# Paediatric Palliative Care

40

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## Introduction

Don't you ever laugh as the hearse goes by,
For you may be the next one to die.
They wrap you up in a big white sheet
From your head down to your feet.
They put you in a big black box
And cover you up with dirt and rocks.
All goes well for about a week,
Until your coffin begins to leak.
The worms crawl in, the worms crawl out,
The worms play pinochle in your snout,
They eat your eyes, they eat your nose,
They eat the jelly between your toes.
A big green worm with rolling eyes
Crawls in your stomach and out your eyes.
Your stomach turns a slimy green,
And pus pours out like whipping cream.
You'll spread it on a slice of bread,
And this is what you eat when you are dead. [1] (Child's nursery rhyme parts of which date back to the Crimean War)

Death, much like sex, is a topic that adults find hard to discuss among themselves let alone with their children [2]. The conversation may be painful for both parties and there is a natural desire to protect children from harm. This ignores the fact that death is an integral part of childhood and that a failure to have an open conversation about childhood mortality can lead to significant misunderstandings at both an individual and societal levels.

## Service Development and Epidemiology

Paediatric palliative care 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 or young person and support for the family. It includes the management of distressing symptoms, provision of short breaks and care through death and bereavement [3].

This definition has profound implications for the provision, planning and funding of services for children and their families with life limiting or life threatening conditions.

Palliative care for children is a relatively new speciality, with the first children's hospice, Helen House, opening in Oxford (UK) in 1982 [4]. Until the middle of the 1990s there was only one consultant specialising in Paediatric Palliative Medicine in the UK. It is a testament to families and professionals that there has been a massive increase in resource provision since then but it is sobering that there are still less than ten senior doctors with a paediatric training working as consultants in this field in the United Kingdom today.

Cancer is the commonest cause of death in children and the second commonest cause (after accidents and violence) in teenagers and young adults in industrialised countries. Cancer represents approximately 25 % of all childhood deaths. This equates to approximately 250 deaths in children aged under 16 years in a country of 63 million [5]. Death however is a very crude marker of need and it should be emphasised that End of Life Care is a small part of a greater palliative care package.

## Models of Disease Trajectory

It is helpful to consider what the disease trajectory for children with a terminal diagnosis is and how it has changed over time. A traditional model views the child's journey as a gradual transition from cure to palliative care but it is also possible to construct a model that is dynamic and more accurately reflects the current situation where a child and family dip in and out of palliative care services as required. In this latter model palliation and curative intent are not seen as distinct,

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separate entities, rather as a reflection of a patient's need at that moment. Both models reflect that palliative and curative care should are two sides of the same coin and there is no defined start or endpoint to palliation (see Fig. 40.1).

Childhood cancer is now probably best thought of as a chronic disease. The majority of relapsed solid tumours still remain incurable but there has been a dramatic increase in the number of options open to families at the time of relapse. It is important to acknowledge that many of these have arisen because families have overcome the fatalistic attitudes of doctors and nurses.

The burden of therapy at the time of relapse should not be under estimated. It may well include further surgery consolidated with myeloablative chemotherapy and stem cell return (an autologous bone marrow transplant), radiotherapy or a phase I/II trial.

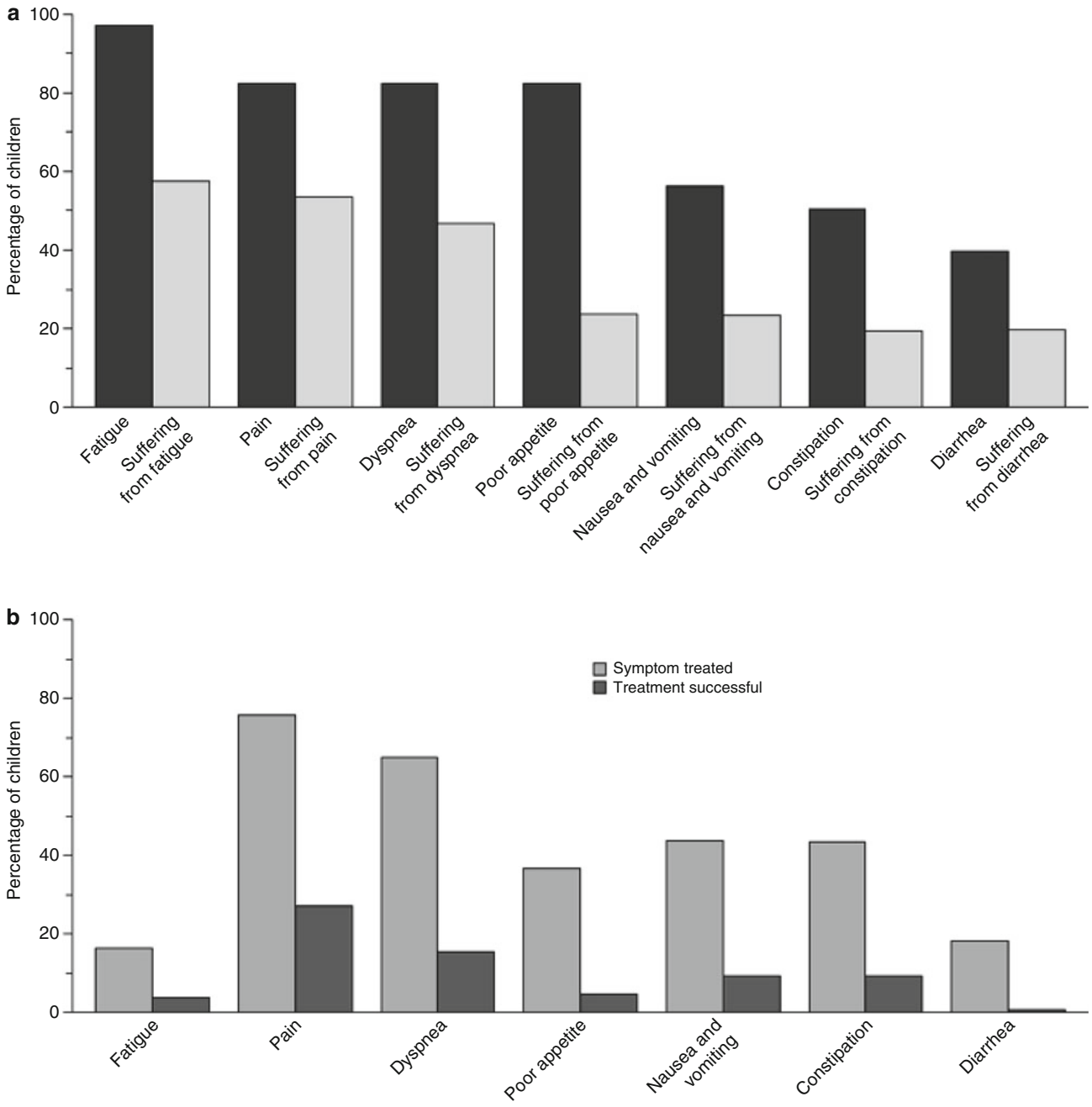


**Fig. 40.1** Models of disease trajectory

## Symptoms at the End of Life

How a child feels and the burden of care has been well researched on both sides of the Atlantic. Wolfe et al. [6] looked at symptoms at the end of life in children dying in Boston. She noted not only the presence of symptoms but also whether they caused suffering. Her team also showed how limited teams were in treating symptoms that did cause suffering (see Fig. 40.2). Furthermore doctors and nurses were unaware of many of the symptoms families described (see Fig. 40.3).

Liben and Goldman [7] found in a UK study that while the symptom constellation was similar that the presence of a dedicated symptom care team enabled a greater success in managing symptoms at the end of life.

A further UK study [8] showed the symptom constellations differed by tumour type when grouped together by type (solid tumour, brain tumour or leukaemia/lymphoma) Surprisingly the symptoms suffered by children with solid tumours and leukaemias was almost identical while children with brain tumours have their own, unique, pattern of symptoms (See Fig. 40.4).

Symptoms should be actively sought and treated. They will change over time through the evolution of the child's disease. Differing members of the multi disciplinary team should be employed as the child and family will disclose different concerns to different professionals. The response of symptoms to interventions should be also be clearly noted. This process should be repeated frequently. Using the mnemonic "I am fine" will allow a systematic approach (See Fig. 40.5)

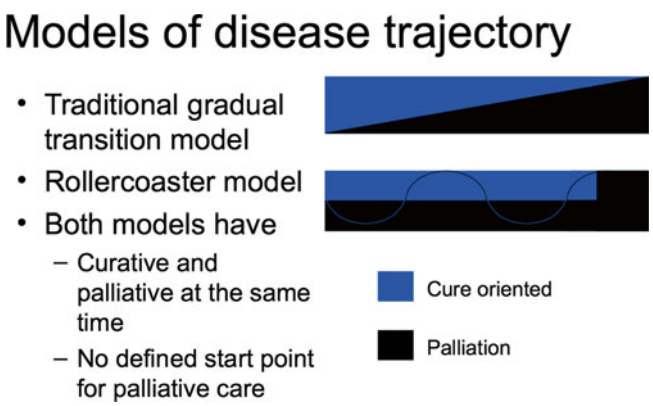


**Fig. 40.2** The degree of suffering from and the success of treatment of specific symptoms in the last month of life. Panel **a** shows the percentages of children who, according to parental report, had a specific symptom in the last month of life and who had "a great deal" or "a lot" of suffering as a result. Panel **b** shows the percentages of children who, according to parental report, were treated for a specific symptom in the last month of life, and in whom treatment was successful (rather than "somewhat successful" or "not successful")

## Barriers to Care

Access to expertise for children and their families is inequitable even within countries with a socialised health care service [9]. Many factors may be associated with this: personal, cultural, social or institutional. It is crucial that all forms of discrimination should be eliminated for this particularly vulnerable patient group.

If a child wishes to enter a phase I/II trial this should be provided as close to home as possible with a recognition that time is limited. It is difficult to balance the hope that an early phase clinical trial offers with the reality that these will very rarely extend the quality or quantity of life.

## Communication

Discussion with the family around the time of relapse is crucial. It should be clear and recognise that the child and family are experts in their own disease. These conversations should be seen as a process rather than a single event and that the parent's and child's needs will need differing approaches even if they have the same requirements.

**Fig. 40.3** Discordance between the reports of parents and physicians regarding the children's symptoms in the last month of life. Data were missing for ten children for whom there was no documentation of clinic or hospital visits in the last month of life and for one child whose records were not available for review.*CI* denotes confidence interval. [a]McNemar's test was used

| Symptom | Reported by parent but not by physician (N = 92) | Reported by physician but not by parent (N = 92) | Kappa statistic (95 % CI) | P Value[a] |
|---|---|---|---|---|
| | Number (percent) | | | |
| Fatigue | 44 (48) | 1 (1) | –0.02 (–0.07 to 0.02) | <0.001 |
| Pain | 15 (16) | 11 (12) | 0.10 (–0.12 to 0.32) | 0.56 |
| Dyspnea | 19 (21) | 10 (11) | 0.10 (–0.11 to 0.31) | 0.14 |
| Poor appetite | 33 (36) | 1 (1) | 0.29 (0.15 to 0.43) | <0.001 |
| Constipation | 31 (34) | 7 (8) | 0.16 (–0.02 to 0.33) | <0.001 |
| Nausea and vomiting | 25 (27) | 18 (20) | 0.06 (–0.14 to 0.26) | 0.36 |
| Diarrhea | 20 (22) | 8 (9) | 0.31 (0.12 to 0.51) | 0.04 |

| Symptom | Entry into study | | | Last month of life | | |
|---|---|---|---|---|---|---|
| | CNS vs leukernia/ lymphoma | CNS vs othersoild | Leukernia/ lymphoma vs other soild | CNS vs leukernia/ lymphoma | CNS vs other soild | Leukernia/ lymphoma vs other soild |
| Pain | – | – | – | NS | p < .01 | NS |
| Weakness | p < .05 | p < .01 | NS | – | – | – |
| Weight gain | p < .05 | p < .01 | NS | p < .05 | p < .01 | NS |
| Weight loss | – | – | – | NS | p < .01 | p < .01 |
| Anorexia | – | – | – | NS | p < .01 | p < .01 |
| Swallowing | – | – | – | p < .01 | p < .01 | NS |
| Excess secretions | – | – | – | p < .01 | p < .01 | NS |
| Headache | p < .05 | p < .01 | NS | p < .01 | p < .01 | NS |
| Dizziness | NS | p < .05 | NS | NS | p < .01 | NS |
| Convulsions | – | – | – | NS | p < .01 | NS |
| Mobility | p < .01 | p < .01 | NS | p < .01 | p < .01 | NS |
| Speech | p < .05 | p < .01 | NS | p < .01 | p < .01 | NS |
| Vision/hearing | p < .01 | p < .01 | NS | p < .01 | p < .01 | NS |
| Anemia | p < .01 | p < .01 | NS | p < .01 | p < .01 | NS |
| Bleeding | p < .01 | NS | NS | p < .01 | p <. 05 | NS |
| No.of significant differences | 8 | 8 | 0 | 9 | 14 | 2 |

**Fig. 40.4** Significance of differences in symptom prevalence between different tumor groups at entry to study and in the last month of life (adjusted for multiple testing). *NS* indicates not statistically significant

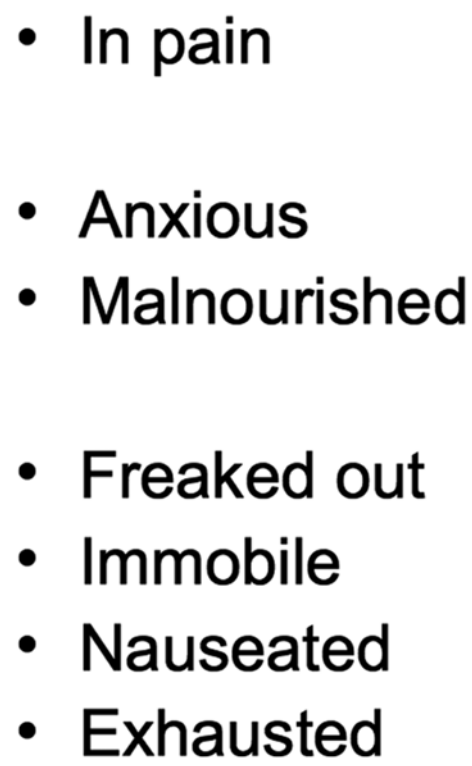


**Fig. 40.5** I am fine

It is now almost universal that parents will be part of a wider social network that is world wide and disease specific. They may well have information on very early trials that is not readily available to clinicians. This has shifted the consultation away from a paternalistic "doctor knows best" model into a much more collaborative conversation in which the doctor's role is to interpret the information that patients have and to direct them towards resources they may not have seen. There should be humility on both sides and a recognition that a second opinion may be useful.

Talking to children about dying is often clouded by mutual pretense [10], here each party understands what is happening but doesn't talk about it. Dangerous topics are avoided, space is given to allow individuals to leave conversations if the pretense is in danger of being shattered. This tactic avoids confirmation of a known, terrifying reality for both parties. This may be a perfectly reasonable way to avoid trauma between individuals but may lead to huge misunderstandings and avoidable fear if it is unrecognised by health care professionals.

## Guidelines on Talking about Death and Dying

There is no "right way" of talking to children (and to the parents and siblings of children) who are dying. Some general pointers can be noted though. (1) It is of paramount importance is to listen to the child and hear what they are saying (and not saying). (2) Have an understanding that children (and their parents) can hold mutually incompatible rational and scientific thoughts about death and dying at the same time. (3) The need and desire for information will ebb and flow over the disease trajectory. (4) Children want to keep those they care about around them-a desire that makes them unlikely to vocalise their needs if they see this as incompatible with their loved ones wishes.

This doesn't mean the fundamental question is "to tell or not to tell" but rather "what to tell, when to tell and who should tell". Children want to know about what their illness

and will try to figure it out for themselves if they are not told. This is especially important in an era when even toddlers can manipulate iPads. However, even prior to the information revolution, a visit to the play room on a children's cancer ward when parents were not present, would quickly confirm that children readily shared detailed knowledge of their disease and prognosis with one another.

In a situation when parents do not wish to discuss aspects of their child's treatment or death with them it is extraordinarily important and beneficial to find out why. Quite often fundamental misunderstandings about what the prognosis is or what treatment entails are uncovered. Again listening to a parents fears and concerns is vital as is an understanding that communication is a joined up process rather than a series of one off events. It is common that the views of individual parents may diverge, not only from one another but also from their child. It is vital that all sides are heard and respected. It is helpful to open dialogue by acknowledging how difficult and almost unreal, the conversation is. It is often also helpful to refer, at some point during the consultation, to previous experience of looking after families in a similar situation. Statements such as "I have found other parents find it really scary to think about their child's death" or "In the past children have tried to protect their parents in this situation-do you think this is happening" help families to engage in topics that they may be avoiding in a bid to protect themselves, their partner or their children. Asking "How would you like me to explain this to your child" is constructive, helpful and can lead to further clarity. Do not expect to have resolution of all concerns after a single consult. Suggest that further discussions with the child, either with you present or not, may help. This need not be straight away-but the groundwork for all future conversations will have been laid. It is vital to have the child's key worker present at as many of these consultations as possible. The reality though is that a parent or child may take advantage of a "corridor conversation" to garner further thoughts. A regular multi disciplinary team meeting allows information sharing and the planning of further consults. Above all, remember a child's needs may well not reflect your own needs or your perception of what their needs are. They may also be very different from the needs of their parents, siblings and grandparents.

Careful notation of a child's and family's wishes are paramount. This will avoid painful repetition of basic information and inappropriate and unwarranted health care interventions. Scotland is the first country in the world to have a legally mandated end of life care plan for children [11]. It allows clear and precise documentation of what a child and family want to happen at the end of life. It is owned by the family and is carried by them and is seen as a positive intervention. It was deliberately designed to show all professionals (including police, ambulance and school) what will be done rather than what wont be done and is used in community, hospice, hospital, education and home environments. Forms, parent information sheets and education packages are all available on line [12].

**Conclusion**

Children dying from cancer form a uniquely vulnerable group. They and their families deserve the same rigour of thought and delivery of care as children who have curable disease. They deserve rapid access to the latest information on novel therapies and symptom control. Access to specialized services should be equitable and independent of class, ethnicity, socio-economic status and geography. Care should be of the highest quality and teams providing palliative care should be driven by a strong evidence base and welcome routine external audit of performance. There should be an institutional and governmental agenda to provide safe and sustainable services.

Winston Churchill once remarked that the mark of a country's civilization was the way it cared for its prisoners. This was taken up by John FitzGerald Kennedy and modified to assert that the mark of a country's civilization was the way it cared for its pensioners. In the Twenty-First century surely the mark of a country's civilization is the way it cares for its dying children.

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