

CARING FOR CHILDREN WITH LIFE-LIMITING AND LIFE-THREATENING ILLNESSES: WHAT THE GP SHOULD KNOW

'Life has meaning only in the struggle. Triumph or defeat is in the hands of the Gods. So let us celebrate the struggle.' (Swahili warrior song featured in the 1992 film *Lorenzo's Oil*, the story of parents' struggle to deal with a child suffering from adrenoleukodystrophy.)

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Death in childhood is not regarded as normal, but despite all our efforts at 'saving children', death in childhood still happens. There is also a subset of children for whom death in childhood is anticipated and can be regarded as a normal outcome of the child's condition (e.g. trisomies, inoperable heart conditions). The death of a child is regarded by the *Diagnostic and Statistical Manual of Mental Disorders (DSM)* as a 'catastrophic stressor' for a parent – on par with experiencing a 'natural disaster'.¹ Yet, why do we learn so little as health professionals about how to deal with it?

The death of a child is regarded by the Diagnostic and Statistical Manual of Mental Disorders (DSM) as a 'catastrophic stressor' for a parent – on par with experiencing a 'natural disaster'.

As a paediatric registrar I tried to get my head around the different types of adrenoleukodystrophy, but it did not prepare me to manage my first case. I remember looking it up in a paediatric textbook at the time, but it seemed as though the book ended where I needed input the most. It informed me that prognosis was poor, that there was no known cure (although there were several experimental therapies and trials) and that care was 'mainly supportive'. But what did this support entail? How did one manage the relentless deterioration and unremitting seizures? How would I know if the child was in pain? How would I know when the end was near? When should I stop treating the pneumonias? What should I say to the exhausted parents?

Then I learnt about paediatric palliative care (PPC), a rapidly growing new specialty worldwide that addresses the care of children with life-limiting and life-threatening illnesses (LLTI). Although not yet recognised as a specialty in South Africa, the 'palliative care approach' is increasingly being taught in medical schools. Even if recognised as a specialty it is unlikely that specialists will be able to supervise the care of all children with palliative care needs. One of the key principles of PPC is to provide care as close to home as possible, which is a view shared with family medicine and primary health care. It is crucial, therefore, that every general practitioner and paediatrician has the core knowledge and skills to manage these children with input from palliative care specialists where necessary (and available).

Definitions

The following terms are key to the understanding and practice of palliative care for children:

Paediatric palliative care (PPC). The WHO states that PPC 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 uses available community resources; it can be successfully implemented even if resources are limited. It can be provided in tertiary care facilities, in community health centres and even in the child's own home.² Other definitions include a focus on the enhancement of the quality of life, the provision of respite and bereavement care.³

Life-threatening. A life-threatening disease is a disease that is potentially fatal, likely to result in imminent death. It includes conditions caused by both natural (e.g. infective) and unnatural (e.g. trauma) factors. Children with life-limiting illnesses may also develop life-threatening complications that are mostly responsible for their death.⁴

Life-limiting. A life-limiting illness is an illness which may not be immediately life threatening but which imposes limits on a person's quality and/or quantity of life.⁵

Symptom control. This is the control of distressing symptoms including pain and 'non-pain symptoms' using both drug and non-drug interventions to relieve suffering in children with LLTIs irrespective of the expected outcome (cure or death).

Psychosocial support. This is care concerned with the psychological and emotional well-being of the patient and their family/carers, including issues of self-esteem, insight into and adaptation to an illness and its consequences, communication, social functioning and relationships.⁶



End-of-life care. This is the care of a person during the last part of their life, from the point at which it has become clear that the person is in a progressive state of decline.⁵

Which children require palliative care and when?

Although end-of-life care is an important part of palliative care for children, palliative care begins when the diagnosis is made. There is no ‘palliative switch point’ as previously thought (Fig. 1), but rather an integration of active cure-focused treatments with palliative care from the time of diagnosis (Fig. 2).⁷

Although end-of-life care is an important part of palliative care for children, palliative care begins when the diagnosis is made. Understanding the disease trajectories for different conditions helps to guide decision making in terms of balancing ‘active treatment’ with ‘palliative care’.

As disease advances (points A, B, C) the amount of aggressive cure-focused treatment decreases and the amount of palliative-focused treatment increases. Understanding the disease trajectories for different conditions helps to guide decision making in terms of balancing ‘active treatment’ with ‘palliative care’.

The UK-based Association for Children with Terminal Conditions (ACT) has recognised four categories of children who would benefit from a palliative care approach.⁸ These categories are defined largely by their disease trajectories (Table I).

Journeying with families along the disease trajectory

Ideally palliative care principles should be applied from the time of diagnosis, through illness, ‘palliation’, terminal care, death and bereavement.

Diagnosis and illness

Diagnoses should preferably be made as early as possible so that appropriate care is provided from the outset. Unfortunately some children present late in their illness or a diagnosis is delayed or even illusive. A critical skill required in communicating the diagnosis (and prognosis) of an illness to a parent or caregiver is that of ‘breaking bad news’ well. Although ‘bad news is always bad

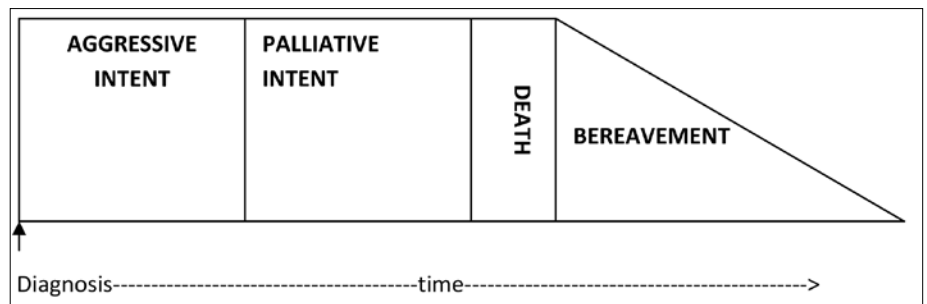


Fig. 1. Traditional palliative care services model.

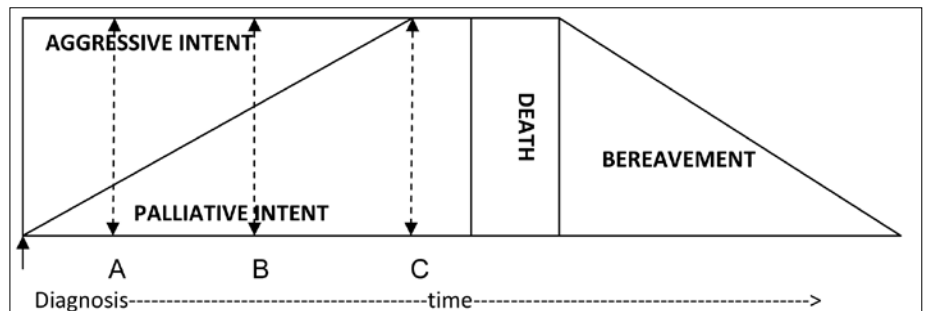


Fig. 2. Modern integrated palliative care services model (Fragar 1997).

Table I. Categories of life-limiting conditions
Group 1: Life-threatening conditions for which curative treatments may be feasible but can fail, e.g. cancer, irreversible organ failure.
Group 2: Conditions where premature death is inevitable, where there may be long periods of intensive treatment aimed at prolonging life and allowing participation in normal activities, e.g. HIV/AIDS on antiretrovirals, cystic fibrosis.
Group 3: Progressive conditions without curative treatment options, where treatment is exclusively palliative and may commonly extend over many years, e.g. many syndromes, inborn errors of metabolism, muscular dystrophy.
Group 4: Irreversible but non-progressive conditions causing severe disability leading to susceptibility to health complications and likelihood of premature death, e.g. severe cerebral palsy, spinal cord insult.

however well it is broken’, there are certain techniques that can be learned and even practised to limit the secondary trauma experienced by parents hearing for the first time that their child is ill.

A useful acronym that provides health care workers with some structure to plan this difficult conversation is BREAKS: setting the **B**ackground (includes information gathering); establishing **R**apport with the family; **E**xploring what is already known and would like to be known; **A**nnouncing the news in manageable chunks (preceded by a warning shot); **K**indling and validating the response; and then **S**ummarising the conversation.⁹

After hearing the news most patients/caregivers experience the same emotions as with any other loss. These emotions were first described by Elisabeth Kubler-Ross (Table II) in the context of bereavement

but are equally applicable at this stage of ‘anticipatory grief’.¹⁰

Table II. Elisabeth Kubler-Ross stages of grief
Stage 1: Denial and isolation
Stage 2: Anger
Stage 3: Bargaining
Stage 4: Depression
Stage 5: Acceptance

Coupled with diagnosis is the disclosure of the condition to the child. Not only is understanding what is happening to their bodies in an age-appropriate manner a basic child right but it also helps to relieve anxiety often caused by the fear of the unknown.

Children with incurable illnesses generally go through various stages of understanding of illness (Table III).¹¹

Table III. Understanding of incurable illness in children over time

- I am sick
- I am sick but I am going to get better
- I am going to keep on getting sick but I will still get better each time
- I am going to keep on getting sick and I won't get better
- I am going to die

Table IV. Stages of awareness

- Stage 1. Closed awareness:** The child is not aware of the problem and those who know it conceal it
Stage 2. Suspected awareness: The child is suspicious that something is wrong but is not certain
Stage 3. Mutual pretence: The child, family and health workers all know but no one talks about it
Stage 4. Open awareness: Everyone knows and is open about it

Table V. Gold standards framework for good end-of-life in children: PEPSI COLA



- P = Physical issues (pain and symptom control)
 E = Emotional issues (addressing fears and anxieties)
 P = Personal issues (relationship and spiritual issues)
 S = Social support (nutrition, finances, housing, etc.)
 I = Information (about the illness, possible end-of-life events)
 C = Control (choice of place of death, knowing what to do in an emergency)
 O = Out of hours/emergency (arrangements to ensure 24/7 access to care)
 L = Late (terminal care plan: last 48 hours)
 A = Afterwards (bereavement support)

trial of therapy (e.g. analgesics for pain) and reassessment.

A second challenge is that many symptom control drugs are 'off-licence' for use, especially in children under the age of 2 years. Balancing the risk of harm (side-effects) with the potential for doing good (relieving suffering) is the basis of decision making in these instances while we await the acquisition of evidence to support these practices.

A good death

While some may think that 'a good death' is an oxymoron, planning for as smooth a transition as possible (where death is inevitable and expected) lays a good foundation for a better bereavement experience.

A useful acronym provided by the Gold Standards Framework for addressing all the issues that need to be considered as part of good end-of-life care is PEPSI COLA (Table V).¹³

Terminal care may include the withdrawal of non-essential medicines (that are no longer helpful) and focusing on symptom control and comfort issues only. In some cases consideration of limiting and even withdrawing feeds (where the enteral route is not useable) may need to be made.

There are several benefits to limiting/withdrawing feeds at the end of life that include:

- induction of ketosis which releases endorphins (assists with pain control) and suppresses hunger

Although there is a natural tendency to want to protect children from bad news it is well known that children know a lot more than they are given credit for. A balance between giving too much information (that may frighten the child) v. giving too little information (that may cause a fear of the unknown) needs to be achieved by giving sufficient information. A useful technique to responding to 'difficult questions' posed by children (e.g. Am I going to die?) is to answer the question with another question (Why do you ask this?). This helps to determine what the child already knows and wants to know further.

In certain instances tangled webs are spun by families (who may include professionals in the collusion) in their attempts to conceal information from their child. Four stages of awareness are described (Table IV).¹²

In dealing with collusion a good approach is to find out (with permission) from each party what they already know and would like to know further and to encourage open and honest communication between child and family. A greater sense of intimacy and support is experienced when the stage of open awareness is reached.

Palliation and terminal care

The word palliate comes from the Latin *pallium*, meaning to cloak. This describes the

relief of suffering through symptom control – a key practice of palliative medicine.

The basic principles of symptom control are the following:

- determine and treat the underlying cause (where possible), including non-physical causes
- relieve the symptom without creating new symptoms and unwanted side-effects
- consider both drug and non-drug interventions
- consider whether treatment is of benefit to the individual patient.

A critical skill required in communicating the diagnosis (and prognosis) of an illness to a parent or caregiver is that of 'breaking bad news' well.

The first challenge in achieving symptom control in children is knowing whether a symptom is being experienced, especially in the non-verbal child. This can be addressed through careful observation and by asking whether the pathology would likely lead to the experience of the symptom in an older child or adult. There is also seldom harm in a

Children

- limitation of excessive secretions that may aggravate pulmonary oedema
- decrease in cerebral oedema
- decrease in abdominal distension caused by feed accumulation in a slowing bowel.

For some this practice is too controversial and emotive and may increase anxiety. In such instances it is best to reach a middle ground between excessive feeding and prolongation of dying and underfeeding and perceived starvation. In my experience 'feeding for comfort' as the child demands is the best and most natural compromise.

Bereavement

Good care does not end when the patient dies, but continues so as to support the family in their grief. Rituals, memorial services, support groups and individual counselling are all well recognised in providing bereavement support. Understanding the development of a mature 'death concept' in children is key to being able to support them in their bereavement journey in an age-appropriate manner.

The death of a child is a well-known risk factor for complicated grief, and timeous referral for professional counselling assistance should be made if this is suspected.

Conclusion

The GP plays an important role in the care of children with life-limiting and life-

threatening illnesses. He/she may have been the first professional to suspect or diagnose the condition and may be the professional the family turns to when 'there is nothing more that can be done' (from a curative perspective), thereby completing the circle. A palliative care approach recognises that

there is 'never nothing more that can be done', does not abandon the patient and family and provides care beyond death into bereavement.

References available at www.cmej.org.za

IN A NUTSHELL

- PPC is the active total care of the child's body, mind and spirit, and also involves giving support to the family.
- Ideally palliative care should begin at diagnosis and can be instituted alongside active cure-focused treatments.
- As disease advances the amount of aggressive cure-focused treatment may decrease while the amount of palliative-focused treatment increases.
- A critical skill required in communicating the diagnosis (and prognosis) of an illness to a parent or caregiver is the skill of 'breaking bad news' well.
- After hearing the news patients/caregivers may experience anticipatory grief which has similar stages encountered in bereavement as described by Elisabeth Kubler-Ross.
- When talking to children about illness and dying a balance between giving too much information (that may frighten the child) vs giving too little information (that may cause a fear of the unknown) needs to be achieved by giving sufficient information.
- Good symptom control using both drug and non-drug measures is a key aspect of the practice of palliative medicine.
- Planning a 'good death' where this is inevitable and expected lays a good foundation for a better bereavement experience.
- The death of a child is a well-known risk factor for complicated grief, and timeous referral for professional counselling assistance should be made if suspected.
- The GP plays an important role in providing palliative care for children. He/she may have been the first professional to suspect or diagnose the condition and may be the professional the family turns to when 'there is nothing more that can be done' (from a curative perspective), thereby completing the circle.

SINGLE SUTURE

Rabies rethink

Rabies may not deserve its reputation. It looks like the immune system sometimes defeats the disease on its own.

This month 8-year-old Precious Reynolds of California became only the sixth person known to survive rabies without receiving a vaccine within a few days. Doctors treated her with the experimental Milwaukee protocol, plunging her into a drug-induced coma to take her brain 'offline' while immune cells scrubbed away the virus. But the protocol is no miracle cure: tried with at least 35 people, only five had previously survived.

Some doctors reckon they may have been infected with a weaker strain of the virus and might have survived anyway. When Craig Hooper and Bernhard Dietzschold at Thomas Jefferson University in Philadelphia, Pennsylvania, gave mice that already had rabies a weakened strain of the virus, the mice cleared the infection on their own. Weaker strains are recognised more quickly by the immune system, which grants immune cells access to the brain, where they clear the virus.

Weakened strains could be an alternative to the Milwaukee protocol. 'We look at rabies as a lethal disease when it appears that there are exceptions,' says Hooper.

New Scientist, 25 June 2011, p. 5.