

Overview of symptoms and their assessment in life-limiting illness



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Chapter:

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Introduction



In 2018 the World Health Organization published a guidance document for healthcare workers delivering paediatric palliative care in which it describes '*the medical and moral necessity of making palliative care accessible to all children in need and their families*' (1, p. 1). Symptom management is an essential part of this process, and in order to provide this minimum standard of palliative care, it is vital that healthcare providers understand the multi-faceted nature of symptoms and their contextual impact on the child with serious illness. It is therefore helpful to identify accurately those children with life-limiting and life-threatening conditions, and to recognize subpopulation-specific patterns of symptom presentation. This may improve access to expertise for good symptom management.

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In the past, judging the severity of symptoms relied on observational variables such as heart rate, respiratory rate, and changes in posture. At best, those observations were elaborated by verbal reports from relatives, other carers, or the child herself. In the twenty-first century, however, symptom evaluation has become more sophisticated, harnessing software that can be easily installed on smartphones (2). These improvements enable more accurate symptom assessment in real time and better patient/professional interface for planning management strategies. There has also been recognition of the diversity of symptom reporting by the child, carer, and healthcare professional and this combined viewpoint provides a valuable global perspective to the patient's symptom experience, enabling more accurate planning of acceptable interventions (2).

Despite improved capabilities for collecting data on patient symptoms, there is a sense that existing barriers continue to prevent the systematic study of children with life-threatening conditions, thus leading to an incomplete impression of the problem. Poorly identified paediatric palliative care populations, poor access to expert palliative care services, and gate keeping by ill-informed healthcare professionals remain key barriers to life-limited children living and dying with well-managed symptoms (3).

The aims of this chapter are:

- To discuss the prevalence of symptoms in paediatric palliative care and within subpopulations of life-limited children;
- To present the historical development of symptom evaluation using pain evaluation as an example;
- To discuss the principles of symptom evaluation;
- To discuss the challenges to comprehensive symptom evaluation;
- To examine the areas where knowledge and expertise are lacking, and the reasons for this;
- To present novel approaches to symptom evaluation;
- To discuss areas for future study;
- To identify the resources available for symptom evaluation.

The importance of accurate symptom assessment

In medicine, the signs of pathology describe effects on a patient that a third party can observe, while its symptoms describe what the patient herself experiences. The obvious way to relieve unpleasant symptoms, if it can be done, is to treat the underlying cause. But not all pathology is reversible, and even where treatment is curative there is often a delay while it takes its effect. In the past there has been a tendency to consider symptom management for its own sake only when curative treatment was no longer possible. Thankfully, health

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professionals are increasingly acknowledging that symptom management should be considered alongside efforts to treat the underlying disease process, and that if reversal of the pathophysiological process is no longer possible, then therapy still needs to be directed at palliating the unpleasant symptoms.

Therefore, in order to manage a symptom successfully it is necessary to recognize and to assess it, to diagnose and to measure it, to evaluate it in a particular individual at a particular point in time and disease trajectory, and then to direct an appropriate therapeutic plan to address it. The management of a symptom may include investigating the underlying pathology and reversing it, but it may also involve therapy to alleviate the resulting symptom alone. The relative importance of these strategies is the basis of goal-directed management.

The prevalence of symptoms in children with life-limiting illness



The World Health Organization's *Essential Medicines List for Children* (4) concludes that the prevalence of symptoms in children at the end of life, and the likelihood that they are controlled by the time of a child's death, remain largely unknown. However, developments in research methodologies are addressing some of these areas and improving evidence-based practice.

- Studies over the past 20 years have clearly demonstrated an increasing number of children living with life-limiting illnesses (5), yet the exact prevalence of these children in most healthcare systems remains largely unknown. Furthermore, trends in this dataset reflect that children are surviving longer with life-limiting illnesses necessitating long-term symptom management and transition to adult services (6).
- There has been a lack of high-quality prospective and randomized controlled trials involving symptoms, their management, and associated distress in children's palliative care. Several retrospective case reviews and interview studies of staff and bereaved parents exist, but these studies are subject to recall and recording bias. With prospective studies the rarity of childhood life-threatening illness, lack of homogenous populations especially in non-malignant disease, and multiple variables in care settings often leads to poor quality research methodologies and inconclusive outcomes. However, utilizing novel approaches to study design such as mixed methods qualitative data collection for symptom evaluation and exposure-crossover design for interventional studies may improve the quality of research in symptom management (7).
- Extrapolation from adult symptom prevalence studies may lead to inaccuracies, as there are differences in the symptom profiles between children and adults, even for comparable diseases such as single organ failure. There are differences in symptom profiles at different ages and

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developmental stages even within the same disease. For example, the symptoms of cardiac failure that are exhibited by an infant (feed intolerance, agitation, and increased sweating) are different from those expressed by an older child (dyspnoea, discomfort due to dependent oedema, and palpitations).

- Children with developmental delay, cognitive disability, and life-limiting disease express symptoms differently from other children, and there may also be distinct perception and processing pathways involved. This poses a challenge both to identifying symptoms and to the development of objective assessment tools in this population.
- There are rare life-limiting diseases that are unique to childhood. However, due to the small numbers of children affected worldwide, it is difficult to obtain reliable data for these populations.
- Despite challenges with obtaining robust data, there are increasing collaborative reviews of existing data such as *Pharmacological Interventions for Pain in Children and Adolescents with Life-Limiting Conditions* (8), and guidance for practice informed by national and international consortia (the NICE guideline *End of Life Care for Infants, Children and Young People with Life-Limiting Conditions: Planning and Management*) (9).

The majority of studies over the past two decades reporting on symptomatology in life-limiting disease in children have focused on those at the end of life and those occurring in specific disease states. Initial studies reported symptom prevalence from the viewpoint of professionals and parents, but more recently some have included the child's perspective in combination with the caregivers' report.

Specific disease states, such as cancer (which accounts for one-third of childhood life-limiting illness) have been studied in the greatest detail. This is most probably due to the fact that this is a relatively homogenous patient group, with a reasonably large, well-defined population who are already involved in treatment studies and so are familiar with the research process. Children with cancer as well as other paediatric disease states, that have received the most attention in the published literature, tend to be cognitively developmentally appropriate for their age, and so are more likely to be able to comply with existing symptom assessment tools and provide self-report.

Studies involving children from other specific disease groups or generalized life-limiting illness groups—such as human immunodeficiency virus/acquired immunodeficiency syndrome (HIV/AIDS), neurodegenerative life-limiting illnesses (e.g. Duchenne muscular dystrophy), cystic fibrosis, and life-limiting genetic and congenital malformation disorders—are emerging as paediatric palliative care practice reaches further into the arena of non-malignant disease.

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Symptom reporting in the paediatric end-of-life care literature

Table 14.1 outlines the most common symptoms of children cared for in an inpatient setting (hospital or hospice) at the end of life—including children with cancer, cerebral palsy, metabolic conditions, and congenital abnormalities—reported in key mixed-cohort studies.

Rank	Carter et al.	Drake et al.	Hunt
1	Dyspnoea	Lack of energy	Pain
2	Seizures/ convulsions	Feeling drowsy	Oral symptoms
3	Prolonged crying	Skin changes	Dyspnoea
4	Terminal agitation	Feeling Irritable	Urinary problems
5	Fear/anxiety	Pain	Skin problems
6	Nausea/vomiting	Swelling of arms or legs	Anorexia
7	Fatigue	Cough	Vomiting
8		Dyspnoea	Psychological Problems
9		Dry mouth	Cough
10		Lack of concentration	Constipation

Source: data from Carter B. et al. (2004). Circumstances surrounding the deaths of hospitalised children: Opportunities for pediatric palliative care. *Pediatrics*. 114(3):e361-6. DOI: 10.1542/peds.2003-0654-F; Drake R. et al. (2003). The symptoms of dying children. *J Pain Symptom Manage*. 26(1):594-603. DOI: [https://doi.org/10.1016/S0885-3924\(03\)00202-1](https://doi.org/10.1016/S0885-3924(03)00202-1); and Hunt AM. (1990). A survey of signs, symptoms and symptom control in 30 terminally ill children. *Dev Med Child Neurol*. 1990;32:341-6. <https://doi.org/10.1111/j.1469-8749.1990.tb16946.x>.

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There have been few studies reporting symptoms at the end of life, but some common observations have been noted:

- The symptom burden at the end of life is high, whether it is reported by patients, parents, or professionals. However, Wolfe and colleagues (10) concluded from their study based on parent proxy reports that there was a high level of suffering and distress, in contrast to the study by Drake and colleagues (11), based on professional proxy reports, which suggested that although the symptom burden was high, the level of distress at the end of life was not.
- Studies of end-of-life care in hospital settings have identified a high proportion of children dying in intensive-care units as opposed to ward settings. However, the tendency to more aggressively manage symptoms and even purposely sedate patients within the paediatric intensive care unit in order to facilitate care, may paradoxically result in the perception of a lower symptom prevalence and proxy-reported distress within this setting when compared with patients admitted to other hospital wards, as posited by Drake and colleagues (11).
- All of the studies are retrospective, but observer variation due to the evaluation of subjective and psychological phenomena such as symptoms at the end of life seems to be present regardless of whether the proxy reporter is a professional or a parent/carer. This is especially so for children who are difficult to assess, such as preverbal children, cognitively impaired children, and those who are unconscious.
- There are a lack of validated tools for assessing symptoms at the end of life, but where the use of such tools confers some consistency of examination, they have nevertheless been utilized. Therefore, tools such as the Memorial Symptom Assessment Scale (which is a subject-reported scale that has been validated in cancer patients) have been used in invalidated settings by proxy reporters. Other studies have adopted a retrospective qualitative interview-based approach.
- All of the studies highlight the lack of reliable data that persists in this area of end-of-life care, and how this can sometimes compromise attempts at intensive symptom management.

Symptom reporting in the paediatric cancer literature

Symptoms reported in children with cancer may be related to a combination of the disease, procedures, treatments such as chemotherapy regimens, and the overall experience of cancer. Some child- and parent-reported symptoms appear to be relatively consistent and they concur with those highlighted by professionals. Goldman and colleagues reported a questionnaire-based study of children and young people with cancer and their caregivers, who consented to report their symptoms at the time of entry to the study and during the last month of life (12). The most commonly reported symptoms were pain, weakness, nausea and vomiting, constipation, and loss of mobility. Collins and

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colleagues, applying the Memorial Symptom Assessment Scale for 10- to 18-year olds (MSAS 10-18), validated this tool in this age group and reported that lack of energy, pain, drowsiness, nausea, cough, lack of appetite, and psychological symptoms were the symptoms most commonly reported by a group of 160 children and young people with cancer aged 10-18 years (13).

Healthcare providers need to be aware that not all patients and caregivers associate their symptoms with their disease or treatment. Woodgate and colleagues recorded a range of symptoms reported by children and caregivers in a longitudinal qualitative study of thirty-nine children, using open-ended formal interviewing and participant observation (14). The most common meaning-assigned symptoms were pain ('I am sore, hurting'), nausea ('I have a sick stomach'), fatigue ('I am wiped out'), and feeling irritable ('I am cranky'). The authors concluded that a greater understanding of symptoms was achieved when they were approached as a multidimensional experience rather than just a side effect of treatment or a disease-related phenomenon.

Interestingly, pain management in paediatric palliative care is an area of expertise that professionals often feel confident about, but the evidence suggests that it sometimes remains inadequately controlled. In a survey of clinical nurse specialists regarding symptoms in a cohort of children with cancer, pain was felt to be poorly controlled at the end of life, with the prevalence increasing from 71 to 92% *after* referral to palliative care 12. Table 14.2 outlines the most common cancer-related symptoms reported in key studies.

Table 14.2 Cancer-related symptoms most commonly reported in key studies

Rank	Wolfe et al.	Jalmsell et al.	Heath et al.
1	Fatigue	Fatigue	Pain
2	Pain	Reduced mobility	Fatigue
3	Dyspnoea	Pain	Poor appetite
4	Poor appetite	Poor appetite	Constipation
5	Nausea/ vomiting	Nausea	Dyspnoea
6	Constipation	Weight loss	Nausea/ vomiting

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7	Diarrhoea	Sleepiness (daytime)	Diarrhoea
8		Vomiting	Seizures
9		Depression	
10		Disturbed sleep (due to pain)	

Sources: data from Wolfe J. et al. (2000). Symptoms and suffering at the end of life in children with cancer. *N Engl J Med*. 42(5):326–33. DOI: 10.1056/NEJM200002033420506; Jalmsell L. et al. (2006). Symptoms affecting children with malignancies during the last month of life: A nationwide follow-up. *Pediatrics*. 117(4):1314–20. DOI: 10.1542/peds.2005-1479; and Heath JA. et al. (2010). Symptoms and suffering at the end of life in children with cancer: An Australian perspective. *Med J Aust*. 192(2):71–5. <https://doi.org/10.5694/j.1326-5377.2010.tb03420.x>.

Symptom reporting in the paediatric HIV/AIDS-related literature

A study by Lavy and colleagues at a hospital in Southern Malawi reported the symptoms and signs in ninety-five children who had been referred for palliative care (15). The majority (75%) had HIV, and 17% had cancer diagnoses. The authors reported that pain was significantly more common in the children with cancer diagnoses than in those with HIV/AIDS. In the cancer group the most common symptoms were pain, cough, and diarrhoea, whereas in the HIV/AIDS group the commonest symptoms were weight loss, fever, and mouth sores. The authors also noted that 47% of the children with HIV/AIDS had either lost their mother or had a mother who was ill with the disease. The authors felt that this contributed to these children having more family and/or social problems than children in the other disease groups. They stated that this highlights the impact of psychological, emotional, spiritual, and social factors on the presentation of physical symptoms, and the need for palliative care to be holistic if symptom management strategies are to be successful.

Symptom reporting in the paediatric respiratory and cardiac disease literature

Few studies have specifically reported on symptoms of paediatric patients with primary respiratory disease in palliative care. In a study that examined the prevalence of symptoms in a cohort dying of cystic fibrosis, dyspnoea (100%), fatigue (96%), anorexia (85%), anxiety (74%), pain (67%), and cough (56%) were found to be most common (16). In contrast, there is a growing body of literature about respiratory symptoms secondary to a variety of non-respiratory underlying medical conditions (e.g. cardiac failure, infection, malignancy, neurologic impairment). The

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respiratory symptoms most commonly described within this literature include dyspnoea, cough, and problems with oropharyngeal secretions.

Symptoms experienced by children with advanced heart disease are often respiratory in nature, including dyspnoea and cough, secondary to left-sided pump failure. But pain and irritability (especially in younger children) are also common, as are the non-specific symptoms of anorexia, fatigue, and poor sleep (17).

Symptom reporting in the literature on progressive neurological disease

As children and young people with progressive neurological conditions deteriorate, they often experience new or worsening symptoms as the direct result of nervous system changes or indirectly because of its impact on other organ systems. Categories of disease for whom these changes can be expected include genetic or metabolic conditions, muscular dystrophies, encephalopathies, cerebral palsy, spinal muscular atrophy, and more (for a complete discussion of the symptoms affecting these conditions, please see Chapter 23, 'Neurological and neuromuscular conditions').

As the nervous system declines, it is not unusual for the child to experience neurologic symptoms such as neuropathic pain, agitation, dystonia, seizures, and other neurologic sequelae as a result. However, the indirect consequences on other systems needs to be anticipated and watched for. Musculoskeletal problems may include progressive weakness and orthopaedic complications. These, in turn, can negatively impact the respiratory system, with resultant insufficiency and all of the symptoms that come with it (cough, dyspnea, fatigue, and headaches). Gastrointestinal symptoms are particularly common in children with progressive neurological decline, including feeding intolerance, aspiration, dysmotility, gastro-eosophageal reflux, and constipation. Particularly among children who are cognitively intact, the psychological effects of experiencing progressive deterioration, with resultant worsening of symptoms and decreased function, can be profound. Examining the child and family's coping on a regular basis is imperative. Finally, it should be recognized that both the direct and indirect effects of progressive neurologic decline can impact general energy level and sleep.

The principles of symptom evaluation



The child or young person requires an individualized symptom management plan, which includes a systematic approach to assessment, identification of reversible causes, treatment options, and plans to re-evaluate management strategies.

A clinical history of the symptom is essential and should include information about the nature and quality of the symptom, temporal factors, intensity, exacerbating and relieving factors, and associated sensations. This will ideally be obtained from the person who is

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experiencing the symptom, but information from other sources may be useful, and in the case of young and non-verbal children this may be the only source of information. This information helps to contextualize the symptom in its particular setting, for a particular individual, in a particular situation, which may not be useful for quantifying the symptom on one occasion but may be particularly helpful when monitoring the situation over time and the effect of interventions.

When formulating a management plan, it may be important to identify an appropriate place of care (ward, day care, home, local hospital, or hospice). Since some places of care may restrict the investigation and management options for a symptom, this discussion focuses the child and their family on goal-directed management strategies that are aimed at relieving discomfort, as opposed to intervention at any cost. It highlights new choices for the child and family and emphasizes the proactive approach of palliative care rather than the misconception held by some families and professionals that palliative care means 'everyone has given up'.

It is important to anticipate symptoms before their onset, to provide the child and family with information about them, and to balance the risks and benefits of the treatment for the individual, at their stage of the illness. This empowers the child and family, helps to relieve some of the anxiety about what the future holds, and allows them to make plans and be involved in care.

For clinicians providing palliative care, developing experience in managing malignant and non-malignant disease states facilitates appreciation of the differences in disease trajectory between these two entities, which may affect the assessment and management of symptoms. For example, opioid-sensitive pain management in a young person with metastatic Ewing's sarcoma, where the disease progresses rapidly over days or weeks and the young person dies shortly afterwards, may involve rapid escalation of medication. Here, the main focus is on ensuring a route of administration that is minimally invasive yet allows rapid titration of analgesia and the ability to re-evaluate frequently. In contrast, pain management in a young person with a neurodegenerative condition, which the child may live with for many years, will focus on chronic pain management strategies that are minimally invasive, minimize side effects, maximize functional ability, and may require consideration of long-term medication effects, such as tolerance and dependence issues.

Pharmacological strategies have their own risks and side effects which must not outweigh the discomfort of the symptom that they are addressing. Non-pharmacological measures include providing psychological support through information and advanced care planning; administration of hot or cold packs; massage; play therapy; distraction therapy; hypnotherapy; physiotherapy; occupational therapy, and other complementary therapies.

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It is important that health professionals understand how children and young people view their symptoms. Everyone—patients, parents, and professionals—makes value judgements about the burden of symptoms which are based on their personal views. It is essential to appreciate how the child or young person views their symptoms, and to acknowledge the meaning of the symptom to them, if rational approaches to management are to succeed (Box 14.1).

Box 14.1 Case example: 'Gina'

This is the story of Gina, provided by the G Frager and J Collins, authors of the first edition of this chapter in the *Oxford Textbook of Palliative Care for Children*, 2006.

Gina was a savvy, independent 16-year-old, who enjoyed cheerleading and a wide circle of friends. She experienced many difficulties during treatment of Ewing's sarcoma. A feisty individual, she toughed it out through an extensive limb salvage procedure, chemotherapy, and radiation therapy, staying hopeful and celebrating the positive aspects of her life.

Gina's course was complicated by obstructive uropathy, necessitating a nephrostomy tube and external bag. Metastases to her lower spine resulted in urinary retention for which she self-catheterized for the kidney not drained by the nephrostomy tube. Gina had significant neuropathic pain managed with a combination of opioids and multiple adjuvants, from which she had significant adverse reactions, requiring many changes to her analgesic regiment.

Despite achieving good analgesia and managing her nephrostomy and self-catheterizations, Gina became despondent because of faecal incontinence secondary to involvement of the cauda equine from spinal cord metastases. She expressed that she felt life was not worth living because of the incontinence.

With various techniques, Gina was able to achieve fecal continence, enabling her to relax in a hot tub, a goal Gina had for her trip to California with her boyfriend, father, and stepmother.

Symptom measurement in children



There is an emerging interest in healthcare quality, which includes patient safety, patient experience, and the effectiveness of care, driving the research and service provision agenda towards measuring outcomes that reflect the impact of healthcare on patients and their families. In the area of paediatric palliative symptom assessment these

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strategies and tools largely focus on cancer symptoms. They include quantitative and qualitative methodology, self-report versus parent, and professional proxy report observational tools, multidimensional and individual symptom scales, and quality-of-life assessment tools (see Table 14.3).

Table 14.3 Summary of some of the features of assessment tools		
Type of assessment tool	Characteristics of the tool	Example
Quantitative assessment tools	<ul style="list-style-type: none"> • May be self-report or observational • Can be used to measure the intensity of a symptom (e.g. pain) • Biophysical measurements or perceived severity states 	<ul style="list-style-type: none"> • Visual analogue scales such as modified Likert Scale and Faeces Pain Scale
Qualitative assessment tools	<ul style="list-style-type: none"> • Descriptive interview data • Patients, carers, and professionals • Focus group interview or individuals • Several different techniques for data analysis (e.g. pragmatic and semantic content analysis or phenomenology) • Questionnaire or participant observation 	<ul style="list-style-type: none"> • Fatigue in children and adolescents with cancer • Focus group data analysed by pragmatic and semantic content analysis techniques • Pain in paediatric oncology: interviews with children, adolescents, and their parents. Structured interview data collection with participant observation

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Self-report tools	<ul style="list-style-type: none"> • Developed for age-appropriate understanding • Suitable for cognitively unimpaired children and older children 	<ul style="list-style-type: none"> • Modified constipation Assessment scale, piloted in children
Carer/parent/professional report scales or mixed-dyad assessment tools	<ul style="list-style-type: none"> • Can allow observational data to be compared with self-report scales and analysed together 	<ul style="list-style-type: none"> • Qualitative study comparing parents' and children's perceived experience of nausea and vomiting
Individual symptom assessment tools	<ul style="list-style-type: none"> • Examine various aspects of one particular symptom • Modified to be age-appropriate for children 	<ul style="list-style-type: none"> • Dyspnoea assessment by means of pictorial scale (Dalhousie Dyspnoea Scale)
Multidimensional assessment tools	<ul style="list-style-type: none"> • Ideally used in self-report setting and age-appropriate examination of the frequency, intensity, and related distress of various symptoms 	<ul style="list-style-type: none"> • Memorial Symptom Assessment Scale (MSAS) validated in the 10-18 years and 7-12 years age groups
Quality-of-life assessment tools	<ul style="list-style-type: none"> • Can be retrospective viewpoint of carers or current view of children and carers • Multifaceted components of life, including physical, emotional, 	<ul style="list-style-type: none"> • Paediatric Quality of Life Inventory (PedsQL)

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	psychological, and social aspects	
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Challenges to comprehensive symptom evaluation



Many of the challenges that are faced when assessing symptoms experienced by children and young people have been described in the children's cancer care literature, but these issues can be extrapolated to non-malignant disease. They include issues that are intrinsic to childhood, such as developmental diversity, variation of the symptom experience in different environmental settings, the vulnerability of adolescents to symptom distress, and the lack of recognition of the child as part of a family. In addition, measuring symptoms in children and young people using appropriate assessment tools, and engaging children and young people in the reporting of symptoms, involve overcoming barriers.

The concept of developmental diversity recognizes that children of different ages and at developmental stages appraise and respond to an illness experience in different ways. This has been researched in children who are not cognitively impaired, which poses other challenges. Younger, non-verbal children who lack the ability to express themselves in expected ways, and who also have fewer coping strategies, are vulnerable.

Current research also suggests that adolescents have an increased vulnerability to symptom distress, perhaps because they are better able to articulate their distress. This group may choose to 'fight the treatment' rather than the disease, leading to challenges with regard to the assessment and management of symptoms.

If developmental diversity is not addressed, there is potential for disparities to occur in the health outcomes for children and adolescents of different developmental stages, even if the therapies are similar. Therefore, it is important to identify age-appropriate assessment tools and interventions.

Another challenge to the accurate assessment of symptoms, even if the appropriate tools are available, is the lack of engagement of children and young people in the process of reporting symptoms. The Advanced Symptom Management System (ASyMS[®]) developed by Gibson and colleagues is an assessment tool that involves the utilization of a two-way process of symptom management by health professionals and children or young people (18). With the help of this tool, the health professional informs the young person of the possible symptoms that they may encounter, and in turn the young person reports back to the professionals information about the symptoms that they are experiencing. This continued dialogue facilitates the provision of an individualized plan. It

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also utilizes technology, such as mobile phones and the Internet, which is familiar to the children and young people for whom the tool is designed.

Some symptom assessment tools, although they have been shown to be reliable and valid, are conceptualized as tools for adults and may lack essential aspects of children's symptom experience. To address these shortcomings, some adult assessment tools are utilized for older children (e.g. the Symptom Distress Scale), while others have been modified for children (e.g. the Memorial Symptom Assessment Scale, validated by Collins and colleagues (13)). There are also new tools which have been developed specifically for children (e.g. the Symptom Screening in Pediatrics tool (19)).

Assessment tools for the younger age groups have several limitations. Some believe that self-assessment tools are of no value for infants and young children who cannot understand and describe their symptoms, and therefore direct observational techniques or parent proxy reports are necessary to determine the symptoms. Research has demonstrated that young children may have difficulty in responding to items on assessment questionnaires, and greater variability in responses can be expected, suggesting that some tools designed for these age groups may not reach an acceptable level of internal consistency. Younger children may require assistance completing the questions, which also queries the validity and reliability of tools available to measure symptoms in this age group. Despite limitations, the routine use of an assessment tool in this age group may be preferable to using no tool at all; at least the former may provide a way to monitor a symptom over time.

There has been variation in the reported accuracy of parent proxy reports, as demonstrated by the fact that, while some studies report a correlation between parent and child's report of symptoms (when both are available), others report disagreement between them. Nonetheless, most studies seem to demonstrate a reasonable correlation between parent proxy reports and children's reports of symptoms, and they are therefore still a valuable information resource in children.

Relatively newer tools go beyond the measurement of a single symptom experience, attempting to capture more information about the true complex symptom experience of patients. Multidimensional assessment instruments, which report on various aspects of symptoms (e.g. frequency, severity, degree of distress), may be more useful measures, as they reflect the associated distress, which documentation of the frequency alone cannot demonstrate. It has been noted that clusters of symptoms may often appear together and influence one another, such as the constellation of pain, sleep disturbance, and anxiety. As a result, measurement tools have been designed that measure groups of symptoms and are able to demonstrate interrelationships between symptoms (20). Finally, overall quality-of-life measurement instruments (e.g. the Paediatric Quality of Life Inventory (PedsQL)) are able to provide a

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temporal document of physical symptoms, and therefore contribute to data about the overall experience of the child and their family.

Among clinicians who lack experience in the palliative care setting, some fears and misperceptions still persist about the use of medications such as strong opioids and benzodiazepines. When these fears are coupled with symptom assessment techniques that have a limited evidence base (and are therefore not commonly used in practice), and which clinicians are not experienced enough to utilize effectively, there is a tendency to underestimate symptoms or even ignore them. This can be damaging to the palliative care process, as it reinforces the belief of the patient and the parents that nothing can be done to relieve the distress and that it must be endured.

Novel approaches to symptom evaluation



The development of mobile telephone and computer technology is now deeply embedded in society, perhaps even more so among the paediatric age group and their relatively young caregivers. Brock, Wolfe, and Ullrich recently conducted a review of such innovations, including mobile apps and electronic programmes, for symptom assessment and their relevance to paediatric palliative care (21). The review demonstrated the applicability of such programmes within the United States, where 95% of Americans own a mobile phone. The review acknowledged that such devices are already frequently used to diarize appointments, document advice, and chart medication administration.

Several web-based approaches have been evaluated, acknowledging that whatever method is used, it must be tested, validated, and allow for the collection of symptoms and health-related quality of life data. The review noted two studies that found no difference in data equivalence scores between information provided by patients using electronic apps versus paper and pencil; the number of recording errors were in fact greater in the paper recording group, in one of the studies, compared to those in the app group. The results therefore support that the electronic data can then be integrated into the medical record, where information becomes available to the clinical team, regardless of where the child or young person may be physically, allowing prompt clinical intervention, particularly when patients and parents may be reluctant to make direct contact with the palliative care team. The sharing of such information allows the team to assess symptoms before a crisis point, reducing the risk of escalating symptoms (Table 14.4).

Table 14.4 Methods of electronic paediatric symptom assessment

Method	Advantages	Disadvantages	Applicability to PPC
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<p>Online tools</p>	<ul style="list-style-type: none"> • Choice of PROs • Can be used in clinical and research environments • Allows wider psychological discussions • Can be used prior to PPC team • Increases parent and clinician satisfaction 	<ul style="list-style-type: none"> • Set-up costs for development and implementation. • Requires inclusion into the medical record. 	<ul style="list-style-type: none"> • Allows the CYP and parents to identify measures of significance • Facilitates discussion with the clinical team • Appropriate for CYP with progressive disease
<p>Mobile apps</p>	<ul style="list-style-type: none"> • User friendly for CYP and parents • Applicable to multiple symptoms • Direct patient/clinician contact • Bespoke interventions on non-pharmacological treatment 	<ul style="list-style-type: none"> • Variability of measures with the apps • Limitation of operation systems being able to host the app. 	<ul style="list-style-type: none"> • Supports and develops CYP and parent coping skills • The availability of online training for mindfulness, guided imagery and relaxation • Appropriate for research
<p>Paediatric Palliative Care (PPC) Team</p>	<ul style="list-style-type: none"> • Efficient assessment during in-person visit • Multidisciplinary team approach to assessment 	<ul style="list-style-type: none"> • Challenges of meeting the needs of CYP outside of hospital • Requires a workforce and capacity to 	<ul style="list-style-type: none"> • Provides human contact and establishes relationships • Can be combined with online

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	<ul style="list-style-type: none">• Supportive data for the role of the team in CYP and parent satisfaction	meet such needs <ul style="list-style-type: none">• Home care is not universally available	tools and mobile apps. <ul style="list-style-type: none">• Provides an assessment and treatment plan
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PRO—Patient Reported Outcome; CYP—Child or Young Person; PPC—Paediatric Palliative Care

Whilst evolving technology is exciting, its development remains in its infancy. A prerequisite for embracing technological development is to recognize that these advances do not replace the interpersonal relationships of the paediatric palliative care team. This risk can be minimized by the wider engagement of patients, families, and professionals in the development of such technology.

Symptom evaluation in paediatric palliative care: Areas for further study



As was highlighted in the first edition of this book, when a US-based study revealed that 76% of parents who were interviewed had reported that avoiding pain and discomfort was important in guiding their decision to withhold life-sustaining intensive-care treatment, it is a sad reflection of our practice that inadequately managed symptoms are a driver for this difficult decision. The reasons for the importance of symptom assessment, the vast knowledge that has already been accumulated in this area, the limitations of the current evidence base, and the avenues for development of appropriate assessment tools have already been discussed in this chapter. However, the following areas still require further development.

- Raising awareness of the importance of paediatric palliative care as a subspecialty of paediatrics dedicated to ensuring a minimum standard of care for life-limited children, and equity of access to child-centred and family-focused care. National recognition of the needs of this group of children—for example, as highlighted by the UK Department of Health direction-setting document, *Better Care, Better Lives*, 2008—is an effective means of addressing carers' and professionals' misconceptions about the symptom assessment aspect of these children's management, and encouraging a more aggressive approach to high-quality expertise.
- Developing seamless continuity of service provision across health, education, and social care settings, so that individualized symptom management plans for a child can be actioned across various care settings.

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- Development of an organized collaborative research agenda is an important means by which a unified approach to accumulating a high-quality evidence base can be achieved in this area despite the small numbers of children in any one clinical centre.
- Symptom assessment needs to be evaluated from the child's viewpoint, including assessment of symptoms in children with non-malignant disease, and in non-verbal and cognitively impaired children. Good-quality assessment tools need to be developed and validated in these populations and made user-friendly, cheap, and accessible, in order to facilitate their use in everyday clinical practice.
- Development of robust means of quality assurance is essential. This will involve developing standardized care practices that can be evaluated and audited. Quality outcome measurements in symptom assessment can be applied not only to service evaluation and improvement, but also to research trials.
- Development of education and training goals is a priority for dissemination of good practice and evidence-based symptom assessment strategies. Grant allocation for clinical research should focus on the provision made by applicants for adequate dissemination and implementation of findings. Further to this, responsible commissioning of services by governments or local-level governing bodies should also ensure the sharing of best practice, and training in and implementation of novel symptom assessment strategies.

It can be seen that symptom assessment in children's palliative care presents challenges that require specific knowledge, skills, and attitudes among those who provide care, rather than merely awareness of the disease and the side effects of treatment. Accurate assessment is the first step in the management of symptoms in children with life-threatening illness, and it is acknowledged that this needs to be child focused as well as disease- and therapy-specific. Management strategies can reflect all of these elements and deliver therapy with the same aggressive, evidence-based approach that is used for curative treatment.

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