An integrated model of provision of palliative care to patients with cystic fibrosis

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Palliative care of patients with cystic fibrosis (CF) is often undertaken by CF teams rather than palliative care teams because of the specialist nature of the disease and the potential role of lung transplantation. We developed an integrated model of provision of palliative care whereby most care is delivered by the CF team using palliative guidelines and pathways, with additional support available from the specialist palliative care team when needed. We report our experience of the terminal care of 40 patients with CF with regard to the circumstances of death, lung transplantation status, specific symptoms and provision of palliative treatments. The transition from disease modifying treatments to palliative care was particularly complex. Patients had a high level of symptoms requiring palliation and most died in hospital. Palliative care is a crucial component of a CF service and requires the specialist skills of both the CF and palliative care teams. *Palliative Medicine* (2009); 23: 512–517

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Introduction

Although the prognosis of patients with cystic fibrosis (CF) continues to improve, many still die in early adulthood. Palliative care for these patients is often undertaken by the CF team rather than by specialist palliative care teams. Palliative care is particularly complex in CF because it is paradoxically intertwined with potential lung transplantation.1–9 When the patient is being considered for transplantation, it can be difficult to address end-of-life issues as the psychological approach of the patient, family and medical team is one of ‘fighting on’ in the hope of transplantation rather than ‘bringing life peacefully to a close’. In many diseases, such as cancer, there is a gradual transition from treatments directed against the disease with the aim of prolonging survival to palliative care focusing on quality of life, symptom relief and end-of-life issues. This is also the case in CF but patients who die whilst awaiting transplantation may have an abrupt, late change from intensive treatment to palliative care, with less time for the patient and family to address end-of-life and bereavement issues. The palliative care ethos has spread from cancer to all diseases and from hospices to all clinical settings but there is a dearth of research on the palliative care needs of patients with CF and uncertainty about the best way of providing palliative care.10,11

In our CF centre, we have developed an integrated model of palliative care whereby most care is provided by the CF team using a palliative care pathway but there is additional support from the specialist palliative care services. We report our experience of the palliative care of patients dying from CF in terms of the circumstances of death, lung transplantation status, specific symptoms and provision of palliative treatments.

Methods

A retrospective review was undertaken of all deaths from CF in patients attending our Adult CF Centre over a 10-year period. The Adult CF Centre is part of a large Respiratory Medicine Department that has a specific interest in palliative care. An integrated care pathway for the dying patient and an associated palliative care guideline are used by the nurses and doctors caring for patients who are terminally ill with various diseases such as lung cancer, end-stage respiratory failure and CF.12 This provides a structured approach in addressing symptom control and the psychological needs of the dying patient. Current medications are reviewed and adjusted, inappropriate interventions are stopped, symptoms are documented, symptom-relieving medications are prescribed and the response is monitored. The plan of care also focuses on communication, religious needs and family support. There is regular support from a specialist palliative care
nurse and physician. Some medications are routinely prescribed to be given as needed for current or common potential symptoms in patients approaching death. These include opiate analgesics for pain and breathlessness, benzodiazepines for agitation, breathlessness and anxiety, anti-emetics for nausea or vomiting and hyoscine for respiratory tract secretions.

A proforma was used to review the case notes in a structured manner so as to identify areas of particular interest: (a) details of the complications of CF, (b) sputum microbiology, (c) cause of death, (d) place of death (e.g. in hospital, at home), (e) lung transplantation status, (f) decisions regarding transition from disease-modifying therapies to palliative care, (g) any advanced directives or end-of-life plans, (h) family members present during dying, (i) specific symptoms during end-stage disease and (j) treatments used to relieve symptoms. The integrated care pathway and additional medical and nursing notes were reviewed using a checklist to identify the frequency of the most prominent symptoms.

Results

In total 212 patients (age range 16–59 years) attended the CF Centre over a 10-year period and 40 patients (21 men) died. The mean age at death was 28.6 (range 17.1–51.6) years. Five patients (13%) died post-lung transplantation of obliterative bronchiolitis or sepsis at a mean of 3 (range 1.5–4.3) years post-transplantation. Thirty-five patients (87%) died of progressive CF lung disease: 27 (77%) had *Pseudomonas aeruginosa* and 8 (23%) had *Burkholderia cepacia* complex infection. Of the 35 patients dying of progressive CF lung disease, 4 (11%) died at home and 31 (89%) died in hospital. One (3%) patient died on the Intensive Therapy Unit (ITU) on endotracheal ventilation. No patient died in a hospice although one patient had spent time in a hospice for symptom control during the advanced stages of the disease. Autopsy was performed on only one patient.

Lung transplantation status

Five (13%) patients died post-lung transplantation. Of the remaining 35 patients, 11 (31%) had been on an active lung transplantation waiting list for a mean of 16.8 (range 2–24) months (Figure 1). Some of those patients had been referred for transplant assessment earlier but had only been put on an active transplantation list when their condition had deteriorated. Two of these patients were admitted to the transplant unit within one week of death but the donor organs were unsuitable and transplantation did not proceed. Four (11%) patients had not been assessed for transplantation at the time of death as they suffered an unexpected acute crisis (e.g. massive haemoptysis, severe exacerbation) (Figure 2). A further 11 patients (31%) had full lung transplantation assessment: 9 were considered unsuitable for transplantation; 2 were suitable for transplantation but were completing tests before going onto an active lung transplantation waiting list. Nine patients (26%) had opted for active medical therapy followed by palliation rather than lung transplantation (Figure 3). These patients, and those considered unsuitable for transplantation, all had major problems such as *B. cepacia* infection, liver disease, renal disease or psychological problems, which made lung transplantation an unappealing option.

Figure 1 Death whilst awaiting transplantation. This 26-year-old woman was accepted for lung transplantation because of progressive cystic fibrosis lung disease with *Pseudomonas aeruginosa* infection. Her chest radiograph shows extensive bronchiectasis with an implanted venous access device in her left subclavian vein. Over the subsequent 23 months, she had frequent admissions to hospital for exacerbations of her lung disease, which were treated by intravenous antibiotics with gastrostomy feeding for nutritional support. She then suffered a further exacerbation with progressive respiratory failure that failed to respond to non-invasive ventilation. She was commenced on an integrated palliative care pathway and died 18 h later. Prominent symptoms in the terminal phase were breathlessness, confusion, headache and some agitation, which were treated by midazolam. Lack of donor organs meant that the hope of transplantation could not be fulfilled.
Transition to palliative care

Of the 35 patients who died without having had a lung transplantation, 16 (46%) were receiving ‘long-term planned palliative care’, 6 (17%) received full active therapy till death and 13 (37%) had an abrupt change from active therapy to palliative care in the last few days of life. The discussion of potential lung transplantation automatically raised the issues of prognosis, risk of dying, the potential role and limitations of transplantation, the role of palliative care and the patient’s wishes. The discussion and planning of long-term palliative care was a complex process typically involving the patient, the family, the CF team and the transplant team. Patients who declined or were unsuitable for lung transplantation had a planned approach, whereby they received full medical therapy for their CF followed by a gradual transition to palliative care, often many years later. As palliative care became the main focus, the aim was to maximise symptom relief and quality of life whilst accepting that progressive deterioration of lung function was inevitable. In most patients, there was a gradual, generally predictable, progression of their CF lung disease but five (14%) patients suffered a sudden, unexpected deterioration due to massive haemoptysis, varicella pneumonia, influenza A infection, a severe exacerbation of P. aeruginosa lung disease, or development of necrotising pneumonia due to B. cenocepacia infection. One patient had unsuccessful cardiopulmonary resuscitation in the context of massive haemoptysis and one patient failed to respond to full endotracheal intubation and ventilation on ITU for varicella pneumonia and P. aeruginosa infection.

Of the 35 patients who died without having had lung transplantation, 22 (63%) had totally implanted venous
access systems, 15 (43%) were receiving gastrostomy feeding, 10 (29%) received non-invasive ventilation via a nasal mask, 2 (6%) had endotracheal ventilation, 1 (3%) had renal dialysis and 1 (3%) underwent dental extractions in preparation for potential transplantation. It was possible to avoid some unpleasant invasive procedures (e.g. gastrostomy feeding) in patients who were no longer being considered for transplantation. In 13 patients (37%), there was a late switch to palliative care, at a mean of 30 h (range 2–96) before death. Typically, this occurred in the context of a sudden unexpected crisis or when an acute deterioration made transplantation no longer feasible. There was a clinical impression that the quality of palliative care and the ability to address end-of-life issues was less good for those in whom the switch to palliative care occurred late in the course of the disease. This seemed particularly to be the case where the hope of lung transplantation could not be fulfilled.

Some specific complications of CF occurred late in the course of the disease in patients receiving long-term palliative care. These included distal intestinal obstruction syndrome in three patients, pneumothorax in two patients and problems relating to totally implanted venous access systems in three patients.

**Terminal symptoms and treatments**

The main symptoms experienced by patients in the terminal stages of their disease are shown in Table 1 and the treatments used for symptom control are shown in Table 2. Breathlessness was a prominent symptom in all patients and was treated by a combination of drug therapies such as midazolam and opiates, non-drug therapies such as non-invasive ventilation, cognitive behavioural therapy and general measures such as oxygen, nursing in an upright position, use of a cool air fan, breathing control methods and reassurance.

Breathlessness was frequently also linked to anxiety and fear in a vicious circle. Many patients had difficulty expectorating sputum so that there was a particular role for palliative physiotherapy in some patients, and hyoscine was used in the last few hours of life. Chest pain was common. A symptom that was particularly difficult to relieve was general malaise often described as ‘feeling awful’. This was probably related to a combination of the effects of hypoxaemia, sepsis and terminal debility. Intravenous anti-pseudomonas antibiotics were given to all patients during the terminal illness with a mixed therapeutic and palliative intent. These were usually omitted in the final 24–48 h when the clinical course was apparent. Ten patients (29%) received non-invasive ventilation via a nasal mask for relief of the effects of hypercapnia such as headache. Most treatment was given by the CF team using palliative care guidelines but there was a particular role for the skills of the specialist palliative care team in managing some difficult symptoms such as anxiety and fear.

<p>| Table 2 | Palliative treatments used in end-stage CF disease |</p>
<table>
<thead>
<tr>
<th>Treatment</th>
<th>Patients (n = 35) (%)</th>
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</thead>
<tbody>
<tr>
<td>Antibiotics</td>
<td>35 (100%)</td>
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<tr>
<td>Opiates</td>
<td>29 (83%)</td>
</tr>
<tr>
<td>Benzodiazepines</td>
<td>20 (57%)</td>
</tr>
<tr>
<td>Hyoscine</td>
<td>4 (11%)</td>
</tr>
<tr>
<td>Cognitive behavioural therapy</td>
<td>4 (11%)</td>
</tr>
<tr>
<td>Non-invasive ventilation</td>
<td>10 (29%)</td>
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**End of life issues**

Discussion of palliative care issues initially involved concepts such as ‘keeping you as well as possible for as long as possible, and then relieving symptoms when the time comes’. Some patients then discussed specific fears such as ‘choking to death’ and some wanted to discuss advanced directives about the circumstances of their death, but many patients did not seem to want to discuss specific end-of-life issues in such detail. Seven patients (20%) had a specialist consultation with the palliative care physician or nurse during the advanced stage of their disease. The issues discussed were variable and depended on the timing of the consultation. Four (11%) patients indicated specific instructions for the terminal stage of their disease. Two (6%) patients specifically indicated that they would like to be at home when they were dying but both of these patients subsequently wanted to be in hospital because of their level of symptoms when they entered terminal stage disease. Other instructions were often quite general such as ‘… parents to be present, but not sad…’

Parents were present during the final illness in 31 (89%) patients and the patient’s spouse in 9 (25%) patients. Four (11%) patients had their young children, aged 4 to 7 years, visiting and three had siblings with CF visiting during the final illness. Many also had visits from other CF patients and the pattern of the final illness was known to be discussed by other CF patients.
Discussion

This study provides a descriptive analysis of the circumstances of death and the palliative care needs of all patients attending one CF centre. It highlights some of the specific issues in palliative care of patients with CF and it outlines the difficulties that arise in the transition from therapeutic to palliative care in the context of potential lung transplantation, the level of symptoms present in end-stage disease and treatment requirements of these patients. Rather than patients being referred to a hospice, palliative care was provided in an integrated manner in the CF centre, with most care being provided by the CF team with the support of specialist palliative care services.

Although most patients had a gradual, progressive deterioration in their CF lung disease that allowed a planned approach to either lung transplantation or palliative care, some suffered a sudden crisis due to major haemoptysis or a severe exacerbation of their lung disease, such that there was an abrupt change from being reasonably well to being terminally ill. For those patients showing a progressive fall in lung function despite increasing treatment, the discussion of potential lung transplantation provided an opportunity for considering the poor prognosis, the risk of dying, the role of transplantation, end-of-life issues and palliative care. In this study, some patients were unsuitable for lung transplantation and therefore had planned palliative care, some died after lung transplantation and some were receiving full therapeutic care whilst on an active transplant waiting list but died before donor organs became available. Lung transplantation is the final treatment option for patients with end-stage CF. However, not all patients are suitable for lung transplantation and the shortage of donor organs results in 30–40% of patients dying of their CF on an active transplant waiting list before donor organs become available.\(^5,9\) It can be difficult, and perhaps inappropriate, to address some end-of-life issues whilst on an active transplant waiting list. Although the patient and family realise that the prognosis is poor and death may occur, and they can therefore address issues such as making a will, the psychological approach is one of ‘fighting on’ in the hope of rescue transplantation rather than of ‘bringing life peacefully to a close’.\(^5\) When the patient dies on an active transplantation waiting list, the family has to cope with the concept of an ‘unfulfilled hope’, although there may also be the concept that ‘everything possible was attempted’. The problem of the transition from therapeutic care to palliation in the context of lung transplantation will continue to be a major difficulty in the palliative care of CF.\(^2,3,6,7\)

In this study, patients dying of end-stage CF had a high level of complex symptoms requiring palliation. Some symptoms, such as pain, were common but quite easy to control by use of analgesic medication. Previous studies have shown a high prevalence of chronic pain in patients with CF although this symptom may be masked by the dominance of other symptoms.\(^13\) Breathlessness was a prominent symptom in all patients. This is a complex symptom that is more than shortness of breath, as it involves a complex interplay of physical, psychological, emotional and functional factors.\(^14\) Opiate and benzodiazepine medications were often helpful in relieving breathlessness, as were general measures such as use of oxygen, use of a fan blowing cool air into the patient’s face, propping the patient up in the upright posture, reassurance and distraction whereby the patient was encouraged to focus on other things rather than on their breathing.\(^14,15\) Breathlessness was often closely linked to anxiety and fear. Cognitive behavioural therapy was useful in helping some patients cope with the vicious circle of symptoms of breathlessness, anxiety and fear. This involves recognition that it is not just the impact of a physical symptom but the patient’s perception and response to the symptom that may be important.\(^16\) There was a particular role for the CF physiotherapist in the terminal stages of the disease in using airway clearance techniques to help with sputum expectoration and breathing control exercises to relieve breathlessness.\(^17\) Some patients had specific fears, such as ‘choking to death’, which could be addressed and allayed. A particularly difficult symptom to alleviate was one of general malaise that was often described as ‘feeling awful’. This may have been due to hypoxaemia, sepsis and debility.

The high level of complex symptoms was probably the main reason why most of the patients in this study died in hospital on the CF ward. Even when patients had indicated in advance that they would like to be at home when dying, they wanted to be in hospital when they entered terminal stage disease. This has also been found in other studies of terminal care of patients with CF.\(^2\) The place of death is likely to be influenced by several factors including the level of services available in the community, the involvement of the primary care team and the attitude and expectations of the patient and their family. CF patients are already known to a multiprofessional team and have often developed confidence in the nurses, physiotherapists and medical staff. In addition to a high level of general symptoms, many patients also had significant specialist CF-related problems. It therefore seemed best to provide their terminal palliative care on the CF ward rather than in a hospice. Most of their care was provided by the CF team but specialist input from the palliative care team was helpful in controlling some symptoms such as anxiety and fear by use of cognitive behavioural therapy and in addressing some end-of-life issues.

The ‘quality of death’ is very important for the patient, the family and other patients attending the CF service. An inadequate level of palliative care in the terminal stages of
the disease is likely to have an adverse effect on the bereavement process of the family, and this may be particularly relevant when siblings with CF are visiting the patient during the final illness. Palliative care is therefore a crucial component of a CF service. The extension of the palliative care skills and ethos from hospices to all clinical settings suggests that in most cases palliative care for patients with advanced CF can be provided by the CF team supported by specialist palliative care input when necessary. Provision of palliative care needs to be flexible to fulfil the individual patient’s needs and circumstances, particularly as to whether the patient wishes to be at home or in hospital as death approaches. Achieving the active total care of CF patients and their families at a time when the patient’s disease is no longer responsive to treatment is challenging and requires the specialist skills of both the CF and palliative care teams.

References