ABC of palliative care

Non-malignant conditions

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Much medical practice is still concerned with control of symptoms rather than cure, and doctors spend considerable time palliating and modifying symptoms associated with incurable disease.

The role of specialists in palliative medicine is to offer what has been learnt about palliation of malignant disease to those caring for patients with progressive, incurable, non-malignant conditions and to share and exchange best practice. Many symptoms experienced by cancer and non-cancer patients are similar: cancer patients' symptoms may be more severe, but those of non-cancer patients tend to be more prolonged.

The approach to controlling pain in progressive non-malignant conditions can be adopted from the strategy for managing cancer related pain. After an accurate diagnosis of the pain, appropriate treatment can be started: the principles of the World Health Organisation's analgesic ladder (discussed in the first article of this series) apply equally to non-cancer patients. The strength of analgesia chosen depends on the severity of the pain, and the choice of adjuvant analgesic depends on the pathogenesis of the pain.

Doctors may be concerned about giving opioids to patients with chronic non-malignant pain. This is a justifiable worry if a systematic approach to selecting patients is not adopted. In the absence of malignancy, underprescribing of opioids for pain is not uncommon. When opioids are prescribed and effective patients will, reasonably, request regular or increased doses. Pseudoaddiction, the seeking of drugs to control pain rather than to feed psychological dependence, should be identified.

Advanced respiratory disease

The commonest chronic respiratory disorder requiring palliation is chronic obstructive pulmonary disease. Like many non-malignant conditions, however, the clinical course is not easy to predict, and a patient's life span with this condition can be decades. The illness may be characterised by frequent exacerbations followed by recovery to baseline status.

Management

If standard treatments—including oxygen, bronchodilators, antibiotics, and corticosteroids—do not adequately relieve symptoms opioids and benzodiazepines should be considered for breathlessness.

An immediate release formulation of morphine will generally ease resistant breathlessness, but use must be judged on an individual basis. Morphine should be started at a low dose, 2.5-5.0 mg every 4 hours, and titrated to an effective dose. For patients with carbon dioxide retention, careful monitoring is vital, and the frequency of dosing may have to be reduced. If an opioid is given a laxative must also be prescribed.

Often patients are anxious, and judicious use of a low dose benzodiazepine can be helpful. Care should be exercised with diazepam as, even at low doses, it will tend to accumulate because of its long half life. Lorazepam is shorter acting.

Discussion, explanations, and planning and non-drug measures are an integral part of management. Rehabilitation is desirable when possible and should be tailored to the individual patient.

A strong body of opinion argues that the skills and philosophies of palliative care should be extended to all care settings. This is optimally delivered by those working in their own specialty—such as neurology, cardiology, and respiratory medicine. Training in basic palliative care should form part of the undergraduate and postgraduate curricula for healthcare professionals. The success and relevance of palliative care will be judged not by the number of specialist teams but by the capacity to influence the care offered to all patients irrespective of diagnosis and place of care

Symptoms common to malignant and non-malignant conditions

Physical

- Pain
- Breathlessness
- Anorexia
- Immobility
- Constipation

Social

- Loss of employment
- Role change
- Fear for dependants

Psychological

- Depression
- Fear and anxiety
- Uncertainty
- Guilt

Existential

- Religious
- Non-religious
- Meaning of life
- Why?

Guidelines for prescribing opioids for pain control in non-malignant conditions

- Careful assessment of patient—Liaise with others as necessary; psychologist, psychiatrist, other physicians
- Careful assessment of pain
- Encourage patient to keep a pain diary
- Apply WHO guidelines for choice of analgesia
- Care needed if previous problems with drug dependency
- Opioids should not reinforce pain behaviour
- One prescriber of opioids
- Make a contract (fixed time, conditions)
- Regular review—Of patient, pain, function, analgesia
- Assess opioid responsiveness of pain
- Overall quality of life should improve
- Function should improve—Can range from improved concentration or enjoyment to substantial increase in mobility

Advanced cardiac disease

The management of cardiac disease at all stages has a substantial palliative component, and, unlike management of cancer, there are few opportunities for cure. This section focuses on palliative care in cardiac failure, as this is the final common pathway in most patients with advanced cardiac disease who do not die suddenly.

Prevalence—Cardiac failure affects 1-2% of the adult population, and the prevalence rises steeply with age (to more than 10% of those aged over 70). It is a disabling and lethal condition that also has a detrimental effect on quality of life. Up to 30% of affected patients require hospitalisation in any year (120 000 admissions annually in the United Kingdom). Mortality is higher than in many forms of cancer, with a 60% annual mortality with grade 4 heart failure and an overall five year mortality of 80% in men.

Clinical aspects—There are several important similarities to and differences from cancer. The now seldom used term cardiac cachexia is as apposite a term in 1997 as it was 40 years ago. Advances in antianginal therapy and interventional techniques have reduced the importance of pain as a dominant feature of cardiac failure.

Management

General

Patients with advanced cardiac failure will be faced with frequent admissions to hospital. Since patients much prefer home management if possible, this should be recognised. Cardiac liaison nurses similar to the highly developed Macmillan system would reduce the number of admissions by early detection of worsening clinical features and by ensuring that patients' homes met all the necessary requirements.

Current indications for hospital admission are

- Need for intravenous therapy
- Persistent paroxysmal nocturnal breathlessness and orthopnoea
- Refractory dependent oedema, despite up to 120 mg oral frusemide twice daily
- Symptomatic postural hypotension
- Fluid leakage from lower limbs
- Development of dysrhythmias.

Dietary advice is important since patients may be obese or cachectic. Frequent small meals are preferred, which should be tailored to each patient's tastes. Tumour necrosis factor and interleukins are implicated in the causation of cachexia, and fish oils may reduce their levels. Supplements of fat soluble and water soluble vitamins may also be necessary to counteract their increased urinary loss and reduced absorption. A small amount of alcohol may help as an appetite stimulant and anxiolytic.

Reducing fluid intake to 1500 ml a day and avoiding excessively salty foods (but not to the extent of making food tasteless) will help in controlling oedema. There is evidence that exercise may lessen breathlessness and improve both quality of life and psychological wellbeing. This must be tailored to patients' individual needs.

Drug treatment

The main emphasis is symptom relief: drugs being given to improve prognosis should be reviewed.

Opioids, combined with antiemetic drugs if necessary, are useful for control of nocturnal breathlessness.

Diuretics also have a key role—orally, intravenously, or in combination depending on the severity of fluid retention. However, awareness of the clinical and biochemical features of overdiuresis is necessary. This can lead to postural hypotension,

Clinical aspects of cardiac failure compared with cancer

Similarities to cancer

- Breathlessness, lethargy, cachexia
- Nausea, anorexia, abnormal taste
- Weight loss (loss of muscle mass countered by fluid retention)
- Constipation
- Poor mobility
- Insomnia, confusion, depression
- Dizziness, postural hypotension, cough
- Jaundice, susceptibility to infection
- Polypharmacy
- Abnormal liver function tests
- Fear of the future

Differences from cancer

- Pain not a major problem
- Oedema a more dominant feature with a different mechanism
- Predicting life expectancy less easy
- Less need for opioids
- Patients mistakenly perceive it as a more benign condition than malignancy
- Anaemia not usually present

Home care for advanced cardiac failure

- Assess appropriateness of the home—Such as comfortable bed or recliner chair, easy access to toilet, family support
- Establish need for oxygen therapy—Balance benefits and risks
- Monitor fluid status and appropriateness of diuretic treatment
- Consider quick release oral morphine 5 mg at night to ease breathlessness
- For night sedation consider temazepam 10-20 mg or, in elderly people, thioridazine 10 mg or haloperidol 0.5 mg
- Assess need for dietary advice, particularly to ensure adequate energy intake
- Ensure optimum treatment of heart failure provided drugs are not causing symptoms
- Regularly consider need for hospital admission

Marked muscle wasting in the arms (left) combined with oedema of the legs (right) in a patient with advanced heart failure

though other causes exist, some of which are shared by patients with cancer.

Digoxin has known symptomatic benefit in advanced heart failure. The optimal dose which avoids toxicity must be found.

Angiotensin converting enzyme inhibitors are of symptomatic benefit, and the dose should be titrated to ensure maximum benefit without adverse effects. Since many patients are volume depleted and hypotensive, small supervised test doses should be given—such as 6.25 mg captopril after 12-24 hours without diuretics. In patients unable to take angiotensin converting enzyme inhibitors other vasodilators should be considered.

Sublingual glyceryl trinitrate is helpful during episodes of breathlessness.

Vaccination—Influenza and pneumococcal vaccination are worth considering despite the advanced nature of the disease.

Counselling and psychological support

The highly developed support network for cancer patients is not available to patients with end stage cardiac disease. Counselling is certainly challenging in this situation because of the high incidence of sudden death (up to 50%) and the misconception of patients, who underestimate the seriousness of the situation. Application of many of the principles of palliative care is needed to optimise this aspect of management.

Symptomatic management of advanced heart failure

Breathlessness

- Oxygen
- Opiates—Regular, quick release oral morphine 5 mg, or intravenous diamorphine 2.5 mg if acutely distressed
- Non-drug measures—Such as fan, positioning, explanation, reassurance
- Diuretics, digoxin
- Vasodilators

Muscle wasting

- Physiotherapy
- Assess diet and energy intake

Fatigue

- Reassess drug therapy

Lightheadedness

- Check for postural hypotension
- Check for drug induced hypotension, from vasodilator or diuretic

Nausea, abnormal taste, anorexia

- Check drug treatments
- Check liver function
- Frequent small meals
- Appetite stimulants such as alcohol
- Consider metoclopramide

Oedema

- Early detection important
- Loop diuretics—Frusemide remains first choice
- Restrict fluid intake to 1.5-2 litres a day
- Mild salt restriction—No salt added at table
- Bed rest in early stages—If out of bed raise lower limbs via foot stool or recliner chair
- Aim for weight loss of 0.5-1 kg a day
- Combination diuretic treatments may be needed—Such as metolazone 2.5 mg on alternate days or bendrofluzide 5 mg/day plus frusemide
- Monitor electrolytes

Advanced neurological disease

Specialist palliative care services have developed a particular expertise in managing patients with progressive and advanced neurological conditions, of which motor neurone disease is perhaps the best studied example. Motor neurone disease (amyotrophic lateral sclerosis) is a disabling disorder of unknown aetiology for which there is no known cure. The mean survival is typically about three to four years.

Motor neurone disease can evoke the most negative attitudes in many medical staff, which are quite often conveyed to patients and their families. However, many of its symptoms can be alleviated by strict attention to detail and applying well established principles. While motor neurone disease is comparatively rare, many of its symptoms are common to other chronic neurological conditions. Patients with motor neurone disease require input from a range of disciplines. A strong commitment to teamwork is needed, and efficient channels of communication must be established and maintained.

The prevalence of symptoms in patients with motor neurone disease is similar to that in cancer patients. In a sample of patients referred to a hospice many of the cancer patients were referred specifically for symptom control, compared with only 15% of patients with motor neurone disease, all of whom had multiple symptoms.

Causes of postural hypotension in terminal cardiac disease and cancer

Cardiac related

- Diuretics
- Vasodilators

Cancer related

- Opioids
- Antidepressants
- Adrenal insufficiency due to metastasis

Common to both

- Bed rest
- Coexistent disease
- Muscle wasting and poor venous tone
- Reduced fluid intake and vomiting

The future of palliation in advanced cardiac disease

- Development of clinical specialist home support nurses to reduce need for hospital admission
- Improved understanding of mechanisms and treatment of nausea and cachexia
- Better early detection and control of oedema
- Improved recognition of need for psychological support and counselling

Specialists involved in caring for patients with motor neurone disease

- Neurologist
- Neurophysiologist
- Palliative medicine physician
- Endoscopist
- Anaesthetist
- Surgeon
- Respiratory physician
- General practitioner
- Physiotherapist
- Occupational therapist
- Speech and language therapist
- Nutritionist or dietician
- Nursing staff
- Social worker
- Patient support groups

Adapted from Smith AM, Eve A, Sykes NP. Palliative care services in Britain and Ireland 1990—an overview. *Palliat Med* 1992;6:277-91

Symptoms of patients on admission to a hospice

	Patients with motor neurone disease (n=124)	Patients with cancer (n=809)
Constipation	81 (65%)	338 (48%)
Pain	71 (57%)	558 (69%)
Cough	66 (53%)	380 (47%)
Insomnia	59 (48%)	235 (29%)
Breathlessness	58 (47%)	405 (50%)

Adapted from: O'Brien T, Kelly M, Saunders CM. Motor neurone disease: a hospice perspective. *BMJ* 1992;304: 471-3

Management of symptoms

Pain

Pain is common and troublesome in motor neurone disease, with 65-70% of patients reporting it to be a major symptom. Pain may occur at single or multiple sites and is often described as aching, cramping, burning, and shock-like. Some of the pain is caused by the stiffness associated with prolonged immobility and will be helped by physiotherapy and passive exercises. Additional relief can be achieved with a non-steroidal anti-inflammatory drug.

Many of these pains will require a strong opioid, and oral morphine is the drug of choice. The doses required are generally quite low, and patients may be maintained on stable doses of morphine for long periods. Although the value of opioids in motor neurone disease is generally accepted, their use is sometimes inappropriately reserved for the "terminal" phase only.

Swallowing and nutritional difficulties

Dysphagia is a common and distressing problem, and patients are often acutely embarrassed by the dribbling and coughing associated with trying to eat, while their carers are often frustrated by how long it takes to complete a meal. Salivary dribbling will usually respond to an anticholinergic agent. If drugs are unsuccessful, radiotherapy should be considered in order to dry up salivary secretions.

The swallowing reflex is often so impaired that patients cannot maintain an adequate fluid and energy intake. They will probably have experimented with a range of foods of varying consistencies, often supplemented by high energy drinks. Despite this they feel hungry and thirsty, and at least part of their continued clinical deterioration is related to their poor nutritional status.

Nasogastric tube feeding is unpleasant and cumbersome and is generally not well tolerated. Early consideration should be given to nutritional support with percutaneous endoscopic gastrostomy tube feeding. Improved nutritional status will result in a greater sense of wellbeing and should enhance patients' quality of life. Each patient will require careful explanation of the procedure and will need ongoing information and support. Complications are rare, with a mortality of 0.3-1.0% and morbidity of 3.0-5.9%.

Respiratory complications

These are a major source of morbidity and mortality in patients with motor neurone disease. Most deaths are associated with catastrophic respiratory muscle weakness. Symptoms such as cough and breathlessness are very common and may be evident at first diagnosis. Routine assessment of pulmonary function, when interpreted in conjunction with symptoms, can provide a useful indicator of disease progression and prognosis.

Management involves a multidisciplinary approach with careful assessment, explanation, and reassurance. Correct positioning enables patients to derive maximum benefit from weakened muscle groups. Oral morphine or low dose diazepam often helps to reduce the subjective sensation of breathlessness. Respiratory tract infections are sometimes difficult to diagnose clinically because of impaired inspiratory effort: symptomatic infections should be treated with appropriate antibiotics.

Ventilatory support is often controversial, yet many patients and families cope admirably with home ventilation and derive great symptomatic benefit and maintain an acceptable quality of life. Ventilatory support should be undertaken only after a comprehensive multidisciplinary assessment. Patients must be fully aware of all that is involved and be assured of adequate support.

Management of symptoms of motor neurone disease

Cough—Methadone linctus 2 mg/5 ml twice daily

Constipation—Combined purgative and softening laxative

Insomnia—Temazepam 10-20 mg, haloperidol 0.5 mg, thioridazine 10-75 mg

Difficulty swallowing oral secretions—Hyoscine butylbromide or hyoscine hydrobromide patch

Poor mobility—Physiotherapy

Response of patients with motor neurone disease to opioids

	Good	Fair	No response
Breathlessness (n = 59)	48 (81%)	2 (3%)	3 (5%)
Pain (n = 49)	36 (74%)	9 (18%)	0

Adapted from: O'Brien T, Kelly M, Saunders CM. Motor neurone disease: a hospice perspective. *BMJ* 1992;304: 471-3

Percutaneous endoscopic gastrostomy feeding is indicated for those patients who feel hungry or thirsty or who are unable to consume sufficient calories to meet their metabolic needs

Complications of percutaneous endoscopic gastrostomy

- Wound infection
- Tube dislodgement
- Peritonitis
- Aspiration
- Septicaemia
- Bowel perforation
- Peristomal leakage
- Gastrocolic fistula

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