be treatable (see Table 16). Reversible conditions can be managed appropriately but cerebral irritability can be difficult to manage with distress and inconsolability lasting for hours.

Gabapentin (Neurontin®) can be considered to manage distress, anxiety and pain. It is effective for cerebral irritability, visceral hyperalgesia and autonomic dysfunction.⁹³ It comes only as an oral preparation (capsules) but these can be split and the contents administered via NGT/PEG tubes. There are minimal-to-no interactions with other medications. It is renally excreted and the dose should be adjusted in renal impairment.

Gabapentin Dosing

Day 1: 5–10/mg/kg (maximum dose 300mg).

Day 2: 5–10mg/kg twice daily.

Day 3 onwards: 5–10mg/kg three times daily.

For marked distress, levopromazine, clonidine or chloral hydrate can be used.

Buccal midazolam (0.3–0.5mg/kg) can be used in a crisis situation to break the cycle of distress. This should not be considered for ongoing treatment of distress, but an option for crisis management.

In cases where irritability and behaviour disturbance are causing significant burden to the family, it can be helpful to consult with a psychiatrist for evaluation and guidance on whether psychotropic medications may be of assistance.

Non-pharmacological management strategies may include:

- A calm and quiet environment.
- Reassurance by touch and the voice of familiar people.
- Use of senses that are intact e.g. reading a story or music (auditory).
- Assess and manage a full bladder.
- Assess and manage pain.

ACUTE DYSTONIC CRISIS

An acute dystonic crisis is characterised by facial and skeletal muscle spasms and oculogyric crises. Acute dystonic reactions are usually a consequence of antiemetics which block central

dopamine receptors. These reactions are commoner in children, especially girls and, generally occur within a few days of starting treatment and may take 24hrs to subside after stopping the drug.

TABLE 16 Sources of pain/irritability in children with neurologic impairment

Somatic pain	
Head, eyes, ears, nose, throat	Headaches, VP shunt malfunction, otitis, corneal abrasion, sinusitis, pharyngitis, corneal abrasion, glaucoma, dental pain
Musculoskeletal	Chronic/acute musculoskeletal pain, spasticity, hip dislocation, fracture, osteomyelitis
Gastrointestinal	Gastro-esophageal reflux, esophagitis, pancreatitis (associated with hypothermia and valproate), ulcer, gallstones, cholecystitis, constipation, rectal fissure, food allergy, retching and vomiting
Renal	Urinary tract infection/pyelonephritis, neuropathic bladder, obstructive uropathy, renal stones (associated with ketogenic diet and topiramate)
Respiratory	Aspiration, reactive airway, costochondritis
Genito-urinary	Testicular/ovarian torsion, inguinal hernia, menstrual cramps
Skin	Pressure sore
General	Medication toxicity, sleep disturbance, obstructive apnoea
Neuropathic pain	
General neuropathic pain	Suggested by pain that appears out of proportion to an identified source or pain behaviours that develop weeks to months following surgery
Visceral hyperalgesia and Central pain	Suggested by pain and/or retching associated with gastric and intestinal feedings, intestinal gas, flatus, and bowel movements
Autonomic dysfunction	Suggested by sweating, increased salivation, flushing or pallor of skin, retching, vomiting, pain localized to the abdomen, agitation, arching, stiffening
Cerebral irritability	Diagnosis of exclusion. May be a feature of severe birth asphyxia and neurodegenerative conditions

NEUROLOGICAL SYMPTOMS

Drugs that may cause Acute Dystonic Crisis:

- Metoclopramide (Maxolon®)
- Haloperidol (Seranace®)
- Levomepromazine (Nozinan®)
- Risperidone (Risperdal®)
- Prochlorperazine (Stemetil®)

Acute treatment for symptom relief can be achieved with IV diazepam. For specific treatment, IV/IM benzotropine (Cogentin®) 0.02mg/kg (max 1mg) either intramuscularly or intravenously is recommended. This can be repeated once, but if the intramuscular route is used, allow 30 minutes to elapse before repeating. The same dose should be given orally, twice daily for the next 24–48 hours to prevent recurrence.⁹⁴

Dystonia as a chronic symptom is treated with various drugs and is mainly dealt with in conjunction with neurologists.

INSOMNIA

Sleep disturbance is common in children with life-threatening illness. The aetiology of insomnia is multi-factorial and is often a combination of physical, psychological and perhaps environmental factors. When depression or anxiety is a factor, consideration should be given to psychotherapy and pharmacologic treatment.

Lifestyle changes, including improved sleep hygiene and exercise may be helpful to improve sleep. Staff caring for patients in the hospital and hospice settings should reduce interruptions, noise and light as much as possible.

Low dose amitriptyline (Endep®) can be helpful as a pharmacologic agent for the management of insomnia in terminally ill children, particularly if neuropathic pain is also present. When clinically indicated, management of persistent pain with a background infusion of opioid or oral controlled release opioid preparation will enhance sleep and minimise the need for breakthrough analgesia. Melatonin can also be considered, particularly in children with neurological conditions and those with an altered circadian rhythm. Stronger sedative medications or psychotherapeutic agents may be required in very severe cases.²¹

The care of a child or young person with End Stage Renal Disease (ESRD) can be challenging and complex. This may include managing co-morbidities, ESRD-related symptoms, renal replacement therapy (RRT) or conservative supportive care. Supportive care needs to be offered to all patients with progressive end stage renal disease and those with a poor prognosis. An integrated early palliative approach may improve the symptom burden for patients pursuing active dialysis.⁹⁵

ESRD is the state in which a patient's renal dysfunction has progressed to the point at which homeostasis and survival can no longer be sustained by the native kidney and maximum medical management. RRT or pre-emptive transplantation may be required. Continual dialysis is based on a combination of biochemical abnormalities and clinical factors which include refractory fluid overload, electrolyte imbalance, acidosis, growth failure, uraemic symptoms and impaired school performance.⁹⁶

Palliative care in ESRD can be initiated whilst the patient requires RRT, such as dialysis or transplantation, and withdrawn if the child or young person improves. RRT may be necessary not only to prolong life, but to maintain a patient's quality of life at an acceptable level. In patients still receiving RRT, palliative care plays a supportive role.

Early integration of palliative care also facilitates discussions about transition to palliation in patients where renal replacement therapy is discontinued because it is thought not to be in the child's best interests. The palliative care team can help with discussions regarding discontinuation or abatement of dialysis, and end of life care.

The median survival time after stopping dialysis is variable and depends on co-morbidities and the rate of decline in renal function. Abatement of dialysis is a less common occurrence in paediatric palliative care compared to adult practice.

Decisions regarding whether to offer or continue dialysis should consider the following factors:

- Prognosis – which can be uncertain and unpredictable.
- Symptoms associated with ESRD.
- Reduction in dose or cessation of medication because of the diminished renal function to avoid accumulation of metabolites.
- Dietary and fluid restrictions.
- Place of end of life care and advanced care plans.
- The severity of other co-morbidities e.g. other organ failure.

In some circumstances, the decision to stop dialysis may not be elective due to contra-indications for continuing either peritoneal dialysis or haemo-dialysis (e.g. difficulty maintaining vascular access). Alternatively, a sudden deterioration (e.g. infection and sepsis resistant to antibiotic therapy) is also possible.

While renal transplantation can be offered to many children, it may not always be possible depending upon the size of the child, availability of donor kidneys and presence of other co-morbidities (e.g. liver or heart failure). Further, it is also possible that a renal transplant may fail or be rejected. Options at the time of renal transplant failure include re-commencement of RRT, consideration of a further transplant and sometimes supportive care only.

Symptom management in End Stage Renal Disease

Symptomatic management for patients with ESRD is usually managed by the nephrologists. Palliative care specialists help in the management of difficult symptoms which are also commonly encountered in other end-stage conditions. ESRD-related symptoms can include pain, dyspnoea, anorexia, nausea, pruritus, peripheral oedema or difficulty with sleep. Symptoms related to the CNS can include malaise, confusion, apathy, twitching, seizures, stupor and coma. Psychological symptoms can include depression and anxiety. Management and treatment of these symptoms are discussed elsewhere in this book.

It is hoped that both health professionals and parents can mutually agree upon various interventions that could or should be offered to the child. This involves balancing benefits and burdens of various interventions with a focus on the child's quality of life. This is often a process and requires multiple discussions. Not all families are able to make these difficult decisions and an approach that is flexible and patient and family centred should be taken. Discussions and decisions around management of issues such as fluid balance (including albumin infusions) and whether it is appropriate to continue performing blood tests (e.g. monitoring of electrolytes) are important to consider in light of the parent's preferences and child's quality of life.

The symptom management of children with ESRD who are dying will require reduced dosing and frequency of medications. Children with ESRD who are dying require half the recommended dose of glycopyrrolate to manage secretions. The presence of terminal agitation is usually managed by a reduced dose of midazolam with

an increased dose interval to avoid excessive drowsiness.⁹⁵ Fentanyl is the opioid of choice for pain or dyspnoea. Other opioids such as oxycodone and hydromorphone can be used short term if fentanyl is not available. They should be used with caution starting at a low dose and reduced frequency.^{97, 98} They should be given on an "as required" basis at the commencement of therapy, rather than regularly or as an infusion, particularly at the commencement of therapy.⁹⁵ Methadone and buprenorphine are opioids that are safe to use in renal failure. Methadone and fentanyl are not readily dialyzable.^{98, 99}

Dermatologic conditions in paediatric palliative care

Dermatological conditions and symptoms in paediatric palliative care are common. Symptoms can be distressing for the child and parents. Children with life-limiting illnesses, like other children, are prone to skin conditions.^{3, 21} Commonly occurring skin problems include itch, rashes, skin ulcers or skin infections. Dermatological symptoms are often attributed to the underlying medical condition, or complications related to treatment. Management may require a multidisciplinary approach especially in the presence of pain or itch which can be difficult to manage. Symptoms amenable to treatment may be relieved by collaboration with a dermatologist. Some symptom presentations may not be obvious and require a high index of suspicion, especially in the unsettled non-verbal child. Commonly occurring symptoms are discussed below.

PRURITUS

Pruritus or itch is an unpleasant cutaneous sensation that provokes the desire to scratch.²¹ Its presence can be both distressing and difficult to manage. The pathophysiological mechanism of pruritus is complex and it involves activation of C-fibres in the skin through mechanical stimuli, or chemical substances such as histamine and several other substances. Neuronal mechanisms have also been identified in the pathophysiology of itch. Neuropathic analgesics, such as gabapentin and pregabalin, have been shown to be efficacious antipruritic therapeutic options. Although further research is required, gabapentin is safe and has been found to be effective in pruritus related to uraemia, cancer, opioid medication and burn injury.^{100, 101}

There are numerous causes of pruritus which include dermatologic conditions, systemic disease, neurologic conditions, uraemia, biliary obstruction, medication side effects, dry skin and psychological factors. Management includes a thorough history to determine the cause, physical examination to evaluate the skin and appropriate investigations which may include a skin biopsy. Some causes of pruritus are discussed below.

Dry skin

Dry skin can cause or exacerbate pruritus. The commonest cause of dry skin in children is atopic dermatitis, also known as atopic eczema. The aetiology of atopic dermatitis includes a complex interplay of genetic and environmental factors and there is no single cause. Another cause of dry skin includes Ichthyosis, a genetic skin disorder, that presents at or shortly after birth and persists throughout life. This usually presents with a chronically dry, scaly skin. Dry skin can present in a spectrum of other medical conditions and can also be associated with dehydration.²¹ The mainstay of dry skin treatment is keeping the skin moist with oil based emollients.

Opioid induced pruritus

The incidence of opioid induced pruritus is variable and commonly seen with the parenteral use of opioids.²¹ The pruritus is mainly localised to the face and trunk. The exact mechanism by which opioids cause pruritus is not known. Treatment may require switching opioids.⁴⁵ Alternatively, a low dose opioid receptor antagonist, such as naloxone, can be trialled.⁷⁸ Other treatment alternatives are serotonin receptor antagonists (e.g. ondansetron)¹⁰² and dopamine (D₂) receptor antagonists (e.g. droperidol).

Cholestasis

The mechanism of cholestasis induced pruritus is unclear. Circulating bile acids and endogenous opioids are thought to play a role. The main management of this pruritus involves decreasing the level of circulating bile acids.¹⁰² Bile acids are reduced by using cholestyramine, rifampicin or phenobarbitone. Other therapies that have been shown to be effective include opioid antagonists such as naloxone and naltrexone.^{21, 78} In some instances surgical interventions may be helpful. Interventions include partial external biliary diversion, stenting of the bile duct and terminal ileal exclusion in patients with intrahepatic cholestasis.²¹

Uraemia

Uraemia induced pruritus is seen commonly in patients with chronic renal failure receiving dialysis.²¹ Therapeutic measures include an enhanced dialysis regimen, correcting any electrolyte abnormalities, and treating hyperparathyroidism. Recent new therapies that have been successfully trialled include the use of gabapentin and UV- B therapy.

General measures for treating pruritus

- Treat the underlying medical conditions.
- Shorter bath times and the use of lukewarm water, with mild or low pH soaps, and the addition of sodium bicarbonate to the bath.
- Use of bath oil or soap substitute such as aqueous cream, bath washes, hydrating or moisturising lotions, and soap free cleansing bars.
- Use shampoo substitute (e.g. oatmeal shampoos).
- Keep the skin moist by regular use of emollients. There are a variety of emollients. Examples include Glycerol 10% in sorbolene cream and aqueous cream (aqueous cream strength can be varied by adding liquid paraffin, white soft paraffin, or olive oil). Urea based creams are useful for very dry skin or coexisting ichthyosis.
- Maintain a cool ambient temperature and avoid rapid temperature changes.
- Prevent the child scratching. Keep the fingernails short, use mittens to cover the hands.

Pharmacological treatment of pruritus

Topical corticosteroids are used for the treatment of atopic dermatitis. Ointment based creams are better at maintaining moisture and the use of wet dressings is recommended if there is severe inflammation and thickened skin. Resistant cases can be referred to a paediatric dermatologist and may require treatment with topical immunosuppressants such as Pimecrolimus.

Antipruritics include either sedating antihistamines (such as promethazine hydrochloride and trimeprazine), or non-sedating antihistamines (such as cetirizine or loratadine). The sedative effects can be helpful in calming any agitation associated with pruritus, particularly at night.

The use of some psychotropic medications has also been described in the alleviation of itch e.g.

- Doxepin – tricyclic anti-depressant.
- Paroxetine – selective serotonin uptake inhibitor.
- Mirtazapine – tetracyclic anti-depressant.

PRESSURE ULCERS

Children who are unable to reposition themselves, and who are bed or chair bound are at higher risk of developing pressure ulcers. Pressure ulcers or sores can develop quickly. Areas likely to develop pressure ulcers are the heels, neck, sacrum and scalp. Risk factors that lead to pressure ulcers are a poor nutritional status, conditions leading to moist skin like incontinence, and poor circulation.

Preventive measures include:

- Reposition patients by lifting to avoid shearing forces every two hours for patients that are bedbound, and hourly if chairbound.
- Use pressure reducing mattresses and gel pads.
- Use absorbent nappies for incontinent patients.
- Protect the skin by using barrier creams.
- Avoid letting the skin become dry – use regular moisturiser.
- Examine pressure areas regularly.

Pressure ulcers can be painful and require appropriate analgesia depending on the severity. Refer to the “Pain” section earlier in this book. Good wound care is essential for wound healing and pain control. Hydrocolloid dressings are used to dress shallow ulcers. Some ulcers can become infected and these require antimicrobial dressings, superficial debridement and wound cleansing.²¹

The management of pressure ulcers also depends on the patient’s prognosis. In patients with a short life expectancy, managing accompanying symptoms of pain and odour may be more important than healing the ulcer. Odour is often well managed with topical metronidazole gel.¹⁰³ Silver sulfadiazine, iodine cleanser or charcoal dressings are other alternative therapies to manage odour. Environmental adjustments to control odours include placing activated charcoal

in a container to absorb odour. Alternate smells that can be introduced to the room using aromatherapy include vanilla, citrus or vinegar.

OTHER SKIN CONDITIONS

Nappy rash

There are many factors that contribute to dermatitis confined to the nappy area. These include excessive skin hydration, friction trauma, irritants such as ammonia from urine, soaps and other creams and powders, and local fungal infection. Recommended treatments include frequent nappy changes, use of barrier creams, and treating any associated candida infections.

Skin infections

Skin infections can occur as a complication of wounds or trauma leading to localised infection such as cellulitis. A wound swab should be taken and appropriate antibiotic treatment instituted. Tinea is another common infection in children. Other uncommon skin infections are herpes zoster and herpes simplex.

Hyperhidrosis

There are several causes of hyperhidrosis (excessive sweating) that are seen in children with life-limiting conditions. Causes include psychological factors, familial dysautonomia, Epidermolysis Bullosa, heart failure, metabolic and endocrine conditions (such as hypoglycaemia and hyperthyroidism) and medication side effects.⁹⁶ Treating the underlying medical condition (if possible) is the preferred management strategy. Anticholinergics medications can also have a role in management.⁹⁶

Epidermolysis Bullosa

Epidermolysis Bullosa (EB) is an inherited blistering condition with a wide spectrum of disease and prognosis. The skin and mucosa is extremely fragile. The most severe forms are junctional EB and recessive dystrophic EB. Complications (dependent on the type of EB) may include early death, formation of contractures, and increased risk of developing squamous cell carcinoma.

Symptoms of EB include varying severity of pain and gastrointestinal symptoms (including gastro oesophageal reflux) as well as bleeding from mucosal blistering.³ Rare complications include the development of dilated cardiomyopathy and renal complications. Care of children with EB include gentle baths, use of lubrication to minimise trauma and avoiding heat and high humidity – which help prevent blister formation. Broken skin should be appropriately dressed and antibiotics should be commenced early to prevent infection.³ Pharmacological and non-pharmacological management strategies should be used to lessen anxiety associated with dressings, and appropriate interventions to manage pain should be used.

The perinatal period of life is defined in different ways. Depending upon the jurisdiction, it starts at the 20th to 28th week of gestation and ends at four weeks after delivery.

Perinatal palliative care is a compassionate model of support that can be offered to parents who find out during pregnancy, or shortly after birth, that their baby has a life-limiting condition. As prenatal testing continues to advance, more families are finding themselves making decisions antenatally in this difficult situation. Perinatal palliative care incorporates the philosophy and expertise of palliative care into the care of this population of families. For parents who receive a life-limiting prenatal diagnosis and who wish to continue their pregnancies, perinatal palliative care provides the opportunity to embrace the life of their baby – before and after birth. It can provide an opportunity “to parent” their child despite limited time.

Ideally, this support begins at the time of diagnosis, not just after the baby is born. It can be thought of as “hospice in the womb”. It includes birth planning and preliminary medical decision-making before the baby is born, as well as more traditional palliative care on the birth suite, post-natal ward, neonatal nursery or at home after birth (if the baby lives longer than a few minutes or hours).

Palliative care can also include medical treatments intended to improve quality of life for the infant. This approach supports families through the remainder of the pregnancy, through decision-making before and after birth, and through their grief. Perinatal palliative care empowers families to make meaningful plans for their baby’s birth, life, and death, and offers dignity to the baby and family in keeping with their spiritual and cultural context.

CONDITIONS REQUIRING PERINATAL PALLIATIVE CARE

Congenital malformations are the leading cause of death in the neonatal group. However, some babies will die in the newborn period because of complications relating to extreme prematurity, or because of severe life-threatening illnesses that do not respond to intensive care interventions (e.g. severe congenital diaphragmatic hernia). It is important that the palliative care approach is integrated into neonatal care units for all patients when required, including for those with extreme prematurity and life threatening illness. Because of this, staffing and policies around bereavement support and follow up are particularly important in the neonatal intensive care environment. Consultation with a paediatric palliative care team can assist with complex cases (e.g. related to symptom management, home care or difficult ethical issues).

Congenital malformations can be multiple, severe and lead to miscarriage, stillbirth or a shortened life span. Conditions include:

- Major structural abnormalities in the brain (e.g. anencephaly, hydranencephaly and holoprosencephaly).
- Chromosomal abnormalities (e.g. Trisomy 18 and Trisomy 13).
- Severe complex congenital heart malformations (e.g. hypoplastic left heart syndrome).
- Severe neuromuscular conditions (usually diagnosed post-natally, although there may be suspicion of a neuromuscular condition with reduced foetal movements or polyhydramnios).
- Severe renal abnormalities (+/- pulmonary hypoplasia).
- Life-threatening skeletal dysplasia.

Prognostication

Predictions of life expectancy can be uncertain. Some infants with life-limiting conditions do live beyond the first year of life.¹⁰⁴ Health professionals need to be comfortable with this uncertainty, and be willing and able to provide support over an extended period of time in such situations. A process of parallel planning is required where outcomes ranging from intrauterine demise, stillbirth, death shortly after birth or a longer time frame of survival are considered. It is also important that parents are supported in a non-judgemental way as they care and make treatment decisions for their child. Early introduction to a paediatric palliative care service, often in the antenatal period, may be helpful to facilitate this. Graduation from the palliative care service could be considered if the baby survives past the expected time frames.

Options available to families

The grief that parents will experience around an antenatal diagnosis of a life-limiting condition can be affected by a lack of consistent medical information and not being given options. Options available to parents in this difficult situation include termination or continuing the pregnancy. Some families (e.g. children with hypoplastic left heart syndrome) may pursue more intensive interventions after birth, such as intensive care and surgical options of management for their child. However, many families will prefer to have a palliative care plan which begins as soon as their child is born. This can be facilitated by an inter-disciplinary team supporting the family. It requires collaboration between obstetric, maternal foetal medicine, neonatology, paediatric and palliative care teams. Formulation of a "Birth Plan" is integral to this process.

STEPS IN CREATING A BIRTH PLAN

If parents elect to continue the pregnancy and for the baby to receive palliative care following birth, a number of important steps must be undertaken to create a birth plan above the standard midwifery or obstetric plan.

This includes an open discussion of all pertinent issues. The discussion needs to be sensitive and non-threatening, with clear and compassionate communication.¹⁰⁵ Holding, touching, seeing, dressing and bathing the baby is often important. Some other important considerations include:

- Mode of delivery (vaginal birth versus caesarean).
- Resuscitation planning – particularly at the time of delivery. In some situations it will be appropriate to not initiate life support upon delivery.
- Pain and symptom management – pain relief may be provided through the intravenous or subcutaneous route. There is also the option to administer medications via enteral, sublingual, buccal or intranasal routes, avoiding invasive procedures. The pharmacokinetics of various drugs is age dependent.

For example, the half-life of morphine is longer and its clearance is less in neonates compared to older children. Therefore the dose of morphine is 0.05–0.1mg/kg orally or 0.025mg/kg intravenously every six hours. For paracetamol, the recommended total daily dose for paracetamol is 60mg/kg/day (compared to 90mg/kg/day in older children).

- Provision of nutrition – this has great meaning to all parents and should be considered (including provision of breast milk). However, feeding is not always possible and/or feasible. In some cases, it can be helpful to discuss the appropriateness of nasogastric tube feeding before birth, so that a clear plan can be documented ahead of time. Drops of breast milk or sucrose can also be used for comfort with or without presence of a sucking reflex.
- The option of home care should be provided to the family. This should include liaison with the family's general practitioner and determination of a local hospital and paediatrician who could care for the child if they required admission for symptom management or inter-current illness. In some jurisdictions, hospice can be explored as a location of care.
- Cultural and spiritual care (e.g. consideration of baptism or other important spiritual rituals for the family). Cultural awareness and sensitivity is also important in this context.
- Memory making (e.g. photographs or sketches, memory boxes with medical items, footprints/handprints, plaster cast and locks of hair).

It is also helpful if the care plan is communicated to all staff (medical, nursing, allied health and pastoral care) **and documented clearly in the maternal medical record**. This is particularly important as the delivery can occur after hours and on weekends. The whole team needs to be aware the plan.¹⁰⁴

Plans for bereavement care and support for the entire family will also be important to consider. Follow up is also necessary for obstetric care with due consideration taken for risks of post natal depression. Depending on the diagnosis, some families may require follow up appointments with genetic services.

THE DYING PROCESS

Parents often wish to know the changes that may occur in their child as death approaches. The dying process is often referred to as the terminal phase of illness. The body begins to shut down as major organ functions are progressively impaired. This is usually a gentle and undramatic series of physical changes which are not medical emergencies requiring invasive interventions.

Parents need to know that these physical changes are an expected part of the dying process. It is very important that families are well supported at this time. If the child is dying at home, 24-hour support from experienced staff that they know and trust is essential. Home visits by the GP, domiciliary nurse and specialist liaison nurse, to assist with managing the child's symptoms are greatly appreciated by the family. This is a very emotionally draining and difficult time for the whole family. It is important to listen to parents' concerns and fears, and when necessary offer guidance and advice on how best to provide comfort to their dying child.

NOISY/RATTLY BREATHING

Excessive secretions, or difficulty clearing pharyngeal secretions, will lead to noisy, gurgling, or "rattly" breathing. Generally this occurs during the terminal phase of the child's illness and is associated with a diminished conscious state. It can also be problematic for children with neurodegenerative diseases or brain stem lesions where swallowing is impaired. Positioning a child on their side or with their head slightly tilted down will allow some postural drainage – this may be all that is required. Reassurance and explanation to the family is essential, as the noise can be very distressing. However, the child is usually unaware and untroubled by the noise and secretions.

Anticholinergic drugs, (e.g. hyoscine hydrobromide or glycopyrrolate), can be used to reduce the production of secretions. For children with chronic conditions, a portable suction machine at home may be of benefit.

INCONTINENCE

During the dying process, there may be a relaxation of the muscles of the gastrointestinal and urinary tracts resulting in incontinence of stool and urine. It is important to discuss this possibility with parents, including how they wish to manage incontinence. If the child is close to death, parents are often reluctant for a catheter to be inserted to drain urine and may choose to use incontinence pads or disposable incontinence draw sheets. It is important for the family that their child's dignity is respected. Disposable draw sheets are also useful for diarrhoea.

EYE CHANGES

The pupils of a person who is dying may become fixed and dilated. Their eyes may become sunken or bulging and glazed. If eyes are bulging (which may occur in neuroblastoma), a small damp bandage placed upon the eye may provide some comfort. Eye secretions can be removed with a warm damp cloth.

RESTLESSNESS AND AGITATION

Generally, a child who is dying will spend an increasing amount of time sleeping. This is in part due to progressive disease and changes in the body's metabolism, but may also be due to progressive anaemia or sedation from opioids required for pain relief.

Some children remain alert and responsive until the moment of death. Others may become confused, semiconscious or unconscious for several hours or days. Restlessness and agitation during the terminal phase is not uncommon and may be due to increasing pain, hypoxia, nausea, fear or anxiety. Agitation may be the child's only way of communicating distress. A calm peaceful environment and the presence of parents and family will assist in relieving the child's anxiety. The child's speech may become increasingly difficult to understand and words may be confused. Even if the child may not be able to communicate, they may be aware of the people around them. Hearing is the last sense to be lost and the family should be encouraged to talk to their child. They may also like to play their child's favourite music, read stories or just sit with and touch their child so the child knows they are not alone. These measures will assist in relieving anxiety.

However, agitation and restlessness may continue if the cause is due to pain, hypoxia, nausea, or metabolic disturbances. Pain relief should be increased and this may be all that is required. If agitation continues, additional drugs may be required. Treatment is then directed at increasing sedation. At this stage oral medications may not be tolerated and alternative routes of medication are essential. Midazolam can be administered via the intranasal, buccal, intravenous or subcutaneous route. Clonazepam can be administered sublingually. Occasionally, there may also be indication for medication to be administered rectally (e.g. diazepam or paracetamol).

CONTINUOUS SUBCUTANEOUS INFUSION

A continuous subcutaneous infusion of a combination of drugs is simple to commence and is well tolerated by most children. A continuous subcutaneous infusion of morphine and midazolam in a syringe driver is effective in controlling pain, agitation and restlessness. A subcutaneous cannula with a side injection port should be used when starting the infusion; this enables bolus doses of medication to be given if required (Appendix 2). If the child is at home, the domiciliary nurse, specialist liaison nurse or G.P. can commence the infusion.

If bolus doses of drugs are needed for breakthrough pain or increasing irritability, parents are generally able to administer bolus doses of morphine or midazolam with appropriate education and support. Doses of medication should be prepared, labelled and stored in the fridge by visiting clinicians. A medication sheet should be provided to parents to record all times and doses of medication they administer.

Occasionally haloperidol (Serenace®) or levomepromazine (Nozinan®) is required when benzodiazepines are unsuccessful at relieving agitation. Regular monitoring of the effectiveness of medication is essential. Side effects (especially dystonic reactions) should also be observed for. Additional drugs can be added to the syringe driver if needed. Morphine, hyoscine, haloperidol, metoclopramide and midazolam are all compatible and can be combined to be delivered as a subcutaneous infusion. Precipitation of drugs may occasionally occur if high doses are required (see Appendix 3).

THE DYING PROCESS

Benzotropine (Cogentin®) should always be available in the home for urgent management of dystonic reactions. See Appendix 7 for recommended dosages of medications.

HOME CARE PACK

For the child dying at home it is important that the treating hospital has dispensed a home care pack containing drugs and other items that may be required in the terminal phase of care. This ensures drugs are available in the home if, and when the child requires them. Without a home care pack there may be a considerable delay in getting drugs or appropriate paediatric sized items required for symptom relief (Appendix 4).

CIRCULATORY AND RESPIRATORY CHANGES

As the heart slows and the heartbeat is irregular, circulation of blood is decreased to the extremities. The child's hands, feet and face may be cold, pale and cyanotic. The child may also sweat profusely and feel damp to touch. Parents may wish to change the child's clothes and keep them warm with a blanket or doona. Respirations may be rapid, shallow and irregular. Respirations may also slow with periods of apnoea. This is called Cheyne-Stokes breathing and is common in the last hours or days of life. This breathing pattern is distressing for parents and siblings to witness, and they need reassurance that it is an expected part of the dying process and is not distressing for the child.

It is important to inform parents that when death occurs the child may be incontinent of urine and stool. There may also be ooze from the mouth and nose, particularly if they roll their child to undress and wash them. Parents who are not aware of these possibilities may become distressed if this occurs.

In our efforts to achieve a peaceful death for the child, it is essential that symptoms are closely monitored and there is ongoing assessment of the effectiveness of therapy given. Early detection of symptoms and appropriate treatment is crucial if we are to achieve a pain free and peaceful death for the child.

WHAT TO DO WHEN A CHILD DIES

The death of a child is an emotionally painful experience and no amount of preparation can entirely prepare a family for when this happens. Additionally, for many parents the death of their child will be the first death they have witnessed.

Parents require gentle and sensitive preparation for what needs to be done when their child dies by a familiar member of the team. Some parents will ask questions as they start to prepare for their child's death, while for others this topic is too painful to discuss prior to their child's death. It is important to reassure parents that it is reasonable to ask questions and be prepared prior to the death if that might be helpful for them. With parental permission, inclusion of the dying child and siblings in this process, and sharing information, may reduce their sense of isolation and encourage expression of their thoughts and wishes.

Parents should be informed of the minimal legal requirements that are to be observed after their child's death; i.e. that a health professional will assess their child, confirm death has occurred and complete the required documentation. When parents are aware there is no urgency for this, they are able to do what is truly important – be with their child and say their goodbyes. It may be helpful to explain the practical tasks that need to be accomplished and a time frame for doing these (e.g. funeral arrangements) and to give parents' permission to make choices that best suit their needs (e.g. choosing a funeral director).

Hospital autopsies and tissue sampling after death

In some cases, a hospital autopsy may be performed for non-coronial cases to confirm the extent of an illness or disease.

This can assist with understanding the cause of death. Understanding a rare disease process, or why the death occurred more suddenly than expected from the primary disease, may be important to understand for both families and medical teams. With any patient's terminal illness, thought as to whether an autopsy would help in understanding of the disease process should be given. The family should be asked if they have any unanswered questions regarding their child's illness and the medical staff should consider if an autopsy would assist with the answer. In many situations the answer will be no.

There is also scope for limited autopsies (e.g. autopsy limited to brain, lung or abdominal organs). Post-mortem tissue samples may also be taken for a variety of purposes, including investigation of the underlying aetiology for metabolic and genetic conditions. Such biopsies usually need to be performed in a timely manner and samples sent to the laboratory urgently. The metabolic service is normally be involved in liaising with other teams and the laboratory about this process. Discussions around the issues of autopsy should be documented in the medical record. Some parents may also give consent for their child's tumour (e.g. brainstem glioma) or a specific organ (e.g. brain) to be donated to the hospital for research purposes.

DEATH AT HOME

When it is likely that a child will die at home, a member of the care team should explain in advance what procedures are required. This may vary in different jurisdictions across the nation. In particular, parents should know who they need to call at the time of the death, and who will visit the home and complete a preliminary death certificate or "pronunciation of life extinct form" (according to State requirements). This may be

WHAT TO DO WHEN A CHILD DIES

required by the funeral directors before they can move the child's body.

There may be the need for prompt action for some families where cultural or religious rituals require specific time frames to be adhered to, or if autopsies are required, or in the rare case if the death is a Coroner's case. Otherwise, parents should be informed that nothing needs to be done in a hurry when their child dies. This is very much a private time for family to say their individual goodbyes. Saying goodbyes and performing 'rituals' are important as they enable parents, siblings, and other family members to express their love, sorrow, relief, regrets and share precious memories.

Washing the child for the last time, dressing the child in special clothes, taking photos, playing favourite music, praying together, touching and cuddling the child, talking to the child, taking foot and handprints, cutting a lock of hair (this must be done with parental permission) and writing a message or poem for the child are all examples of rituals that families have found helpful.

A death certificate will need to be completed but does not necessarily need to be done at time of death. A doctor (e.g. GP, paediatrician) who has consulted the child in the past three months must complete this, and pre-death identification of this person may avoid unnecessary stress at the time of death. This requirement may vary between jurisdictions. It will be important to know at this time whether the family want cremation or burial, as those choices may define the paperwork required. The death certificate is usually collected by the funeral director; this may be billed as a cost to the parents.



Parents often ask if the police need to be phoned once the child has died. As the child is dying of a progressive disease, and death is expected, the police do not need to be contacted at the time of death. However, it is prudent to check in advance that there are no indications to notify the Coroner (e.g. a child under the care of Child Safety or Protection Services).

Similarly it is helpful to discuss with families whether or not they should call an ambulance. There is no need to call an ambulance after a child has died. In many states, if an ambulance is called prior to death, resuscitation will be attempted irrespective of the information given by family members. It may be helpful for the family to have a letter for the ambulance, a resuscitation plan, or Allow Natural Death document that explains the illness and expected treatments.

At the appropriate time for the family, a funeral director will need to be called. Many families would have already chosen the funeral director they wish to use. Parents need to negotiate with the funeral director the time they will attend to collect the child's body. It is very important that parents remain in control of the timing and they are not hurried. Some families may choose to keep their child's body at home for some time, including days and perhaps until the funeral.

WHAT TO DO WHEN A CHILD DIES

If this is the case, the family will need information and support to manage the child's body and the changes which occur post mortem. They should be advised of changes including colour, stiffness, and fluid leakage, and to keep the body in a room as cold as possible. A family may choose to have the body leave the home temporarily for embalming with the funeral director. From a cultural or religious perspective, some families may require prompt burial or cremation even within hours of death. Early identification of these requirements will necessitate meticulous pre-planning, including pre-engagement of a funeral director, cultural/spiritual advice, family preparation and prompt processes for death certification.

Funeral directors are on call 24 hours a day and parents can phone them at any time. There may be an extra cost if they attend the home after normal working hours. When preparing parents for what happens at the time of death, it may be helpful to explain how the funeral director will transport their child. It is a legal requirement that a body is transported in a body bag. The funeral director can be asked to leave the child's face uncovered as they are moved to the vehicle, but they will need to close the body bag for transport. Families may like their child to leave with a doona, pillow, favourite toy or other item. These can be collected later from the funeral director. It is helpful for the GP or palliative care coordinator to be aware of parent's desires and plans before the child dies so that appropriate support can be offered at the time of death.

DEATH IN HOSPITAL

Many of the principles discussed regarding what to do when death occurs at home also apply for when death occurs in hospital. It is important that the family is given as much time as they need to perform important rituals and say their goodbyes. Some families may need to spend hours with their

child before they are ready to say goodbye. It is important for hospital staff to find the balance between respecting a family's need for privacy, and identifying their need for support. In the busyness of the acute hospital setting, it may be necessary to advocate and ensure the family is allowed sufficient time to say goodbyes to the child and the staff who have cared for them.

This may require some management of both time and staff. The time needed to support staff and allow for debrief also needs to be considered. In non-acute settings (for example hospice) there is often more perceived time and space available for everyone to do this.

It is our experience that families find not taking their child home and leaving the child's body extremely painful and distressing. Families would have travelled to hospital and back home many times with their child during their illness. They may elect to have the funeral director collect their child's body from the ward, rather than go to the hospital mortuary. Parents may also wish to walk with their child on their final journey out of the hospital to the funeral director's vehicle or mortuary viewing room. If this is their wish, a member of the team should walk with the family and be available to support them when they leave their child's body. When walking with the child through the hospital, staff need to consider who is nearby and 'clear the way' (from other families/staff) before the child is moved from the ward. Parents may want to carry their own child for this journey and this should be assisted wherever possible.

Taking the child home from the hospital after the death can be an option for some families. This can be discussed with sensitivity and careful consideration of the support needs of the family, community capacity, transport requirements, funeral director engagement, and legal requirements such as certification.



It is not uncommon for parents and teenagers to ask about the possibilities of organ donation as they approach end of life. Questions regarding organ and tissue donation should be discussed with the family by a member of the team who has knowledge of the processes involved.

While many diagnoses render a child unsuitable as an organ donor, they may be able to donate specific tissue for transplants, including cornea, heart valves, skin and bone. The underlying condition, presence of infection, and size of the child influence whether a child would be a candidate for tissue donation.

Mechanical ventilation is not required for tissue donation and the tissue can be retrieved several hours following death. The child does not have to die in hospital to be a tissue donor. However, pre-planning is important to achieve tissue donation. If the child dies at home, the procedure for retrieval of tissue is coordinated through the State Donor Coordinator and the funeral director.

Families are able to spend time with their child following retrieval of tissue if they wish. Support is provided to the family from the Transplantation Service including provision of basic (anonymous) information about the recipient of their child's donated tissues.

A Donor Family Support program is available with ongoing free of charge counselling and other supports through a bereavement counsellor.

Further sources of information on tissue and organ donation are available through:

Donate Life Network

The Organ and Tissue Authority
Tel: 02 6198 9800
<http://www.donatelife.gov.au/>

Australian Capital Territory Tel: 02 6174 5625

New South Wales Tel: 02 8566 1700

Queensland Tel: 07 3176 2350

Northern Territory Tel: 08 8922 8349

South Australia Tel: 08 8207 7117

Western Australia Tel: 08 9222 0222

Victoria Tel: 1300 133 050

Tasmania Tel: 03 6270 2209

An organ and tissue donor coordinator is available for consultation after hours in all states.

FUNERALS

In many societies, funerals are an important part of the grieving process and help to acknowledge the importance of the child's life and offer support to the family and the whole community. A funeral is an important event for many families and can be conducted in a range of ways. Funerals enable a family and community to express some of their emotions and thoughts about the person who has died, including how they have been affected by the person's life and death, and about numerous other issues of importance to the individuals and family involved. For many people it is also an opportunity to express elements of their faith or spirituality.

It is current practice in Australia and New Zealand that funeral directors guide families and their spiritual or religious representatives, or a civil celebrant, through the planning process including the necessary paperwork and conduction of the ceremony. This may be as much or as little as the family requires and in some situations a family may choose not to use a funeral director but to manage the process themselves, or not to have a funeral ceremony at all.

It may be helpful for the health professional to raise the issue of funerals before a child's death. Families may have thought about funerals privately and appreciate the opportunity to explore the options further even though such discussions can be distressing. Families may find it helpful to have some time to think through the decisions that need to be made about the funeral before the death has occurred. While it may be upsetting to do this, it can allay anticipatory anxiety and it may be easier to have made some decisions earlier rather than in the heightened distress of the hours and days after the death of the child.

Some families however, may not wish to discuss funerals prior to the child's death and will discuss this at the time it is required.

Funeral costs can vary significantly and it can be helpful for the family to talk this through with one of the team members. Financial assistance to assist families in arranging the funeral may be available and should be considered with appropriate social work assessment and supports.

Funeral directors and ministers of religion or civil celebrants have the primary responsibility to assist a family through the important issues of planning and holding the funeral.

Families should be encouraged to do what best suits their individual needs and meets their cultural and spiritual beliefs. Some examples of decisions to consider:

- music
- cemetery plot or place for ashes
- photographs,
- releasing balloons, doves or butterflies or planting a tree
- readings
- eulogies
- inclusion of siblings
- open or private ceremony
- memorial card
- pallbearers
- public notices
- inviting particular people to participate in special ways.



An issue that is often raised is whether or not children should attend funerals. The answer should be individualised but the general rule is that attending the funeral is helpful for a child if they are given appropriate preparation and support. It is also helpful for some children to be involved directly with the funeral process. This might include choosing the clothes for the child to wear, decorating the coffin, choosing music, writing or drawing something to place in the coffin with the child, or saying something at the funeral service in the case of older siblings or close friends.

Children should never be forced to attend, though attendance often allows the child to feel included in this important family and community event and helps prevent misunderstandings about what has happened to the deceased child's body. Preparation for a child to attend the funeral that is helpful includes:

- Allowing the child to help plan the funeral if appropriate.
- Enabling the child to ask questions about what will happen.
- Giving the child accurate information about what to expect.

It is especially important to prepare children for the likelihood that many people, including their mum and dad, will be very sad and might cry (but that it is alright to cry and they will be okay). It is also important to tell them what will happen to the coffin with the child's body in it (e.g. it will be put into the ground or curtains will close around it at the end of the funeral so it cannot be seen and then later it will be cremated).

It is essential to apply principles of good communication with children when preparing to attend a funeral. It is especially important to listen to their questions such as *"Why is he put into the ground?"*, *"Can she feel anything?"*, *"What if he's not dead?"* and *"What will her ashes look like?"*.

These questions need honest, simple, accurate and sometimes repeated answers given in an atmosphere of care and nurture for the grieving child. Advise parents to remember to use reflective questions rather than assuming the answer that is required, as children's questions may have more than one answer. An example of a reflective question is: *"Tell me what you think and then I can tell you what I know?"* It can be a process which is quite distressing for families, especially if they have not been prepared, but one which is essential for children.

GRIEF AND ANTICIPATORY GRIEF

A simple working definition of grief is that it is the cluster of thoughts, emotions, behaviours and experiences that are related to a loss.

Anticipatory grief is the grief that is associated with an impending or expected loss. It is a significant form of grief within palliative care. Many families will experience grief with impact of the illness and impending death of their child, and also with the loss of 'normal family life' as it was before diagnosis. The loss of a child also presents unique issues of grief for parents as they face their inability to protect their child, and the loss of their hopes and dreams for that child's future.

The sick child will also be experiencing grief as they experience the losses associated with their illness. These include loss of their healthy self and ability to do things independently, loss of 'normal life' and routine activities such as going to school, playing sport, going out with friends, and consequently a sense of isolation as many relationships change and diminish. They may also experience the loss of hopes and dreams for their future, particularly for adolescents.

Grief is a natural process through which people adapt to the changes in their lives which involve loss. While many changes involve losses at levels that are not of major concern, losses associated with the death of a loved one are of enormous significance and bring with them powerful and distressing expressions of thought, emotion, behaviour and experience.

Thankfully, grief has become an issue that society is now more willing to face than it has in the past. However, many people find it difficult to know how to communicate with a bereaved family or a family that is facing an imminent bereavement. Consequently such families often find that there may seem to be general avoidance of them or of the issue that is most dominant in their lives (the death or palliative care of their child).

Having a clear framework for understanding grief and anticipatory grief will enable effective work with families. Importantly, a framework will not only guide work with families but can also be used when working with families' support networks. By sharing an appropriate understanding of grief and anticipatory grief, the anxieties and concerns of a family's support network can be reduced, enabling them to be of greater assistance to the family.

Tasks of grief and anticipatory grief

There is an extremely broad range of normal and adaptive grief reactions, including a depth of emotional and existential pain that is difficult for most people to observe, and conversely, avoidance and suppression of emotion that can be hard to understand. However, it is important to be able to view the big picture and not be too quick to judge a family's expressions of grief as pathological.

Importantly, health care professionals must realise that it is not their role to eliminate experiences and expressions of emotional pain in the context of grief or anticipatory grief. Such a role would be impossible and any attempt to achieve it would deny the reality of the family's experiences.

William Worden provides just one framework for understanding the process of grief that focuses on the “tasks” which grieving people need to achieve.¹⁰⁶ The tasks that he identifies are:

1. To accept the reality of the loss.
2. To work through the pain of grief.
3. To adjust to the environment in which the deceased is missing.
4. To emotionally relocate the deceased and integrate the loss into their ongoing lives.

There are many additional frameworks available and parents/caregivers should be directed to them.

Clearly tasks 1 and 2 are tasks which also exist in the anticipatory grief phase. Task 3 takes place in anticipatory grief, primarily in terms of adjusting to the changing environment as the disease progresses, and in terms of preparations for the changes that will occur once the child has died. In the anticipatory grief phase, a fifth task of “identifying and pursuing quality of life” should be added.

This framework also identifies the following goals of providing psychosocial support to a family experiencing anticipatory grief:

- Facilitating understanding of the reality of the loss that is faced.
- Enabling expression of and exploration of feelings.
- Helping overcome blocks to adjustment in the changing physical and emotional environment.
- Assisting in the identification and pursuit of quality of life.

It is not the health professional’s role to achieve these tasks, but to gently facilitate the family’s movement towards them. This occurs as the family faces, and gradually works on, the tasks in the process of caring for a child receiving palliative care. They are tasks which extend into and blend with tasks of grieving. As health professionals we must be informed by the family’s own interpretation of their experience.

For example, health professionals may feel a family is “in denial” about the seriousness of their child’s condition – but a family may simply have developed a way of life that allows them to live day to day. Families may appear to be planning for the future and ignoring the immediate situation, but when this is explored more deeply parents may say *“I have to keep some hope alive, I can’t keep hearing my child’s bad news all the time”*. Similarly they may view “hope” in terms of a “pain free day”, or a day *“when we didn’t have to think about the future”*.

Importantly, in palliative care and in grief work, psychosocial support is not limited to formal psychosocial interventions such as formal counselling or therapy, but extends into every interaction with a family and its social network. The skill that is required is to see how each interaction can assist the family’s movement towards achievement of key tasks from the framework identified above.

BEREAVEMENT SUPPORT



A child's death has a profound lifelong impact on a family and they need recognition, acceptance and validation of their experience. Families are constantly reminded of what could have been had their child survived. Seeing other children playing, commencing school, starting high school, graduations, 21st birthdays, marriages and other events that mark the passage of life, serve as reminders of what could have been, and often bring about renewed pain.

It is important to understand that all people, adults and children, react to the death of a family member differently. Health professionals develop relationships with the dying child and the family. This also requires acknowledgement of the personal impact of caring for the family. As professionals who have known the family and the child who has died, we can assist families to acknowledge and live with their experiences. This can be achieved by being available to listen to their feelings of helplessness, anger, guilt, regret, relief and sadness, and by exploring lost hopes, dreams and their search for meaning.

Throughout the course of their child's life, parents will have developed important relationships with staff and other families in the health care setting. Parents may wish to revisit the hospital and talk with people who knew them and their child, and shared a part of their journey.

It is helpful for the treating medical practitioner to offer to meet with the family in the weeks or months after the child's death. This provides an opportunity for parents and siblings to reflect on their experience, and to ask questions that may help to clarify some of the events that occurred during treatment and in the terminal phase of their child's life. Some families will receive this offer appreciatively whilst other families may not wish to accept the offer at all.

As health professionals, it is important to listen carefully to bereaved parents and other family members. Gratuitous and possibly insensitive general advice is never helpful. Each family's grief is a unique and deeply personal experience requiring the flexible support from carers. It is helpful to avoid some words and phrases such as "moving on", "closure" and "we know how you feel". Once again listening is often more healing than talking.

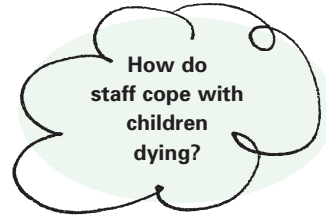
A real fear for some parents is that their child will be forgotten. Phone calls to families at significant times such as the child's birthday, anniversary of death and other special times are appreciated. Health professionals are at times reluctant to contact families for fear of making things worse. However, the child who has died will always be in the thoughts of their family, and knowing that others remember is important and comforting.

Siblings of children who have died may also have their own fears or misconceptions of the experience. It is important to include them in conversations and to help them create memories for the present time and into the future. A memory book created by the sibling to keep can sometimes help to alleviate this fear. It also provides an opportunity to say goodbye creatively and to explore feelings through the use of drawings, photos and writings.

It is important to remember that children's expression of their grief after a death can appear to be less intense and persistent than for adults. Siblings and other children can often be quite concrete and matter-of-fact discussing the child's death and grieving parents can easily misinterpret this. Parents will require sensitive reassurance at these times that such behaviours are quite normal. Children may need to revisit their understanding of their brother or sister's death and the emotional impact of this on themselves as they grow older.

Families will experience varying degrees of emotional and physical pain in the months and years following the death of their child. It is important for health professionals to know what local bereavement services are available for families who wish to access them. Different models for providing bereavement support include individual, couple and family counselling and bereavement support groups. Each person is unique and should be encouraged to access the type of support that suits them best.

SUPPORTING THE STAFF



The death of a child has an impact on staff at both a professional and personal level. Working in a palliative care setting, the death may be perceived by staff as 'good' or 'bad', with each child's death having a different impact on individual staff. As health professionals we have our own expectations of 'what a good death should look like'. These expectations may have been influenced by experiences in our personal lives. The relationships which have formed, perhaps during the more active treatment phases, can also impact on the experience for individual staff members or teams. It may be challenging for a staff member to maintain the professional boundary of relationships when they relate to a family more closely than others.

In order to maintain the staff's own health and wellbeing, as well as sustaining the quality of the service provided, staff support, or "care for the carer", is an important consideration.

A model for supporting staff might include the following elements:

- Psycho-education – the provision of information on the impact of stress and trauma.
- Training in coping skills and strategies.
- Structured systems of workplace support including opportunities to reflect on specific events and the potential for their personal impact.

Continued exposure to events such as children dying, without acknowledgement of its impact, can lead to a reaction that could be considered 'burn out' or 'vicarious trauma'.

Burn out can be described as the accumulated experience of working in an environment that impacts upon your emotional wellbeing, the personalisation of your work and any sense of accomplishment in the work environment.

By contrast, vicarious trauma may result from the experience of working directly with the child, and the 'trauma' associated with witnessing their care and its impact on them and their family.

Possible responses to these experiences can be characterised by:

- Strong emotions such as fear, anger, helplessness, sadness.
- A change in cognitive processes such as memory, attention, and concentration.
- Behaviours such as irritability, social withdrawal and self-medication.

Potential strategies which may be useful in managing these responses can be organised into personal and workplace strategies.

Personal strategies

For the individual, a strategy which is generally manageable by staff is to effectively develop self-care features into their day to day lives. An adequate diet, sleep and exercise are all important for general fitness and in promoting resilience. Finding a balance in life between work and "non-work" is also very important and is an effective strategy for health professional's self-care in the longer term.

Learning specific strategies for managing the stress caused by exposure to trauma can also give staff a greater sense of control. These specific skills include: identifying and challenging negative thinking, time management skills, problem solving techniques and relaxation training.

There is not a "one size fits all" approach to self-care and as such it is important to explore all options that may meet the needs of the individual staff member's life-style. Quite simply one needs to "work" at their self-care.

Workplace strategies

It is important that the individual work unit where a child dies should have in place a range of options for "caring for the carer" and these need to suit the diversity of staff responses to their workplace experiences. All staff within the work unit, whether they are working in clinical or operational streams can be affected when a child within their care dies. Where possible staff should avail themselves of any structured systems of trauma support that exist in the workplace. Of course, these also need to meet the needs of the individual staff member. Broadly these workplace strategies can be broken down into proactive or reactive approaches within operational and psychological categories (see Table 17).

BEREAVEMENT

TABLE 17 Workplace strategies for staff support

<i>Intervention level</i>	<i>Pre-Event (Proactive)</i>	<i>Post-Event (Reactive)</i>
Operational	Policy/procedure	Operational debriefing
Psychological	Psycho-education Peer support training	Peer support Psychological debriefing

Peer support strategies involve training staff in workplaces to manage the effects of the traumatic events to which they are exposed. This may involve developing skills to more effectively support each other and, where appropriate, refer to outside supports.

Operational debriefings focus more directly on the specifics of the care provided and are also a useful strategy. The focus of these sessions is to consider the care provided, namely what was done, by whom, and with what resources. This type of debrief is generally undertaken by appropriately trained line managers or supervisors and considers what additional learning can be gained from reflecting on the type of care provided.

Psychological debriefs can occur when the peer support resources of a specific workgroup are exceeded. These forums may involve follow-up of the group and/or individuals as required. The opportunity for a more 'supportive reflection' of the experience may be useful to assist staff to incorporate the experience into their paradigm of practice. An example of this is a specific memorial activity which is organised by the work facility and provides the opportunity within a supportive, public forum to share individual emotional experiences.

It is suggested that staff familiarise themselves with services for employee assistance within their specific workplaces. For Queensland Health employees, "Employee Assistance Services" can be contacted on **1300 361 008** (24/7) or enquiries can be emailed to EAP@health.qld.gov.au

A health system should seek to make improvements in six areas or dimensions of quality, which are listed below.

These dimensions require that health care is:

- **Effective** – delivering health care that is adherent to an evidence base and results in improved health outcomes for individuals and communities, based on need.
- **Efficient** – delivering health care in a manner which maximizes resource use and avoids waste.
- **Accessible** – delivering health care that is timely, geographically reasonable, and provided in a setting where skills and resources are appropriate to medical need.
- **Acceptable/patient-centred** – delivering health care which takes into account the preferences and aspirations of individual service users and the cultures of their communities.
- **Equitable** – delivering health care which does not vary in quality because of personal characteristics such as gender, race, ethnicity, geographical location, or socioeconomic status.
- **Safe** – delivering health care which minimizes risks and harm to service users.¹⁰⁷

Most paediatric palliative care services and hospices are involved in quality improvement (QI) activities. This involves changes in practice or care that lead to better care and outcomes for patients and families. Implementing evidence based practice into routine day to day care is a key objective of quality improvement. Health services require a strong culture of quality, and also a willingness to collaborate with other services within this context. “The Standards for Providing Palliative Care for all Australians” has been established to define the standard of care that will best meet the needs of dying patients and their caregivers (see appendix 5).

These standards incorporate the unique needs of dying children and their families.

Activities that can assist with quality and service improvement include:

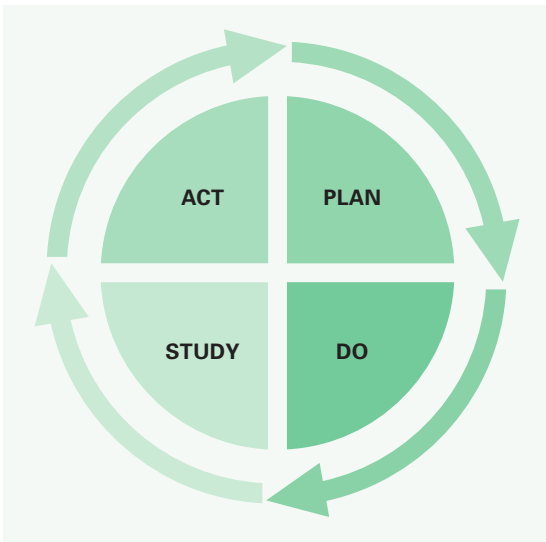
- Clinical governance.
- Development of policy, procedures and guidelines related to care.
- Audits of practice and documentation.
- Analysis of clinical and critical incidents
- Routine review of the care of patients after they die (i.e. death reviews, mortality and morbidity meetings).
- Incorporating child and parent feedback into how a service operates (both families currently caring for a child with a life limiting condition, and also bereaved parents and families).
- Monitoring clinical indicators (see appendix 6).
- Self-assessment programs (e.g. National Standards Assessment Program/NSAP).
- Peer review and mentorship (e.g. NSAP).
- Collaborative projects (e.g. through the Australian and New Zealand Paediatric Palliative Care Reference Group and NSAP).

The ‘plan, do, study, act’ (PDSA) cycle¹⁰⁸ uses simple measurements to monitor the effects of change over time. Changes and improvements do not need to be large and can be implemented over small time frames (weeks to months). Larger improvements occur through successive cycles which become linked cumulatively to each other and across organisations. Reflective practice and building on prior learning and experience also assist this process. Such change can be based on existing ideas and research, or through practical ideas that have been proven to work elsewhere.¹⁰⁹

Within the paediatric context, family centred care is also important in the context of patient or child centred care. Family centred care can be achieved by:

- Better communication with parents and carers.
- Treating all family members with dignity and respect.
- Embracing the family as part of our team of carers.
- Providing prompt feedback.
- Accepting a joint decision-making process
- Enabling carers with the skills to provide ongoing care.
- Improving overnight carer accommodation.¹¹⁰

FIGURE 10 PDSA Cycle ¹⁰⁸

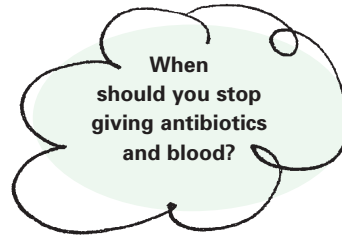


Palliative care is an active branch of medicine and as such is governed by the ethical standards that apply to all members of the health care professions involved. There are particular issues and rights that pertain to both palliative care and paediatrics.

Palliative care, including pain and symptom management, can be seen as a basic human right. In the United Kingdom, *Together for Short Lives* has developed a charter listing the rights of dying children and their families.⁸ Similar to standards, the charter is a useful tool for professionals striving to deliver optimal paediatric palliative care. Other relevant documents in this context include “The Korea Declaration”, “The Joint Declaration and Statement of Commitment on Palliative Care and Pain Management as Human Rights”, the “Declaration of Montreal” and the “UN Convention on the Rights of the Child”.^{111, 112}

EXTENT OF SUPPORTIVE CARE

The extent of treatment that should be given to children in the palliative or terminal phase of their illness often raises ethical concerns for health professionals. It is important to consider the intent of therapeutic interventions to judge the ethics of giving or withholding a particular treatment. If the intent of the intervention is to prevent the child from dying, when death is inevitable, then it is ethically acceptable to withhold the treatment. **If the intent is to control symptoms, however, then all interventions need to be considered in the context of the current clinical goals for each patient.**



ADVANCED CARE PLANS

The concept of Allow Natural Death (AND) offers a new conversation for families and medical teams as they navigate the difficult terrain of end of life decision making. Resuscitation orders and policies around resuscitation status have long been associated with Do Not Resuscitate (DNR) or Not for Resuscitation orders (NFR). They allowed an opportunity to have an open and frank discussion about end of life decision-making. However, DNR and NFR discussions can be negatively perceived by the family as they focus on what should **not** be done rather than what can be done. The AND conversation addresses these issues from the perspective of the child and family’s quality of life and what is in the child and family’s best interest. The AND conversation focuses on what can be done at the end of life and what can be added to the process rather than solely on what is to be withdrawn. Conversations about allowing a natural death may open up more opportunities to explore and manage a good death. The Paediatric Acute Resuscitation Plan (PARP) is another tool which emphasizes symptom management, privacy and dignity during end of life care.

Decision making at the end of life requires a collaborative approach that encompasses the wishes of the child, and the knowledge and

expertise of the family and medical team. It is important for parents to retain control over decisions relating to their child's life and death and to consent to interventions. Even quite young children will have a view on treatments offered and their wishes should be part of the decision making process. Discussions about allowing a natural death can help to focus on the activities and interventions that might lead to a good death.

Ideally decisions on the extent of intervention can be made ahead of time and written into a plan for the family. These may include both positive and negative instructions, for example *"I wish to receive platelet transfusions"*, and *"I do not wish to receive antibiotics"*, or *"No more blood tests"*. These directives may include the extent of resuscitation and should also include spiritual and social aspects of care. It is vital that these directives are clear to all involved in the child's care and are made with the child's parents. Hospital policy will vary as to how these will be documented for inpatients. Advanced care plans including AND directives will alter as the condition of the child changes and it is essential that they are regularly reviewed and dated (Appendix 1).

However, some families are unable to make these difficult decisions ahead of time and it is vital that a flexible and patient-centred approach is taken.

DOUBLE EFFECT OF DRUGS

In the control of pain the dose of opioid required might be close to the level that depresses respiration, raising concerns that it may hasten the time of death. This is usually only in the terminal phase of the illness when the child often requires sedative drugs in addition to opioids. This differs from euthanasia in that the intent is to control distressing symptoms of pain and agitation, not to cause death.

CONFIDENTIALITY, PRIVACY AND DISCLOSURE

Palliative care is a specialty that requires a multidisciplinary team approach to optimise care. The ethical principle of confidentiality of information given to one member of the team is sometimes in conflict with the functioning of a large team. It is important to consider the intent of disclosing information; if it will have a significant impact on the interaction of another team member with the patient it is ethically reasonable to share the information, if it is "of interest only" it is not. The problems relating to confidentiality can often be avoided if the need for sharing of information is discussed with the family.

EUTHANASIA

Euthanasia is the deliberate and painless termination of life of a person afflicted with an incurable disease leading inevitably to death. Allowing a person to die differs from euthanasia in intent, in act and in professional mandate.³ The intent in euthanasia is to cause death immediately. The intent in discontinuing life prolonging treatment is to cease hindering an inevitable process from reaching its timely end.¹³ While there are both vehement supporters and opponents in the community, the practice of euthanasia is illegal in Australia and New Zealand.

Parents and other family members will sometimes ask about euthanasia directly or indirectly. Whilst it is correct to state the legal position, it is more important to find out the parents' real concerns and their understanding of what euthanasia is. The parents' concerns for their child's suffering can almost always be dealt with by good palliative care. It may also be that the parents' understanding of euthanasia is different from the standard medical one, and includes concerns about the need for resuscitation and limitation of pain relief due to the double effect of analgesia.

RECOMMENDED BOOKS FOR FURTHER READING

Listed in the following pages are books that may be helpful to health professionals and teachers wishing to increase their knowledge on paediatric palliative care issues. There is also a list of storybooks that may be useful in assisting children to express their feelings associated with loss and grief. These books are only a sample of the literature available and you may wish to look further for suitable resources.

Before recommending books to a family read the book first. Do you think the child will understand the concepts in the book? Who has died or is dying? Is it the child themselves, a sibling, parent, friend or pet? Should the book be about an animal or a person? Is the main character of the story someone with whom the child could identify?

Further information on these topics and book loans may be available through the local children's hospital, palliative care or hospice.

Books for young children

Gentle Willow A Story for Children about Dying

Joyce Mills and Michael Chesworth.
Magination Press: New York, (1994).
(5 to 8 years)

I Had a Friend Named Peter – Talking to children about the death of a friend

Janice Cohn and Gail Owens.
Morrow Junior Books: New York, (1987).

Beginnings and Endings with Lifetimes in Between

Bryan Mellonie & Robert Ingpen.
Puffin Books: Melbourne, (2005).
(3 to 6 years)

How I Feel

Alan Wolfelt

Goodbye Forever

Boulden Publishing

Saying Goodbye

Boulden Publishing

When Someone Very Special Dies

Marge Heegaard

The next place

Warren Hanson. Waldman House Press (1997).

When dinosaurs die:

A guide to understanding death

Laurene Krasny Brown & Marc Tolon Brown.
Demco Media (1998).

I miss you: A first look at death

Pat Thomas. Barron's Educational Series (2001).

The invisible string

Patrice Karst. DeVross Publications (2000).

Books for older children and teenagers

How Teenagers Cope with Grief – Something I've never felt before

Doris Zagdanski.
Hill of Content: Melbourne, (1990).

The Grieving Teen

Helen Fitzgerald

Straight Talk about Death for Teenagers

Earl Grollman. Beacon Press: Boston, (1993).

Thoughts – A teenager's response to a crisis

Darren Crewe.
Ronald McDonald House: Melbourne, (1997).

The grief book: strategies for young people

Elizabeth Vercoe & Kerry Abramowski.
Black Dog Books, (2004).

RESOURCES

When a friend dies: A book for teens about grieving and healing

E. Gootman & Pamela Espeland.
Turtleback Books, (2005).

10 pack journals: What on earth do you do when someone dies?

Trevor Romain Company, (2008).

Books for siblings

Two weeks with the Queen

Morris Gleitzman.
Pan Macmillan: Sydney, (1989).

Where's Jess

Joy and Mary Johnson
A short simple story to help children ages 2–5 years cope with infant sibling loss.

Am I Still a Sister?

Alicia Sims
An 11 year old girl tells the story about her brother who died from brain cancer.

Books for parents

Shelter from the Storm – Caring for a child with a Life-Threatening Condition

Joanne Hilden and Daniel Tobin.
Da Capo Press: Cambridge, (2003).

Living with a seriously ill child

Jan Aldridge. Sheldon Press: London, (2007)

A Child Dies: A Portrait of Family Grief

Joan Hagan Arnold & Penelope Buschman.
The Charles Press Publishers: Philadelphia, (1994).

Are You Sad Too? Helping Children Deal with Loss and Death

Dinah Seibert, Judy Drolet and Joyce Fetro.
ETR Associates: California, (1993).

Talking About Death – A Dialogue between Parent and Child

Earl Grollman. Boston: Beacon Press, (1990).

After the Death of a Child: Living with Loss through the Years

Ann Finkbeiner
A comforting book that examines the continuing love a parent feels for their child and ways to preserve that bond even after death.

The Worst Loss: How families heal from the death of a child

Barbara Rosof
A practical book by a child psycho-therapist who has worked with bereaved families.

The Bereaved Parent

Harriet Sarnoff Schiff New York
Crown Publishers Inc., (1977).
A classic book written by a bereaved mother for parents whose child has died and for all who want to help them.

Coping with grief

Mal McKissock & Dianne McKissock. ABC Books for the Australian Broadcasting Corporation (1995).

35 ways to help a grieving child

Amy R. Barrett & Dougy Centre for Grieving Children.
Dougy Centre for Grieving Children (1999).

Beyond Words – Grieving when your child has died

Andrew Thompson & Tricia Irving Hendry.
Skylight: New Zealand, (2012).

Books for grandparents

Grandparents cry twice: Help for bereaved grandparents

Mary Loud Reed.
Baywood Publishing Company (2000).

When a grandchild dies:

What to do, what to say, how to cope

Nadine Galinsky

Gal in Sky Publishing Company (1999).

Books for professionals

Teaching children with life-limiting illnesses: a manual for schools

The Children's Hospital at Westmead

Grief in children: a handbook for adults

Atle Dyregrov

Jessica Kingsley Publishers, London, (2008).

Loss, change and bereavement in palliative care

Pam Firth, Gill Luff, David Oliviere

Open University Press, (2005).

Parents and bereavement: a personal and professional exploration of grief

Christine Young and Tracy Dowling

Oxford University Press, (2012).

Websites where books and other resources can be sourced

www.innovativeresources.org

St Luke's provides popular resources for working with children and teens, and lists books that suit children, teens, parents and professionals.

<http://www.skylight.org.nz/>

Skylight support people facing any kind of tough life situation of change, loss, trauma or grief – whatever the cause and whatever their age. They also assist those wanting to help them.

<http://www.openleaves.com.au/>

Open Leaves Books is a bookseller in the areas of counselling, therapy, loss and grief.

LIST OF ORGANISATIONS AND WEBSITES

Listed below are organisations and websites that may be useful to families and professionals working with them. The organisations listed are particularly relevant to Queensland, Australia. Similar organisations should be available for families in other states, territories and countries.

Information and advocacy organisations

CareSearch Palliative Care Knowledge Network

CareSearch is an online resource designed to help those needing relevant and evidenced based information and resources about palliative care.

There are sections designed specifically for health professionals and others for patients, for carers, and for family and friends.

<http://www.caresearch.com.au/>

Palliative Care Australia

Tel: 1800 660 055

Website: <http://www.pallcare.org.au>

This site has online access to the Department of Health & Ageing's Palliative Care resource "Journeys", "Spiritual Care", "National Palliative Care Service Directory", "Standards for Providing Quality Palliative Care for All Australians" and "Palliative Care & Primary Care – an overview of selected resources to support Primary Care Providers in Palliative Care."

Palliative Care Queensland

Tel: 1800 660 055

Website: <http://www.palliativecareqld.org.au/>

The Palliative Care Information Service Qld

Tel: 1800 772 273

Email: info@pcis.org.au

<http://www.pcis.org.au/>

International organisations

Children's Hospice International

<http://www.chionline.org/>

Together for Short Lives

<http://www.togetherforshortlives.org.uk/>

Children's Project on Palliative/Hospice Services (ChiPPS)

<http://www.nhpco.org/resources/pediatric-hospice-and-palliative-care>

International Children's Palliative Care network (ICPCN)

<http://www.icpcn.org/>

World Health Organisation – Cancer pain and palliative care program

<http://www.who.int/cancer/palliative/en/>

Children and family support

I Give a Buck

This charity assists families caring for children with life-threatening or life-changing illness or condition. Assistance can be in the form of emergency funding, equipment, early intervention and the provision of iPads.

<http://www.igiveabuck.org.au/>

Make A Wish Foundation of Australia

Make-A-Wish grants wishes to children between the ages of three and 18, who have been diagnosed with a life-threatening medical condition.

Toll free Ph: 1800 032 260 for your nearest office

<http://www.makeawish.org.au>

Queensland Kids

Queensland Kids is a not-for-profit charity with the vision of creating an innovative and sustainable children's hospice and respite facility addressing a significant gap in Queensland's paediatric landscape.

Tel: 07 3040 2510

<http://www.hummingbirdhouse.org.au>

Smile Foundation

SMILE provides financial assistance and case management services to affected families and supports research relating to rare childhood medical conditions.

<http://www.smilefoundation.com.au/>

Starlight Children's Foundation

Starlight provides programs integral to the total care of seriously ill children with the aim of lifting the spirits of the child, giving them the opportunity to laugh, play and be a child again. This includes "A Starlight Wish" which gives the whole family a break from the stress of their child's illness. Fulfilled wishes provide lasting memories for the child and family. "Livewire" is dedicated to meeting the needs of adolescents (and their siblings) living with serious illness, a chronic health condition or disability.

Livewire.org.au provides a safe and supportive online community for 10–20 year olds where members can connect and share experiences with others who understand what they are going through.

Tel: 1300 727 824 for your nearest office

<http://www.starlight.org.au>

The Steve Waugh Foundation

The Steve Waugh Foundation is working to help change things for children with a rare disease by giving hope, providing medicine, equipment and treatment, supporting education and research, partnering with other like agencies and organisations as well as supporting specific projects and programs.

<http://www.stevewaughfoundation.com.au/>

TLC for Kids

TLC for kids is a National charity that supports sick children and their families through the moments in hospital where fear and distress can take over. Their distraction services facilitate kids through frightening procedures, and in the crucial 48 hour following any emotionally challenging situation in hospital.

Tel: 1300 361 461

<http://www.tlcforkids.org.au/>

Variety Australia

Empowering Australian children who are sick, disadvantaged or have special needs to live, laugh and learn. By giving practical equipment, programs and experiences.

<http://www.variety.org.au/>

Children's hospice services

Bear Cottage

Manly, Sydney, New South Wales

Tel: (02) 9976 8300

<http://www.bearcottage.chw.edu.au/>

Very Special Kids

Malvern, Melbourne, Victoria

Tel: 1800 888 875

<http://www.vsk.org.au/>

Clinical support organisations

(some of these organisations are able to provide respite/"short breaks")

Better Start

The Better Start for Children with Disability initiative provides funding for early intervention services. Children with an eligible diagnosis must register for Better Start before they turn six years old. A child will have until they turn seven to access funding. Services that can be provided include: audiology, occupational therapy, orthotics, physiotherapy, psychology and speech pathology.

It is a national program (administered by the Department of Families, Housing, Community Services and Indigenous Affairs).

Tel: 1800 242 636

<http://www.betterstart.net.au/>

Centacare

<http://www.centacarebrisbane.net.au/>

Commonwealth Respite and Carelink Centres

Tel: 1800 052 222

<http://www9.health.gov.au/ccsd/>

Disability Services Queensland

Tel: 13 74 68

<http://www.communities.qld.gov.au/disability/>

FSG Australia

<http://www.fsg.org.au/>

Mamre Association

<http://www.mamre.org.au/>

Montrose Access

<http://www.montroseaccess.org.au/>

Paradise Kids

<http://www.paradisekids.org.au/>

St Vincent's Hospital

<http://svphb.org.au/>

Xavier Children's Support Network

<http://www.xcsn.org/>

Community and palliative care nursing organisations

Anglicare

Tel: 1300 785 853

<http://www.anglicaresq.org.au/>

Bluecare

Tel: 07 3377 3377

<http://www.bluecare.org.au/>

RESOURCES

Cittimani Hospice Service

Tel: 07 5445 0822

<http://www.cittamanihospice.com.au/>

Karuna

Tel: 07 3632 8300

<http://www.karuna.org.au/>

Ozcare

Tel: 1800 692 273

<http://www.ozcare.org.au/>

Cancer specific organisations

Australia and New Zealand Haematology and Oncology Group

<http://www.anzchog.org/>

Brainchild Foundation

Tel: 1300 272 462

<http://www.brainchild.org.au/>

Camp Quality

Tel: 1300 662 267

<http://www.campquality.org.au/>

Canteen

Tel: 1800 226 833

<http://www.canteen.org.au/>

The Cancer Council Australia

Tel: (02) 8063 4100

<http://www.cancer.org.au/>

Cancer Council Queensland

Ph: 13 11 20

<http://www.cancerqld.org.au/>

Childhood Cancer Support

Ph: 07 3252 4719

<http://www.ccs.org.au/>

Leukaemia Foundation of Queensland

Ph: 07 3318 4418

<http://www.leukaemia.org.au/>

National Cancer Institute

<http://www.cancer.gov/>

Oncolink (resources for children and families)

<http://www.oncolink.org/>

Redkite

Tel: 1800 334 771

www.redkite.org.au

Non-cancer disease specific organisations

Australian Leukodystrophy Support Group

Tel: 1800 141 400

<http://alds.org.au/>

Cerebral Palsy League

Tel: 1800 275 753

<http://www.cpl.org.au/>

Cystic Fibrosis Australia

<https://www.cysticfibrosis.org.au/>

DEBRA: Epidermolysis Bullosa

<http://www.debra.org.au/>

Epilepsy Australia

Tel: 1300 852 853

<http://www.epilepsyaustralia.net/>

Heartkids

<http://www.heartkids.org.au/>

Australian Mitochondrial Disease Foundation

<http://www.amdf.org.au/>

Mucopolysaccharide and Related Diseases Society

<http://www.mpssociety.org.au/>

Muscular Dystrophy Australia

<http://www.mda.org.au/>

Short Gut Syndrome Families' Support Group

<http://www.shortgutsupport.com/>

Spinal Muscular Association of Australia

<http://www.smaaustralia.org.au/>

Bereavement support

Australian Centre for Grief and Bereavement

This organisation's website provides a range of resources about grief and how to support grieving children, adolescents and adults. It also has a collection of stories, narratives, poems and creative expressions contributed by bereaved individuals who want to share insights from their own grief journey.
<http://www.grief.org.au>

Australian Child & Adolescent Trauma, Loss & Grief Network

This website has key resources that can assist adults in understanding the impact of trauma, loss and grief experiences on children and young people, and how to best help young people who have been affected by grief.
<http://www.earlytraumagrief.anu.edu.au/>

Bereavement Care Centre

The Bereavement C.A.R.E. Centre was established at Randwick in 1981 and in Lewisham in 1986 by Mal and Dianne McKissock. The Centre's goals are to provide comprehensive and accessible counselling and support services for the terminally ill and their families, and for those recently bereaved (i.e. approximately the first two years after the death of someone with whom they have a significant relationship).
<http://www.bereavementcare.com.au/>

Centrelink

Centrelink offers a Bereavement Payment that may be available to assist families adjust to changed financial circumstances after the death of a partner, child, or someone they cared for.

To be eligible for a Bereavement Payment families need to be eligible for other payments from Centrelink or the Department of Veterans' Affairs. The Bereavement Payment is usually paid as a lump sum.

Centrelink also has Social Workers who may be able to assist with claiming payments, counselling and support during times of grief.
Tel: 136 150
<http://www.humanservices.gov.au/families>

Center for Good Mourning

The Center for Good Mourning provides support and assistance given to bereaved children and families in Arkansas through education, program development and grief support programs. Their website includes resources and a regular newsletter.
<http://www.archildrens.org/Services/Center-for-Good-Mourning.aspx>

Compassionate Friends

(Support for bereaved parents, siblings and grandparents)

The Compassionate Friends offers support in the grief and trauma which follows the death of a child at any age and from any cause. The Compassionate Friends offer a range of free services to bereaved parents and families including:

- Branches across Queensland
- Support group meetings
- Country support groups
- Telephone and email support
- Grief information and library
- Newsletters
- Annual Candlelight Remembrance Service
- Annual Grief and Loss Seminar

<http://www.tcfaustralia.org.au/>

The Dougy Centre

The Dougy Centre provides support in a safe place where children, teens, young adults, and their families grieving a death can share their experiences. Their website provides resources

for parents, teachers, and others helping children in grief.

<http://www.dougy.org/>

Medicare Better Access Initiative

The Better Access initiative provides Australians with access to mental health professionals through Medicare. In order to receive a Medicare rebate for mental health services, there must be a referral to a mental health professional by an appropriate medical practitioner (GP, Psychiatrist or Paediatrician). The doctor must first make an assessment that the services of a mental health practitioner are needed. The costs will vary depending on the length of the session and the fee being charged by the professional. www.health.gov.au/mentalhealth-betteraccess

Paradise Kids

Paradise Kids help children learn life-skills to deal with grief through death, loss or illness. This loss can include the death or approaching death of a parent, grandparent or sibling, or the child's own chronic life-limiting illness. Programs include grief and loss support services and a holiday house.

Tel: 07 5574 6853

<http://www.paradisekids.org.au/>

RD4U

RD4U ('road for you') is a website designed to support adolescents after the death of someone close to them. It is a space where young people can read about other people experiences of grief, share their thoughts and feelings on a message board, offer advice to others and get information about ideas for coping.

<http://www.rd4u.org.uk/>

Reachout.com

Reachout.com is Australia's leading online youth mental health service and provides an online support space for adolescents dealing with grief and loss.

The service offers:

- Expert information on how adolescents can tackle life's challenges and become mentally fit and resilient
- Real-life stories of grief and loss
- Forums
- Videos

<http://au.reachout.com/Tough-Times/Loss-and-grief>

SANDS

(Miscarriage, stillbirth, and newborn death support)

Sands aims to facilitate healthy grieving following the death of a baby through miscarriage, stillbirth, newborn death or termination for medical purposes.

<http://www.sands.org.au/>

SIDS and Kids Queensland

SIDS and Kids provide a range of services including:

- A 24 hour bereavement support line
- Face-to-face and telephone counselling for parents, siblings and extended family
- Workshops
- Support meetings
- Information kits and potential funeral funding assistance.

<http://www.sidsandkids.org/>

Skattle

Skattle offers a unique buddying support model to assist children, teenagers and their families who are experiencing difficulties with loss. This loss can include the death of a loved one and that associated with a medical diagnosis.

<http://skattle.org.au/>

Skylight

Skylight support people facing any kind of tough life situation of change, loss, trauma or grief – whatever the cause and whatever their age. They also assist those wanting to help them. New Zealand Tel: 0800 299 100 or 04 939 6767 <http://www.skylight.org.nz/>

Young Minds

Young Minds is a leading UK charity which provides information on improving the emotional wellbeing and mental health of children and young people. Their website provides resources that may be helpful in assisting your child/children during their grief. <http://www.youngminds.org.uk/>

Memory making

Heartfelt: giving the gift of photographic memories

Heartfelt is a volunteer organisation of professional photographers from all over Australia dedicated to giving the gift of photographic memories to families that have experienced stillbirths, premature births, or have children with serious and terminal illnesses. All services are provided free of charge. Tel: 1800 583 768 <http://www.heartfelt.org.au/>

Impressionable Kids

Create framed hand and feet sculptures. Tel: 1300 885 868 <http://www.impressionablekids.com.au/>

Smallprint

Create silver jewellery keepsakes (e.g. fingerprints or drawings). Tel: 1800 762 557 <http://www.smallprint.com/>

Journals

The American Journal of Hospice and Palliative Care

<http://ajh.sagepub.com/>

BMC Palliative Care

<http://www.biomedcentral.com/bmcpalliatcare/>

BMJ Supportive and Palliative Care

<http://spcare.bmj.com/>

Death Studies

<http://www.tandfonline.com/>

Grief Digest

<http://www.centering.org/>

International Journal of Palliative Nursing

<http://www.ijpn.co.uk/>

Journal of Hospice and Palliative Nursing

<http://journals.lww.com/jhpn/pages/default.aspx>

Journal of Pain and Symptom Management

<http://www.journals.elsevier.com/journal-of-pain-and-symptom-management/>

Journal of Pediatric Oncology Nursing

<http://jpo.sagepub.com/>

Journal of Palliative Medicine

<http://www.liebertpub.com/jpm>

Journal of Social Work in End of Life and Palliative Care

<http://www.tandfonline.com/>

Pain

<http://www.iasp-pain.org/>

Palliative Medicine

<http://pmj.sagepub.com/content/current>

Pediatric Blood and Cancer

<http://onlinelibrary.wiley.com>

RESOURCES

Local resources

This page has been designed for individual organisations to develop a local resource list. If you are aware of local health professionals with an interest and/or experience in palliative care, record their names here for future reference.

Paediatrician/s

Community Nursing Service/s

General Practitioner/s

Local Hospital (Paediatric Ward)

Local Hospice/Palliative Care group/s

Social Work Services

Other

APPENDIX 1

APPENDIX 1. PALLIATIVE CARE ACTION PLAN

Patient Sticker

Parents _____

Siblings _____

Other family _____

Accommodation _____

Treating team consultant _____ Nurse coordinator _____

G.P. _____ Paediatrician _____

Regional hospital _____ Contact _____

Domiciliary nurses _____ Contact _____

Social worker / Occupational therapist / Psychologist / Others

*Indicates the care coordinator

Diagnosis _____ Date of diagnosis _____

SIGNIFICANT PAST HISTORY

CURRENT STATUS

ADVANCED CARE PLANS

APPENDIX 1

<i>Date</i>	<i>Current symptoms</i>	<i>Management</i>

<i>Date</i>	<i>Potential symptoms</i>	<i>Management</i>

APPENDIX 2. GUIDELINES FOR CONTINUOUS SUBCUTANEOUS INFUSIONS

Saf-T-Intima Indwelling Catheter and NIKI T34 Pump*

* Some service may use alternative pumps

Purpose

1. To administer a continuous infusion of medications via an indwelling subcutaneous catheter.
2. To reduce the need for repeated injections.

Background information

While the route of choice for medications in the paediatric palliative care setting is enteral (e.g. oral, NG or PEG), this is not always possible. Many drugs are well absorbed subcutaneously and this route removes the need for intravenous infusions or intramuscular injections¹¹⁴ Continuous subcutaneous infusions of medications using a portable syringe driver can provide good symptom control. Combinations of drugs are sometimes used to control several symptoms. A significant advantage of continuous subcutaneous infusion is that plasma levels of drugs are more stable and good symptom control can be achieved without the peaks and troughs that result from intermittent drug administration.

At times it may be appropriate to use the child's Central Venous Access Device (central line or port) if present to administer medications by infusion.

Indications

When medications are unable to be given by another route, e.g:

- enteral or intravenous,
- due to nausea/vomiting, dysphagia, mal-absorption, gastro-intestinal obstruction,
- patient no longer responsive to enteral medications
- reduced level of consciousness/ severe weakness

Often used for patients receiving palliative care.

Key points

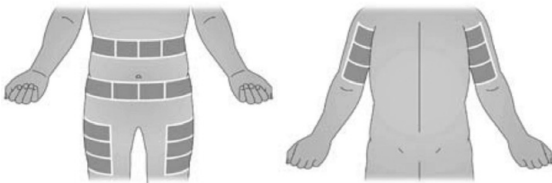
1. Use an aseptic technique to prepare and set up the infusion.
2. More than one drug can be mixed and administered in the same syringe. Where more than two drugs are required to manage multiple symptoms (such as pain, secretions and nausea) care must be taken to ensure compatibility, particularly with higher doses of midazolam. Check compatibility with a pharmacist. Common drugs delivered in combination are listed in Appendix 3. Dexamethasone often precipitates in an infusion, but can be given in bolus doses if clinically indicated. Consider the need for two subcutaneous devices if bolus medication is required or anticipated.
3. Normal saline or water for injection (WFI) are the preferred diluents for most medications. Dilute the drug as much as possible to reduce risk of irritation and preserve the insertion site. For infants or delicate sites, minimal volumes of 10ml are usually well tolerated.

4. Select the most appropriate injection site with adequate subcutaneous tissue (see Fig 1). Rotate the site with each insertion.
5. Catheter of choice is the **BD Saf-T-Intima 24** gauge with Y-port. This catheter can remain in place for as long as the site remains free of complications. After two weeks, consider rotation of the insertion site to avoid afterhours replacement or complications.
6. The **NIKI T34** portable syringe driver is suitable for continuous SC infusions. A loading dose of the medication (e.g. analgesia, anticonvulsant) may be required at the start of the infusion to ensure therapeutic drug levels are reached quickly.

Site selection

Preferred sites in children: abdomen; upper thigh; upper arm where there is more subcutaneous tissue.

FIGURE 1¹¹⁵



Avoid:

- Bony prominences, as there is less subcutaneous tissue and absorption will be reduced.
- Joints, as more likely to be uncomfortable and easier to dislodge.

- Oedematous, infected, broken, or bruised skin.
- Recently irradiated skin areas.
- Sites in contact with wheelchair harnesses or seatbelts

Equipment

- Topical anaesthetic cream (EMLA/LMX4) or ice if cream is not tolerated or available
- Saf-T-Intima 24g (yellow) indwelling catheter.
- NIKI-T34 infusion pump
- 10/20/30/50ml BD Plastipak luer lock syringe, or a 25ml Terumo luer lock syringe (No other syringe brands)
- Prescribed medication and diluent
- Minimum volume extension set
- Occlusive transparent dressing
- Non-sterile gloves
- Fixomull/hypafix

Additional equipment:

- 9 volt spare battery (non-rechargeable)
- ± Lock box and key

APPENDIX 2

1. Prepare Medication syringe

Procedure	Additional information
Prepare prescribed medication using a clean technique and in accordance with hospital or home care protocols.	
<p>Syringe size & dilution: Volume of undiluted, prepared drug(s): ≤10ml: 10ml syringe can be diluted with up to 10ml volume ≤20ml: 20ml syringe can be diluted with up to 18ml volume >20ml: 30ml syringe can be diluted with up to 22ml volume <50ml: 50ml syringe can be diluted with up to 33ml volume</p>	<p>Check compatibility of diluent as some drugs require WFI or 5% dextrose.</p> <p>Note – with 50ml syringe the volume will not be accommodated in the locked box.</p>
Attach extension set to the syringe and prime the line.	Keep end caps in place until ready to connect to the patient.
Label according to hospital protocol.	
If immediately commencing the infusion, prime the Intima once the extension tubing has been connected.	Always change the extension tubing at the syringe when the medication infusion order changes. The volume of the extension tubing is 2ml and therefore takes several hours to infuse.

APPENDIX 2

2. Insertion of Saf-T-Intima catheter

Procedure	Additional information
Explain procedure to child/parent and gain consent.	
Choose appropriate site and apply topical anaesthesia 45-60 minutes before procedure.	Ice applied for 5 minutes can also be used.
Remove topical anaesthesia with soap and water Perform Hand hygiene.	
Clean skin with 0.5-2% chlorhexidine in 70% alcohol or an alcowipe and allow to dry .	
Apply gloves (non-sterile).	
Pinch the skin between thumb and forefinger.	Ensures subcutaneous tissue is identified.
Grasp the Saf-T-intima wings with bobble-surface facing down towards patient. Bevel upright. Remove clear plastic needle cover.	
Insert at an angle of 20–45° to skin.	If blood is visible in the cannula, remove and insert a new cannula in another site – at least 3cm from previous site.
Hold the wings of the catheter firmly. Pull back on the introducer to reveal the needle encasement. Hold the Y-connector with one hand and gently pull the yellow encasement away with the other hand in one smooth single movement. Secure the catheter and wings only with a transparent dressing. Replace the injectable bung with a needle free bung (eg.Smartsite).	
Loop the extension set and secure with fixomull/ hypafix tape.	
Dispose of equipment, remove gloves and perform hand hygiene.	
Check all lines are secure.	

APPENDIX 2

3. Setting the NIKI T34 infusion pump

Prior to looking after a patient with a NIKI T34, all staff must have received instruction on how to operate this pump. Online training is available.

Procedure	Additional information
Check there is sufficient battery life to complete the infusion – change the battery if in doubt.	At least 40% battery life required for 24 hour infusion.
Connect the extension set to the available Y-connector of the Saf-T-Intima cannula.	
Switch the pump ON: Wait for the pump to pre-load.	The machine will calibrate itself.
Measure the drawn up syringe against the NIKI T34 and press either FORWARD or BACK to align actuator to the syringe plunger.	Do not attempt to manually move the actuator as this can damage the device.
The pump will display “LOAD SYRINGE”. Lift the barrel arm clamp and align the syringe in place with numbers facing operator. Lower the clamp.	If syringe not aligned correctly, the screen will alert which sensor is incorrect.
The Niki T34 will automatically detect which syringe is loaded. Use the ▲▼ keys to confirm syringe brand and size and press YES.	
Use the ▲▼ keys to change: Volume Duration Rate	
Confirm settings displayed on the screen and press YES. Start Infusion? Confirm YES.	
Press and hold INFO to lock/unlock keypad.	
Press NO/STOP prior to switching OFF.	

Observation and documentation:

- Refer to drug specific protocols for frequency of observations. These may be modified in the end-of-life setting. Discuss with the primary treating team or Palliative Care team if unsure.
- Site appearance: observe and document at least four hourly, monitoring for irritation, inflammation, excessive redness, tenderness, presence of a haematoma or leaking at the insertion site.
- Syringe/line contents appearance: absence of clouding, crystallisation or colour change.
- Monitor infusion delivery, including: infusion rate setting, volume remaining (correlate the volume remaining with the time remaining), volume infused.
- Check battery status (shown as a percentage on the Niki T34 display panel).
- Check parents know whom to contact if they experience problems with the infusion.
- Document any problem(s) noted and action taken.

At completion of infusion or when removing the subcutaneous infusion device:

- Ensure disposal of any remaining syringe contents per hospital/community nursing service protocols.

Adapted from Children and Adolescent Health Service (CAHS) Paediatric Nursing Practice Manual Section 2.5.8 Princess Margaret Hospital Perth Western Australia, 2012.

APPENDIX 3. SYRINGE DRIVERS AND DRUG COMPATIBILITIES

The combination of an analgesic and anxiolytic is the most common preparation, but an antiemetic and anticonvulsant are frequently also required. The requirements for each child will vary and the prescription should be assessed and written on an individual basis.

The following drugs can usually be combined in an infusion if indicated:

- Midazolam
- Morphine or other opioid
- Hyoscine hydrobromide or Glycopyrrolate
- Metoclopramide, cyclizine or ondansetron
- Haloperidol

Usually drugs are prepared in normal saline or water for injection. As the infusion rate is slow, discomfort is usually not a problem if drugs are mixed with water for injection. There is a tendency for precipitation to occur when more than one drug is used and when drugs are mixed with normal saline. For many infusions there is little diluent required to be added to the syringe as the volume may be complete with the drugs required.

Specific drug notes

Midazolam (0.2–1mg/kg/day) acts as a sedative and anticonvulsant agent and is suitable for an agitated or distressed child. Additional boluses can also be given.

Haloperidol (50–100µg/kg/day) can also be used for agitation and restlessness. It also has an antiemetic effect and is less sedating than midazolam. Extrapyramidal effects can occur however.

Metoclopramide can cause skin reactions, but is generally well tolerated, and is an effective antiemetic.

Cyclizine can also be used for severe nausea, but is only compatible with water for injection and there is no evidence of compatibility with other agents.

Hyoscine hydrobromide or **glycopyrrolate** may be added if excessive secretions are problematic.

A **Phenobarbitone** infusion can be used for refractory seizures or sedation; however it needs to be administered on a separate infusion site as it is not compatible with other medication.

Some medications **should not** be administered subcutaneously. These include:

antibiotics, diazepam, chlorpromazine and prochlorperazine, as they often cause skin reactions at the injection and infusion sites.

The Syringe Driver – Continuous Subcutaneous Infusions in Palliative Care provides a more exhaustive reference of various drug compatibilities used in palliative care¹¹⁶. Alternatively, you can also liaise with your pharmacist.

APPENDIX 4. CONTENTS OF HOME CARE PACK

To facilitate early treatment of symptoms and the availability of specific drugs, it is advisable that families living away from their treating hospital are given an initial home care pack. This pack should contain medications that may be required for general symptom relief and the terminal phase of the illness.

Contents for consideration:

- Medical summary of current condition and goals of care.
- Copy of any relevant care plan where applicable (e.g. Allow Natural Death/End of Life Care Plan; or equivalent).
- Symptom management plan.
- Medication supply related to symptom management plan.
- Terminal care medication suggestions include opioid – morphine (of various strengths), benzodiazepine – midazolam, anticholinergic – hyoscine hydrobromide (in parenteral formulation), anti-emetic – metoclopramide.
- Medication supply related to suggested terminal care starting dosages.
- Contact details of relevant health professionals including an afterhours contact plan.
- Equipment for commencing subcutaneous infusion or intermittent dose (Appendix 2).
- Spare subcutaneous access devices: drawing up needles; syringes; alcohol swabs.
- 1ml syringes for bolus doses, bungs/stoppers and labels.
- Emla® cream or similar.
- Water and Normal Saline for injection.
- Sharps container.
- Syringe driver – with details of who can setup and commence as well as a troubleshooting plan.

Equipment for home care

Listed below is equipment that may assist in caring for the child at home.

- Pressure relief mattress as prescribed or recommended by an occupational therapist e.g. over lay, gel-pad or ripple.
- Incontinent supplies e.g. disposable draw sheets, nappies.
- Bed pan, urinal and commode.
- Wheelchair or buggy, walking aids, manual handling belt, hoist.
- TENS machine (if used prior).
- Oxygen concentrator.
- Portable oxygen cylinder.
- Portable suction, disposable suction catheters.
- Mouth care equipment e.g. toothettes.
- Nasogastric feeding equipment.
- Specimen jars (urine and faeces).
- Spare batteries for the infusion pump.
- Consider the need for appropriate IDC – particularly if spinal cord complications are a risk.
- Consider extra supplies that may be required if bleeding is a risk.

APPENDIX 5. STANDARDS FOR PROVIDING QUALITY PALLIATIVE CARE FOR ALL AUSTRALIANS

Standard 1

Care, decision-making and care planning are each based on a respect for the uniqueness of the patient, their caregiver/s and family. The patient, their caregiver's and family's needs and wishes are acknowledged and guide decision-making and care planning.

Standard 2

The holistic needs of the patient, their caregiver/s and family, are acknowledged in the assessment and care planning processes, and strategies are developed to address those needs, in line with their wishes.

Standard 3

Ongoing and comprehensive assessment and care planning are undertaken to meet the needs and wishes of the patient, their caregiver/s and family.

Standard 4

Care is coordinated to minimise the burden on patient, their caregiver/s and family.

Standard 5

The primary caregiver/s is provided with information, support and guidance about their role according to their needs and wishes.

Standard 6

The unique needs of dying patients are considered, their comfort maximized and their dignity preserved.

Standard 7

The service has an appropriate philosophy, values, culture, structure and environment for the provision of competent and compassionate palliative care.

Standard 8

Formal mechanisms are in place to ensure that the patient, their caregiver/s and family have access to bereavement care, information and support services.

Standard 9

Community capacity to respond to the needs of people who have a life-limiting illness, their caregiver/s and family is built through effective collaboration and partnerships.

Standard 10

Access to palliative care is available for all people based on clinical need and is independent of diagnosis, age, cultural background or geography.

Standard 11

The service is committed to quality improvement and research in clinical and management practices.

Standard 12

Staff and volunteers are appropriately qualified for the level of service offered and demonstrate ongoing participation in continuing professional development.

Standard 13

Staff and volunteers reflect on practice and initiate and maintain effective self-care strategies.

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APPENDIX 6

APPENDIX 6. CLINICAL INDICATORS FOR PAEDIATRIC PALLIATIVE CARE

1	Multi-disciplinary assessment undertaken?	Yes / No
2	Care Plan created?	Yes / No
3	Information resources provided to families?	Yes / No
4	Link with GP/community nurse established?	Yes / No
5	Offered respite/"short break" services?	Yes / No
6	Offered spiritual support?	Yes / No
7	Offered community services?	Yes / No
8	Offered support for siblings?	Yes / No
9	Equipment needs assessed?	Yes / No
10	Equipment provided in a timely manner?	Yes / No
11	Family goals identified and modified as required?	Yes / No
12	Financial assessment completed?	Yes / No
13	Difficult symptoms discussed?	Yes / No
14	Written plan about managing difficult symptoms provided?	Yes / No
15	Choices for care options discussed and documented?	Yes / No
16	Support during end of life care provided?	Yes / No
17	Preferences for location of end of life care discussed with caregivers?	Yes / No
18	Access to the right location at the right time?	Yes / No
19	Accompanied by family member or friend at the time of death?	Yes / No
20	Presence of unresolved issues between family and team?	Yes / No
21	Bereavement key support worker identified?	Yes / No
22	Death review completed?	Yes / No

Source:

Department of Pain Medicine and Palliative Care, The Children's Hospital at Westmead, 2008.

Australian and New Zealand Paediatric Palliative Care Reference Group, 2009.

National Standards Assessment Program Paediatric Self-Assessment Workbook, Palliative Care Australia, Canberra, 2011 version 2.

APPENDIX 7

APPENDIX 7. COMMONLY USED DRUGS AND DOSES

Drug and formulation	Indication	Route, dose and frequency	Ceiling dose
Amitriptyline 'Endep®' <i>Tablets: 10mg, 25mg, 50mg</i>	Neuropathic pain May assist sleep	PO 0.2–1mg/kg/dose Nocte (Can also be given twice daily)	50mg/day
Benzotropine 'Cogentin®' <i>Amp: 1mg/ml Tablets: 0.5mg, 2mg</i>	Extrapyramidal side effects Dystonia	IV/IM < 12yrs: 0.02mg/kg/dose stat. > 12yrs: 1–2mg/dose stat. Repeat after 15min if required	2mg/dose
Carbamazapine 'Tegretol®' <i>Tablets: 100mg, 200mg Tablets CR: 200mg, 400mg Controlled release tablets should be given every 12hrs Liquid: 20mg/ml</i>	Seizures Neuropathic pain	PO/PR 2.5mg/kg/dose twice daily or 5mg/kg/dose nocte, increase over 2 weeks to 5–10mg/kg/dose Q8–12h	2g/day
Chloral Hydrate Liquid <i>50mg/ml, 100mg/ml</i>	Sedation	PO/PR 10–20mg/kg/dose Q6 h prn Can use higher doses (up to 50mg/kg) with care	1g/dose
Chlorpromazine 'Largactil®' <i>Tablets: 10mg, 25mg, 100mg Liquid: 5mg/ml, 10mg/ml Amp: 25mg/ml</i>	Agitation Nausea	PO/IV 0.1–1mg/kg/dose Q6 – 8h	< 5yrs: 40mg/day 5–12yrs: 75mg/day > 12yrs: 300mg/day
Clonazepam 'Rivotril®' <i>Tablets: 0.5mg, 2mg Liquid: 2.5mg/ml Amp: 1mg/ml + diluent 1 drop = 100mcg 25 drops = 1ml</i>	Agitation/Anxiety Seizures Dyspnoea	PO/Sublingual < 12yrs: 0.01mg–0.05mg/kg/dose Q8–12h > 12yrs: Initially 0.5mg/dose Q8–12h IV/SC 0.125–0.50mg stat (slow). Repeat if required.	0.2mg/kg/day for children 20mg/day for adults 1mg/dose (IV/SC)
Codeine (S8) <i>Tablets: 30mg Liquid: 5mg/ml</i>	Analgesic	PO/PR 0.5–1mg/kg/dose Q4–6h	60mg/dose
Cyclizine (SAS) <i>Tablets: 50mg Amp: 50mg/ml</i>	Nausea	PO/IV/SC 0.5–1mg/kg/dose Q6–8h Can be administered as continuous infusion. (Use water for injection)	50mg/dose

APPENDIX 7

Drug and formulation	Indication	Route, dose and frequency	Ceiling dose
Dexamethasone <i>Tablets: 0.5mg, 4mg</i> <i>Amp: 4mg/ml</i>	Cerebral Oedema Spinal cord compression	PO/IV/SC 0.25–0.5mg/kg/dose Q6–12h Seek specialist advice: A bolus dose of 1–2mg/kg can be given initially prior to urgent imaging if available/appropriate.	20mg/dose
	Anti-inflammatory (peripheral nerve compression, pain, bowel obstruction)	PO/IV/SC 0.1–0.25mg/kg/dose Q6–12h	8mg/dose
	Nausea	PO/IV 0.1–0.25mg/kg/dose Q6h–daily	8mg/dose
Diazepam ‘Valium’ <i>Tablets: 2mg, 5mg</i> <i>Liquid: 1mg/ml</i> <i>Amp: 5mg/ml</i> <i>Enema: 5mg</i>	Anxiety Muscle spasm	PO 0.05–0.3mg/kg/dose Q6–12h IV 0.1–0.3mg/kg Q4h–12h	40mg/day
	Seizures	IV 0.1–0.3mg/kg/dose stat. PR 0.3–0.5mg/kg/dose Repeat at 15–30 minute intervals if required.	10mg/dose
Diclofenac ‘Voltaren®’ <i>Tablets: 12.5mg, 25mg, 50mg</i> <i>Tablets (EC): 25mg, 50mg</i> <i>Suppositories: 12.5mg, 25mg, 50mg, 100mg</i>	Anti-inflammatory	PO/Rectal 1mg/kg/dose Q8–12h with food	50mg/dose
Docusate Sodium ‘Coloxyl®’ <i>Tablets: 50mg, 120mg</i>	Constipation	PO < 3yrs: consider poloxamer drops 3–6yrs: 50mg Q8h – Daily 6–12yrs: 50–120mg Q8h – Daily > 12yrs: 100–150mg Q8h – Daily	< 12yrs: 240mg/day > 12yrs: 500mg
Docusate Sodium ‘Coloxyl® with Senna’	Constipation	PO 1–2 tablets/dose Q8h – Daily	6 tablets/day
Domperidone ‘Motilium®’ <i>Tablet: 10.mg</i>	Nausea Gastrointestinal stasis	PO 0.2–0.4mg/kg/dose Q6–8h	20mg/dose
Gabapentin <i>Tablets: 600mg; 800mg</i> <i>Capsules: 100mg; 300mg; 400mg</i> <i>Capsules may be opened and dissolved in water.</i>	Neuropathic pain Itch	PO 5mg/kg/dose Daily – Q8h (increase dose over days – weeks). Day 1: Daily Day 2: Q12h Day 3: Q8h	20mg/kg/dose
Glycopyrrolate ‘Robinul®’ <i>Tablets: 1mg (SAS)</i> <i>Amp: 0.2mg/ml (can be given orally)</i>	Reduce secretions	PO 40–100mcg/kg/dose Q6–8h IV/SC 4–10mcg/kg/dose Q6–8h Can be administered as continuous infusion. Use water for injection.	400mcg/dose

APPENDIX 7

Drug and formulation	Indication	Route, dose and frequency	Ceiling dose
Haloperidol 'Serenace®' Liquid: 2mg/ml Tablets: 0.5mg, 1.5mg, 5mg Amp: 5mg & 10mg/ml.	Agitation/Delerium Nausea/Vomiting	PO/IV/SC < 12yrs: 0.01–0.1mg/kg/dose Q8–12h > 12yrs: 500mcg–2.5mg Q8–12h Can be administered as continuous infusion.	7.5mg/day
Hyoscine N-Butylbromide 'Buscopan®' Tablet: 10mg Amp: 20mg/ml	Antispasmodic (renal, gastrointestinal)	PO/IV/SC 0.5mg/kg/dose Q6h	20mg/dose
Hyoscine Hydrobromide Tablets: 0.3mg (Kwells®) Amp: 400mcg/ml Patch: 1.5mg (Scopoderm®) (SAS)	Reduce secretions	PO 2–7yrs: ¼ tablet/dose Q6h > 7yrs: ½–1 tablet /dose Q6h IV/SC 6–10mcg/kg/dose Q6h Can be administered as continuous infusion. Transdermal < 3yrs: ¼ patch Q72h 3–9yrs: ½ patch Q72h > 9yrs: 1 patch Q72h	400mcg/dose
Ibuprofen 'Nurofen®' Liquid: 20mg/ml Tablet: 200mg, 400mg	Anti-inflammatory	PO 5–10mg/kg/dose Q6–8h with food	400mg/dose
Lactulose 'Duphalac®' Liquid: 0.67g/ml liquid 70%	Constipation	PO 1ml/kg/dose Q12h – daily	45ml/dose
Levomepromazine 'Nozinan®' (SAS) Tablet: 25mg Amp: 25mg/ml	Nausea and vomiting	PO /IV/SC < 12yrs: 0.1mg–1mg/kg/dose Q12h – daily > 12yrs: 6.25–25mg Q12h – daily Can be administered as continuous infusion.	25mg/day 50mg/day
	Terminal restlessness	IV/SC 0.3–3mg/kg/day as a continuous infusion.	
Loperamide 'Imodium®' Capsule/tablet/melts: 2mg	Diarrhoea	PO 0.05–0.1mg/kg/dose Q8–12h	2mg/dose
Lorazepam 'Ativan®' Tablets: 1mg, 2.5mg	Anxiety Anticipatory nausea and vomiting. Dyspnoea	PO 0.02–0.05mg/kg/dose Q8–24h Can be administered sublingually	2.5mg/dose

APPENDIX 7

Drug and formulation	Indication	Route, dose and frequency	Ceiling dose
Macrogol 3350 and electrolytes 'Movicol®' 'OsmoLax®' 'ClearLax®'	Constipation	PO Movicol ® 2–5yrs: 1 x Movicol-Half sachet Daily 6–11yrs: 1 x Movicol sachet/dose Daily > 12yrs: 1–2 x Movicol sachet/dose Daily (May need to increase to Q8h). ClearLax ® Children 2–6yrs: dissolve 1/2 of contents of 1 sachet in any 100ml liquid, then drink Adults, children greater than or equal to 6yrs: dissolve contents of 1 sachet in any 120–250ml liquid, then drink. Osmolax ® Children 4–5yrs: initially one 8.5g scoop mixed with 1/2 cup liquid once daily. Children 6–12yrs: two 8.5g scoops (or one 17g scoop) mixed with 1 cup liquid once daily; adjust dose up or down to produce regular soft stools. Adults, children > 12yrs: one 17g scoop mixed with 1 cup (approx 240ml) liquid once daily. Chronic constipation: may reduce to one 8.5g scoop mixed with 1/2 cup (approx 120ml) liquid daily, according to individual response.	Titrate dose to effect
Metoclopramide 'Maxolon®' Tablet: 10mg Syrup discontinued. Amp: 10mg/2ml	Nausea and vomiting	PO/IV/SC 0.1–0.2mg/kg/dose Q6h Can be administered as continuous infusion.	10mg/dose
Midazolam 'Hypnovel®' Amp: 5mg/ml, 15mg/3ml, 50mg/10ml. Plastic 5mg/ml amps can be used nasally or buccally.	Agitation Seizures Dyspnoea Premedication	Buccal/PO/Intranasal < 20kg: 0.3–0.5mg/kg/dose > 20kg: 5–10mg/dose Dose can be repeated. IV/SC 0.1–0.2mg/kg/dose Q4h (Dose can be titrated) Can be administered as continuous infusion 1–5mcg/kg/min (to make this infusion add 3 x bodyweight (kg) of midazolam inmg to 50ml of normal saline or dextrose solution and infuse at rate of 1–5ml/hour) or 0.2–1mg/kg/24 hr as a starting dose.	10–15mg/dose – higher in refractory cases

APPENDIX 7

Drug and formulation	Indication	Route, dose and frequency	Ceiling dose
Morphine sulphate (S8) Immediate release Tablet: 'Sevredol®' 10mg, 20mg 'Anamorph®' 30mg Liquid: 'Ordine®' 1mg/ml, 2mg/ml, 5mg/ml, 10mg/ml Amp: Morphine sulphate: 5mg/ml, 10mg/ml, 15mg/ml, 30mg/ml Morphine tartrate: 120mg/1.5ml, 400mg/5ml Controlled Release MS Contin® – tablet, suspension Kapanol® – capsule (see page 48)	Analgesic for moderate – severe pain	PO Neonate – 3 mo: 0.05–0.1mg/kg/dose Q4–6h 3–6 mo: 0.1mg/kg/dose Q4–6h > 6 mo: 0.2–0.5mg/kg/dose Q4–6h SC/IV Neonate – 3 mo: 0.025mg/kg/dose Q6h 3–6 mo: 0.05mg/kg/dose Q6h > 6 mo: 0.1–0.2mg/kg/dose Q4–6h	Increase dose as required – no max dose.
	Dyspnoea	30–50 % of the dose used for pain PO 0.05–0.1mg/kg/dose Q4–6h SC/IV 0.025–0.05mg/kg/dose Q4–6h	
Naloxone 'Narcan' Amp: 400mcg/ml	Acute respiratory depression related to opioids Itch	IV 5 mcg/kg. Repeat every 2–3 minutes until adequate respirations are established. Caution in children receiving chronic opioid therapy. IV 0.5mcg/kg may assist itch. Consider low dose infusion (0.5mcg/kg/hr).	100mcg/dose
Octreotide	Bowel obstruction or Intractable diarrhoea and gastrointestinal fluid loss	IV/SC 1–10mcg/kg/ dose Q8h Can be administered as a subcutaneous infusion. 25mcg/kg/24 hours. Initial dose for older children is 300mcg/day. Titrate to effect over 24–48 hours.	500mcg/dose 1000mcg/day
	Acute variceal bleeding	Load 1mcg/kg/dose then 1–5mcg/kg/hour (increase every 8 hours if no effect)	50mcg/hr
Omeprazole 'Losec®' Tablets: 10mg, 20mg Capsule: 20mg Some hospitals prepare a mixture for small doses and nasogastric tubes.	Antacid Reflux Oesophagitis/ Gastritis	PO 0.5–1mg/kg/dose Q12h – daily	30mg/dose
Ondansetron 'Zofran®' Tablets/wafers: 4mg, 8mg Liquid: 4mg/5ml Amp: 4mg & 8mg/2ml Suppository: 16mg	Nausea and vomiting	PO/IV/SC 0.15mg/kg/dose Q8h PR (> 12yrs) 16mg/dose Daily	8mg/dose

APPENDIX 7

Drug and formulation	Indication	Route, dose and frequency	Ceiling dose
<p>Oxycodone (S8) Immediate Release Tablet: 5mg (Endone®) Capsule: 5mg, 10mg, 20mg (Oxynorm®) Liquid: 1mg/ml Amp: 10mg/ml, 20mg/2ml Suppository : 30mg (Proladone®)</p> <p>Controlled Release Oxycontin® (See page 48)</p>	Analgesic for moderate – severe pain	PO 0.1–0.25mg/kg/dose Q4–6h	Increase dose as required – no max dose.
<p>Paracetamol ‘Panadol®’ Liquid: 24mg/ml, 48mg/ml, 50mg/ml, 100mg/ml, Tablet/capsule: 500mg Amp: 500mg; 1g Suppositories; 125mg, 250mg, 500mg</p>	Analgesic Antipyretic	PO/PR/IV 15mg/kg/dose Q4–6h For neonates and children weighing <10 kg to approximately 1 year of age the IV dose is 7.5mg/kg/dose Q6h	< 6 mo: 60mg/day > 6 mo: 90mg/day
<p>Paraffin Liquid Liquid: 50%</p>	Constipation	PO 1–3ml/kg/dose Daily	50ml/day
<p>Phenobarbitone Tablet: 30mg Liquid: 3mg/ml, 10mg/ml Amp: 200mg/ml</p>	Seizures	PO/IV/SC (slow) 2.5–5mg/kg/dose Q12 – Daily (maintenance) Loading dose: IV (slow)/SC (slow)/IM 20–30mg/kg Can be administered as continuous infusion. Essential to dilute injection in 10 times the volume of water and use a separate subcutaneous site if given as subcutaneous infusion.	600mg/day – higher in refractory cases 1g/dose
<p>Phenytoin ‘Dilantin®’ Infatab: 50mg (chewable) Capsules: 30mg, 100mg Liquid: 6mg/ml Amp: 50mg/ml</p>	Seizures	PO/IV 2mg/kg/dose Q8–12h (maintenance) Loading dose IV 15–20mg/kg over 20 minutes (can be given in 3 parts over 6 hours orally)	100mg/dose – higher in refractory cases 1.5g
<p>Poloxamer ‘Coloxyl drops®’ Liquid: 10%</p>	Laxative	PO < 6 mo: 10 drops/dose Q8h 6–18 mo: 15 drops/dose Q8h 18 mo–3yrs: 25 drops/dose Q8h	
<p>Prochlorperazine ‘Stemetil®’ Tablets: 5mg Amp: 12.5mg/ml Suppository: 5,25mg</p>	Nausea and vomiting	PO/IV/IM/PR 0.2mg/kg/dose Q6–12h Caution with IV use due to cardiovascular side effects. Give as a slow IV push (maximum rate is 5mg/min, preferably slower)	Oral 10mg/dose

APPENDIX 7

Drug and formulation	Indication	Route, dose and frequency	Ceiling dose
Promethazine 'Phenergan®' Tablets: 10mg, 25mg Liquid: 1mg/ml Amp: 25mg/ml	Nausea and vomiting Itch Sedation	PO/IV/SC 0.125–0.5mg/kg/dose Q6–8h	150mg/day
Ranitidine 'Zantac®' Tablets: 150mg, 300mg (effervescent tabs 150mg) Liquid: 15mg/ml Amp: 10mg/ml, 25mg/ml	Antacid Reflux Oesophagitis/ Gastritis	PO 2–4mg/kg/dose Q8–12h IV 1mg/kg/dose Q8h	300mg/day (POI) 50mg/dose (IV)
Senosides 'Senokot®' Tablets/chocolate squares: 7.5mg	Constipation (Stimulant Laxative)	PO 2–6yrs: ½–1 tablet/dose Nocte 6–12yrs: 1–2 tablet/dose Nocte >12yrs: 1–4 tablet/dose Nocte	30mg/day
Sucralfate Tablet: 1g Some pharmacies make cream.	Mucosal bleeding/ oozing	½–1 tablet dispersed in water and applied topically as required	
Tranexamic Acid 'Cyclokapron®' Tablet: 500mg IV formulation available (SAS)	Antifibrinolytic for bleeding Also used for mouthwash	PO 15–25mg/kg/dose Q8–12h 500mg tablet dissolved in 5–10ml water (or 5% mouth wash) Rinse 10ml in mouth for 2 minutes then spit out Q6h	1.5g/dose

The drugs included are those commonly used for symptom management. This information is designed to be a dose guide only. Each patient's dose requirements may vary and should be adjusted based on the clinical situation. Readers should also refer to larger texts on Palliative Care for further drugs, indications and side effects. All care has been made to ensure that doses are accurate, but the user is advised to check these carefully and to consult the above references and the text of this guide for potential toxicities. Contact with an experienced pharmacist is also advisable. The authors shall not take responsibility for any errors in publication of drug doses or in drug administration.

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- Ceiling dose:** Increases in dose beyond the ceiling dose may not elicit any further increase in therapeutic effect. The addition of an alternative drug should be considered (either to replace or be used in conjunction with the existing therapy).
- Enteric coated:** Coated with a material that permits transit through the stomach to the small intestine before the medication is released. This may minimise adverse gastrointestinal effects. These tablets/capsules cannot be crushed or chewed.
- S8:** (Schedule 8) Poisons schedule. Special restrictions apply regarding prescribing, dispensing and supply.
- SAS:** Special Access Scheme Medications unavailable in Australia that may be imported via the Therapeutic Goods Administration (TGA). Special documentation required and supply issues as these items are sourced from overseas suppliers.

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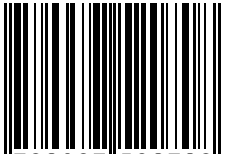
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