ABC of palliative care

Special problems of children

Ann Goldman

The death of a child has long been acknowledged as one of the greatest tragedies that can happen to a family, and care for seriously ill children and their families is central to paediatrics. It is only recently, however, that the needs for palliative care of children with life limiting illnesses and their families have been considered as a speciality within paediatrics: the most suitable approaches to care are still being developed, and the provision of services nationally is uneven and sometimes inadequate.

Which children need care?

Fortunately, deaths in childhood that can be anticipated, and for which palliative care can be planned, are rare. A recent joint report by ACT (Association for Children with Life Threatening or Terminal Conditions and their Families) and the Royal College of Paediatrics and Child Health offers the most up to date information about epidemiology (see box of further reading).

Paediatric palliative care may be needed for a wide range of diseases, which differ from adult diseases and many of which are rare and familial. The diagnosis influences the type of care that a child and family will need, and four broad groups have been identified.

Palliative care may be needed from infancy and for many years for some children, while others may not need it until they are older and only for a short time. Also the transition between aggressive treatments to cure or prolong good quality life and palliative care may not be clear—both approaches may be needed in conjunction, each becoming dominant at different times.

Numbers of children with life limiting illness

Annual mortality from life limiting illnesses
- 1 per 10 000 children aged 1-17 years

Prevalence of life limiting illnesses
- 10 per 10 000 children aged 0-19 years

In a health district of 250 000 people, with a child population of about 50 000, in one year
- 5 children are likely to die from a life limiting illness—Cancer (2), heart disease (1), other (2)
- 50 children are likely to have a life limiting illness, about half of whom will need palliative care at any time

Groups of life limiting diseases in children

Group	Examples
Diseases for which curative treatment may be feasible but may fail	Cancer
Diseases in which premature death is anticipated but intensive treatment may prolong good quality life	Cystic fibrosis HIV infection and AIDS
Progressive diseases for which treatment is exclusively palliative and may extend over many years	Batten disease Mucopolysaccharidoses
Conditions with severe neurological disability that, although not progressive, lead to vulnerability and complications likely to cause premature death	Severe cerebral palsy

Aspects of care in children

Child development

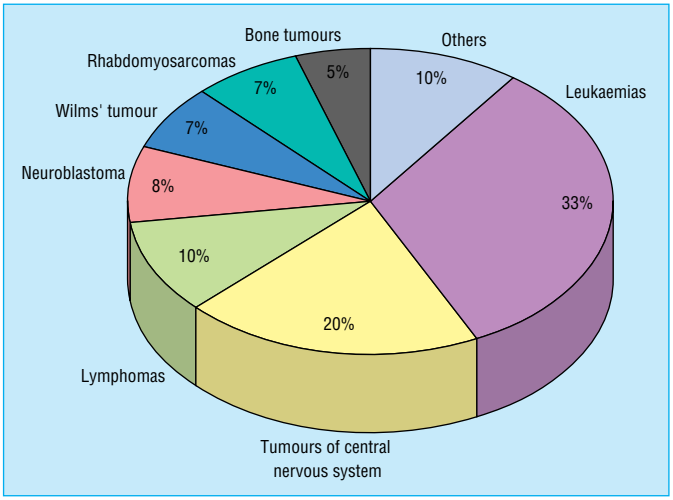
An intrinsic aspect of childhood is children's continuing physical, emotional, and cognitive development. This influences all aspects of their care, from pharmacodynamics and pharmacokinetics of drugs to their communication skills and their understanding of their disease and death.

Care at home

Most children with a life limiting disease are cared for at home. Parents are both part of the team caring for the sick child and part of the family and needing care themselves. As their child's primary carers, they must be included fully in the care team—provided with information, able to negotiate treatment plans, taught appropriate skills, and assured that advice and support is accessible 24 hours a day.

Assessing symptoms

Assessing symptoms is an essential step in developing a plan of management. Often a picture must be built up through discussion with the child, if possible, combined with careful observations by parents and staff. There are formal assessment tools for assessing pain in children that are appropriate for different ages and developmental levels, but assessment is more difficult for other symptoms and for preverbal and developmentally delayed children.



Range of malignant disease found in children

Methods of assessing pain in children
- Body charts
- Faces scales
- Numeric scales
- Diaries
- Colour tools
- Visual analogue scales
- Observation of behaviour

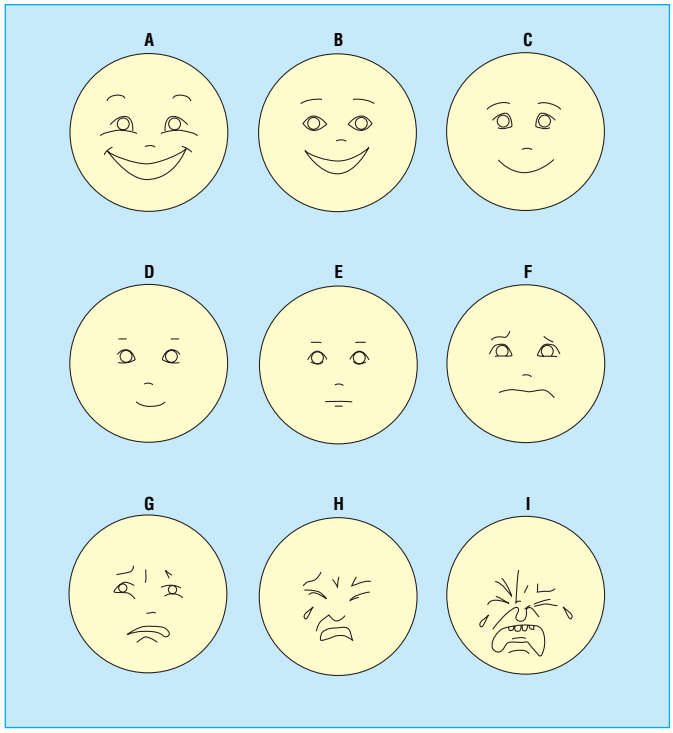
It is also important to consider the contribution of psychological and social factors for a child and family and to inquire about their coping strategies, relevant past experiences, and their levels of anxiety and emotional distress.

Managing symptoms

Many of the symptoms that children suffer and the approaches to relieving them have not been studied formally. Until definitive information becomes available, treatment is often based on clinical experience and adapted from general paediatric practice and palliative care of adults.

Many of the drug doses and routes used in palliative care are not licensed for children, and responsibility lies with the clinician prescribing them. In all situations the management plan should consider both pharmacological and psychological approaches along with practical help.

Children often find it difficult to take large amounts of drugs, and complex regimens may not be possible. Doses should be calculated according to a child's weight. Oral drugs should be used if possible, and children should be offered the choice between tablets, whole or crushed, and liquids. Long acting preparations are helpful, reducing the number of tablets needed and simplifying care at home. If an alternative route is needed some children find rectal drugs acceptable; they can be particularly useful in the last few days of life. Otherwise, a subcutaneous infusion can be established or, if one is in situ, a central intravenous line can be used. Parents are usually willing and able to learn to refill and load syringes and even to resite needles.



Faces scale used to measure pain effect in children (adapted from McGrath P. *Pain in children. Nature, assessment and treatment.* New York: Guildford Press, 1990)

Specific problems

Pain

The myths perpetuating the undertreatment of pain in children have now been rejected. However, most doctors lack experience in using strong opioids in children, which often results in excess caution. The World Health Organisation's three step ladder of analgesia is equally relevant for children, with paracetamol, dihydrocodeine, and morphine sulphate forming the standard steps.

Opioids—Laxatives need to be prescribed regularly with opioids, but children rarely need antiemetics. Itching with opioids in the first few days is quite common and responds to antihistamines if necessary. Many children are sleepy initially, and parents should be warned of this lest they fear that their child's disease has suddenly progressed. Respiratory depression with strong opioids used in standard doses is not a problem in children over 1 year old, but in younger children starting doses should be reduced.

Adjuvant analgesics—Non-steroidal anti-inflammatory drugs are often helpful for musculoskeletal pains in children with non-malignant disease. Caution is needed in children with cancer and infiltration of the bone marrow because of an increased risk of bleeding. Neuropathic pain may be helped by antiepileptic and antidepressant drugs. Pain from muscle spasms can be a major problem for children with neurodegenerative diseases and may be helped by benzodiazepines and baclofen. Headaches from raised intracranial pressure associated with brain tumours are best managed with gradually increasing analgesics. Although corticosteroids are often helpful initially, the symptoms soon recur and increasing doses are needed. The considerable side effects of corticosteroids in children—rapid weight gain, changed body image, and mood swings—usually outweigh the benefits. Headaches from leukaemic deposits in the central nervous system respond well to intrathecal methotrexate.

Children and pain

- Children's nervous systems do perceive pain
- Children do experience pain
- Children do remember pain
- Children are not more easily addicted to opioids
- There is no correct amount of pain or analgesia for a given injury

Analgesic doses

Paracetamol
- Oral dose 15 mg/kg every 4-6 hours
- Rectal dose 20 mg/kg every 6 hours
- Maximum dose 90 mg/kg/24 hours, 60 mg/kg/24 hours in neonates

Dihydrocodeine
- Ages < 4 years 500 µg/kg orally every 4-6 hours
- Ages 4-12 years 500-1000 µg/kg orally every 4-6 hours

Morphine

Immediate release preparations
- Ages < 1 year 150 µg/kg orally every 4 hours
- Ages 1-12 years 200-400 µg/kg orally every 4 hours
- Ages > 12 years 10-15 mg orally every 4 hours
- Titrate according to analgesic effect and provide laxatives

12 hourly preparations
- Ages < 1 year 500 µg/kg orally every 12 hours
- Ages 1-12 years 1 mg/kg orally every 12 hours
- Ages > 12 years 30 mg orally every 12 hours
- These are guidelines to starting doses, but many patients may start at higher doses after titration with immediate release morphine preparation every 4 hours

Diamorphine
- A third of total 24 hour dose of oral morphine
- Subcutaneous 24 hour infusion

Feeding

Being unable to nourish their child causes parents great distress and often makes them feel that they are failing as parents. Sucking and eating are part of children's development and provide comfort, pleasure, and stimulation. These aspects should be considered alongside a child's medical and practical problems with eating. Children with neurodegenerative disorders or brain tumours are particularly affected. In general, nutritional goals aimed at restoring health are secondary to comfort and enjoyment, although assisted feeding, via a nasogastric tube or gastrostomy, may be appropriate for those with slowly progressive disease.

Nausea and vomiting

These are common problems. Antiemetics can be selected according to their site of action and the presumed cause of the nausea. In resistant cases combining a number of drugs that act in different ways or adding a 5-HT_3 antagonist can be helpful. Vomiting from raised intracranial pressure should be managed with cyclizine in the first instance.

Neurological problems

Watching a child have a seizure is extremely frightening for parents, and they should always be warned if it is a possibility and advised about management. A supply of rectal diazepam at home is valuable for managing seizures. Subcutaneous midazolam can enable parents to keep at home a child with severe repeated seizures. Children with neurodegenerative disease will often already be taking maintenance antiepileptic drugs, and the dose and drugs may need adjusting as the disease progresses.

Agitation and anxiety may reflect a child's need to express his or her fears and distress. Drugs such as benzodiazepines, methotrimeprazine, and haloperidol may help to provide relief, especially in the final stages of life.

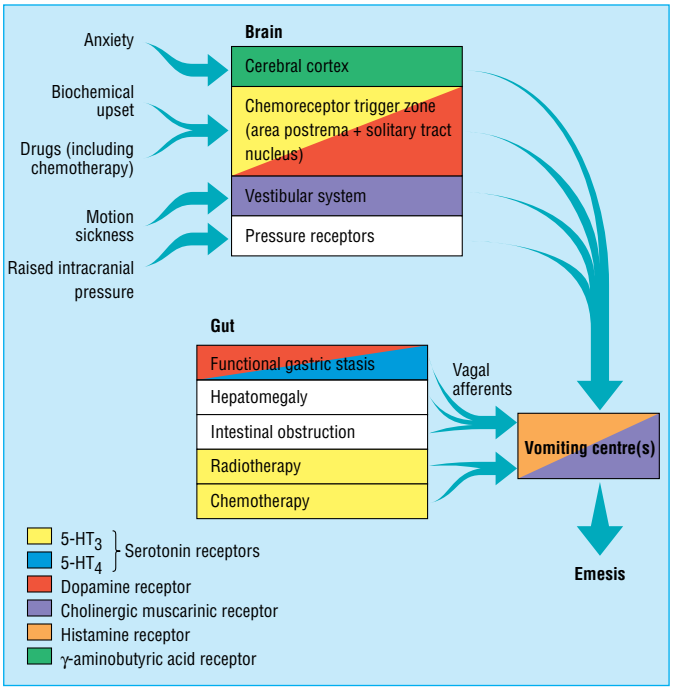
Support for the family

The needs of children with a life threatening illness and their families are summarised in the report by ACT and the Royal College of Paediatrics and Child Health. Families need support from the time of diagnosis and throughout treatment as well as when the disease is far advanced. Professionals must be flexible in their efforts to help. Each family and individual within a family is unique, with different strengths and coping skills. The needs of sibling and grandparents should be included.

The family of a child with an inherited condition have additional difficulties. They may have feelings of guilt and blame, and they will need genetic counselling and information about prenatal diagnosis in the future. When an illness does not present until some years after birth several children in the same family may be affected.

Families who maintain open communication cope most effectively, but this is not everyone's pattern. Children almost always know more than their parents think, and parents should be encouraged to be as honest as they can. Play material, books, and other resources can be supplied to help with communication, and parents can be helped to recognise their children's non-verbal cues.

Sick children need the opportunity to maintain their interests and to have short term goals for as long as possible. Education is an essential part of this, as it represents their normal pattern and continues relationships with their peers. Providing information and support to teachers facilitates this.



The emetic process—pathways of emesis and the neurotransmitters involved

Uses of antiemetic drugs

Cause of vomiting	Treatment
Raised intracranial pressure and sensory stimuli via cerebral cortex	Cyclizine Ondansetron Dexamethasone (only in resistant cases)
Drugs and biochemical upset	Phenothiazines Metoclopramide Domperidone Ondansetron Haloperidol
Gastrointestinal tract stimuli	Metoclopramide Domperidone Ondansetron

Support that every child and family should expect

- To receive a flexible service according to a care plan based on individual assessment of needs, with reviews at appropriate intervals
- To have a named key worker to coordinate their care and provide access to appropriate professionals
- To be included in the caseload of a paediatrician in their home area and have access to local clinicians, nurses, and therapists skilled in children's palliative care and knowledgeable about services provided by agencies outside the NHS
- To be in the care of an identified lead consultant paediatrician expert in individual child's condition
- To be supported in day to day management of child's physical and emotional symptoms and to have access to 24 hour care in the terminal stage
- To receive help in meeting the needs of parents and siblings, both during child's illness and during death and bereavement
- To be offered regular respite, including nursing care and symptom management, ranging from parts of a day to longer periods
- To be provided with drugs, oxygen, specialised feeds, and all disposable items such as feeding tubes, suction catheters, and stoma products through a single source
- To be provided with adaptations to housing and specialist equipment for use at home and school in an efficient and timely manner without recourse to several agencies
- To be helped in procuring benefits, grants, and other financial assistance

Bereavement

Grief after the death of a child is described as the most painful and enduring. It is also associated with a higher risk of pathological grief. Parents suffer multiple losses. Siblings suffer too and may have difficulty adjusting; they often feel isolated and neglected, as their parents can spare little energy or emotion for them.

Helping the bereaved family involves

- Support and assessment through the tasks of normal mourning—Most families do not need specialist counselling but benefit from general support and reassurance, supplied if possible by those who have known the family through the illness
- Information—Such as support groups and the Child Death Helpline (freephone 0800 282986); many parents value the opportunity of talking with others who have also experienced the death of a child
- Referral for specialist bereavement counselling if needed
- Gradual withdrawal of contact.

Ann Goldman is CLIC consultant in palliative care at Great Ormond Street Hospital for Children, London.

The ABC of palliative care is edited by Marie Fallon, Marie Curie senior lecturer in palliative medicine, Beatson Oncology Centre, Western Infirmary, Glasgow, and Bill O'Neill , science and research adviser, British Medical Association, BMA House, London. It will be published as a book in June 1998.

Communicating with children about death

Factors to consider

- Child's level of understanding
  - Of illness
  - Of death
  - Of own situation
- Child's experience
- Family's communication pattern

Methods of communication

- Verbal
- Play
- Drama
- Art
- School work
- Stories

The loss of a child

- Multiple losses for parents
  - The child who has died
  - Their dreams and hopes
  - Their own immortality
  - Their role as parents
- Stress on marriage
- Change in family structure
- Grief of siblings and grandparents

Further reading

- ACT, Royal College of Paediatrics and Child *Health guide to the development of children's palliative care services*. 1997. Available from ACT, 65 St Michael's Hill, Bristol BS2 8DZ (Tel 0117 922 1556, Fax 0117 930 4707.)
- Royal College of Paediatrics and Child Health. *Prevention and control of pain in children: a manual for health care professionals*. London, BMJ Publishing, 1997. (Tel 0171 383 6185, Fax 0171 383 6662.)

Lesson of the week

Early diagnosis of pyoderma gangrenosum is important to prevent disfigurement

A J Harris, P Regan, S Burge

Pyoderma gangrenosum should be suspected in patients with non-healing ulcers

Department of Dermatology, Stoke Mandeville NHS Trust, Aylesbury, Buckinghamshire HP21 8AL

A J Harris, *dermatology registrar*

S Burge, *consultant dermatologist*

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Pyoderma gangrenosum is a destructive inflammatory disease that responds well to immunosuppression, though surgery may exacerbate the lesions. Diagnosis is based on clinical findings. We report the case of a 24 year old man with non-healing ulcers to highlight the importance of considering pyoderma gangrenosum in patients with this symptom.

Case report

A previously fit 24 year old white man was referred to the department of dermatology with a 12 month history of a painful, enlarging ulcer on the right side of his jaw. A year earlier his general practitioner had excised a cyst from his right ear lobe, but the wound had dehisced and become ulcerated. Four attempts were made to close the wound surgically over the next year. On the first and second attempts the wound was debrided and closed immediately. Because the defect was too large to close on the third and fourth attempts a skin graft was used. On each occasion the wound failed to heal and the ulcer grew. Skin donor sites on the thighs healed normally. *x* Ray films of the mandible were normal. A decaying tooth was extracted from the patient's left lower jaw and the tooth socket also failed to heal.

Cultures of debrided tissue were negative for bacteria, mycobacteria, and fungi. Cultures from the tooth socket were also sterile. Histological examination identified a non-specific mixed inflammatory cell infiltrate with areas of necrosis. The surgeons suspected dermatitis artefacta, and referred the patient to a dermatologist.

When the patient was examined by the dermatologist the ulcer extended over the angle of the jaw and affected the right ear (fig 1). It had a haemorrhagic base with indurated, violaceous, undermined borders. Pus exuded from the ulcerated cartilage of the right ear. The ear lobe was partially destroyed. A small ulcer on the nape of the neck, an ulcerated left nipple, and an ulcer where the tooth had been extracted were also found. All had been precipitated by trauma.

A diagnosis of pyoderma gangrenosum was considered. The patient had a normal full blood count and erythrocyte sedimentation rate. Urea, electrolyte, and C reactive protein concentrations were normal, and liver function tests gave normal results. Tests for rheumatoid factor, antinuclear antibodies, and