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End of life care for patients with cystic fibrosis

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Abstract

Palliative care is an approach that improves quality of life for patients and their families facing problems associated with a life-threatening illness. Care planning is particularly important in CF, where predicting a time of death is extremely difficult. The patient and family should receive realistic information about health status and further options of care. Particularly important is the explanation that treatment does not stop during the terminal phase of the disease, instead the primary aim is to alleviate unpleasant symptoms. More invasive end of life care is becoming the norm in patients awaiting lung transplantation. Terminal care should be organised in the place chosen by the patient and their family. Ideally terminal care should not end when the patient dies, instead psychological and spiritual support should continue to bereaved families.
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Keywords: Cystic fibrosis; End of life; Palliative care

1. Background

People with cystic fibrosis (CF) experience a slow deterioration of lung function coupled with numerous disease complications which may continue for many years. This long, chronic phase means that patients undergo an intensive daily therapy regimen making it difficult to predict prognosis. Constant therapeutic demands, limited predictability of the terminal phase and difficulty in defining the point that treatment ceases to be effective, reflect differences with oncology where the decision to introduce alternative therapies is well identified [1]. Given these time sequences a patient with cancer, their family and clinicians may find it easier to accept and deal with death. The timing of death is difficult to predict in CF, therefore active and palliative treatment usually continue at the same time, this may help to explain why most patients with CF die in hospital [2,3]. Different family members are involved with providing or supporting care throughout the various life stages of patients with CF, these can be parents, siblings, partners or the children of parents with CF. The term family used throughout refers to all these people, acknowledging changes at different ages.

Although the majority of patients with CF die following overwhelming pulmonary infection, technological advances have provided the CF team with alternative treatment choices for the very sick patient. Lung transplantation is a viable option for patients with CF, although it must be acknowledged that not all patients are suitable and organ availability remains an issue [2]. The situation differs dramatically around Europe: from countries without the option of lung transplantation, to countries with long waiting lists and countries where lung transplantation can be performed in weeks or even days when urgent. When lung transplantation is chosen the CF team, in partnership with the transplant team must provide careful assessment and preparation as many patients initially believe that transplantation is a cure and underestimate the commitment necessary both before and after the operation [3].

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A number of CF Centres are re-evaluating the possibility of intensive care, especially for those patients awaiting lung transplantation. Both invasive mechanical ventilation (IMV) and non-invasive ventilation (NIV) are choices that must be considered whenever appropriate [4,5]. Even though the mortality rate for patients undergoing IMV remains high, recent research reports some improvement, particularly for transplanted patients [6–9]. Although these data are insufficient to be able to derive firm conclusions, it is clear that where suitable, these options should be proposed and discussed with the patient and family.

In summary, the characteristics of the end of life phase in CF include:

- Difficulty in establishing a terminal phase
- A reluctance in some instances to introduce palliative care
- Active treatment continued until the time of death
- Possible lung transplantation
- The possible use of invasive or non-invasive mechanical ventilation

It is clear that CF teams need to agree end of life care policies for their individual patient populations. Although many CF Centres routinely offer symptom control and palliative care concurrent with active treatment, there still remains a number of places where this does not happen. Palliative care should be given a wider definition and considered not only as an option for terminally ill patients but as being similar to all therapies (pharmacological or not) where relief from pain and other distressing symptoms is provided. Thus, palliative care can be automatically integrated with ongoing therapies at every stage of the disease. The control of symptoms does not interfere with the strategy of clinical improvement, but is an objective in and of itself. This definition makes the role of palliation in chronic disease very different from the usual cures of the last days of life. The acceptance of a wider use of palliative care (pharmacological, psychological and spiritual) can for example allow the treatment of chronic pain, nausea, anxiety and dyspnoea, which may be ignored in these patients even during the terminal stages.

Treating the symptom of pain also involves the psychological and spiritual aspects of palliation and has been defined by the World Health Organisation (WHO) [10] for both adults and children.

"Palliative care is an approach that improves the quality of life of patients and their families facing problems associated with a life-threatening illness. This is provided through the prevention and relief of suffering by means of early identification, accurate assessment and treatment of pain and other problems, physical, psychosocial and spiritual. Palliative care:

- Provides relief from pain and other distressing symptoms
- Affirms life and regards dying as a normal process
- Intends neither to hasten nor to postpone death
- Integrates the psychological and spiritual aspects of patient care
- Offers a support system to help patients live as actively as possible until death
- Offers a support system to help the family cope during the patient's illness and in their own bereavement
- Uses a team approach to address the needs of patients and their families, including bereavement counselling, if indicated
- Will enhance quality of life, and may also positively influence the course of illness
- Is applicable early in the course of illness, in conjunction with other therapies that are intended to prolong life, such as chemotherapy or radiation therapy, and includes those investigations needed to better understand and manage distressing clinical complications."

"Palliative care for children represents a special, albeit closely related field to adult palliative care. The WHO's definition of palliative care appropriate for children and their families is as follows; the principles apply to other paediatric chronic disorders [10]:

- Palliative care for children 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 makes use of 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 children's homes."

2. Care planning

Planning is essential and should occur early in the course of the illness. The care team should be open, honest, and sensitive to family and cultural issues. Care planning is a process of structured discussion and documentation woven into the regular process of care. The goal is to ensure that a patient and their family's wishes will be respected [11].

Care planning is particularly important in CF, where predicting a time of death is extremely difficult [12]. The terminal phase of the disease is usually heralded by an increased frequency and severity of exacerbations, dependence on oxygen therapy, a relentless deterioration in pulmonary function and increased pain in some patients. The patient and family should receive realistic information about health status and further options of care. Particularly important is the explanation that treatment does not stop during the terminal phase of the disease, instead the primary aim will be to alleviate unpleasant symptoms [13]. The wishes of both the patient and their family concerning treatment options and place of death should always be respected and supported whenever possible. Early management planning in the terminal phase of the disease not only allows the patient and family to prepare for the moment of death, but also provides time for important decisions to be made such as putting affairs in order and planning a funeral. End of life care should be considered in patients who do not agree to further treatment and are in the

terminal stages of the disease, or who do not fulfil the criteria for lung transplantation or who do not wish to undergo lung transplantation. It is important to remember that patients who are on the lung transplant waiting list should not be excluded from palliative care options.

3. Therapy

3.1. Palliative care

The main symptoms requiring medical management in the terminal phase of the disease are dyspnoea, nausea, pain, anxiety, and confusion. The terminal phase of pulmonary disease still requires oxygen therapy, nebulised medication and physiotherapy. Often patients display symptoms of breathlessness and can become distressed, both NIV and benzodiazepines or antidepressants can be helpful in these instances. One of the more common drugs used in terminal dyspnoea is morphine and its derivates [14]. These can be administered via a variety of routes suitable to patient needs including oral, transdermal, sublingual and intravenous. Occasionally morphine can be nebulised; however the effectiveness of drug absorption varies between patients. Retrospective studies have shown that patients can tolerate levels of opiate sufficient to manage dyspnoea without respiratory depression. Intravenous morphine of less than 5 mg per hour has been reported to control dyspnoea in the majority of patients with CF [15–17]. Opiate medication (opioids) may be combined with benzodiazepines, antidepressants, anxiolytics and antiemetics. The combination of drugs should be adjusted to the individual needs of the patient (Table 1). Palliative sedation is the monitored use of medications to relieve refractory and unendurable symptoms, by inducing varied degrees of unconsciousness. The intent of palliative sedation is control of suffering, not to hasten death (Table 2) [18].

In the final phase of the disease it is uncommon to intubate and ventilate individuals; however NIV is common for symptom relief and as an aid to chest physiotherapy. Before commencing this form of therapy counselling is important for both the patient and their family as they need to understand that NIV is not being used to prolong the process of dying, but to help with symptoms such as breathlessness and to clear secretions from the lungs. In the terminal phase of the disease psychological, spiritual and social support is mandatory.

Table 1
Treatment for dyspnoea

Morphine (short acting)
- Suggested starting dose: children 0.15–0.3 mg/kg, adolescents and adults 2.5–5 mg every 4 hours PO
- End of life period – Infusion IV or SQ dose should be 33–50% of the oral dose
- Drug may be mixed with benzodiazepines, antidepressants and hyoscine.
- Inhalations: 2.5–5 mg/dose

Hyoscine
- Suggested starting dose 10 mg q 8–12 h PO or 7.5–10 mg q 5–12 h PR and then 20–40 mg/24 h infusion IV or SQ

Note: These treatment options and doses are provided as a guide only, most hospitals will have their own protocols which should be followed.
Abbreviations: IV, intravenous; SQ, subcutaneous; PO, per os; PR, per rectum.

3.2. Lung transplantation

With the wider adoption of lung transplantation as a rescue therapy for end stage CF lung disease, end of life care for CF has changed [19]. For patients on the lung transplant list, the goal is to keep the patient as well as possible until potential donor organs can be found. Given this goal, more aggressive interventions such as NIV or IMV, intensive care, enteral feeding, and indwelling catheters, are appropriate [11,20,21].

3.2.1. Prediction of short-term mortality

A significant difficulty in end of life care for patients with CF is the inability to predict short-term survival with accuracy. Short-term prognostic ability is limited both by the relatively high degree of inter-patient variability and the natural course of the pulmonary decline. Some attempts to develop prognostic markers for mortality in CF have initially focused on pulmonary function testing [12,22–24]. These studies are motivated by the need to provide an appropriate time for lung transplant listing given the waiting time for suitable organs. As a consequence, they usually focus on prediction of mortality within a two year period and are of limited use in making plans for end of life care.

Other studies have focused on clinical signs of advancing disease and other useful clinical indicators of poor short-term survival. These include the inability to maintain metabolic compensation for chronic respiratory acidosis, presence of pulmonary hypertension, an accelerated rate of decline in pulmonary function despite aggressive therapy, the lack of response to prolonged intravenous antibiotic therapy, and weight loss that cannot be halted or reversed by supplemental feeding or parenteral nutrition [25–28]. Despite the lack of convincing evidence on prognosis, prediction of the appropriate time to offer lung transplantation to CF patients remains largely subjective. This concept is reinforced in a recent update of guidelines for the selection of candidates for lung transplantation where the authors have reported that decision making will be supported by several indicators of disease severity. These indicators include: FEV_1, increased oxygen need, hypercapnia, the need for non-invasive ventilation, functional status (e.g. six minute walk test), and pulmonary hypertension [28–30] (Tables 3 and 4).

3.2.2. Impact of lung transplantation on end-of-life care

Two single centre studies have focused specifically on the effect of lung transplantation on end of life care. In the study by Dellon et al. [31] patients on the lung transplantation waiting list were more likely to die in the intensive care unit (ICU), more likely to remain intubated on the last day of life, and more likely to have the discussion about terminal care delayed. This meant that patients were unable to participate in any decision making plans. A similar study by Ford and Flume [19] found that patients on

Table 2
Drug protocol for palliative sedation (adapted from the Hospice & Palliative Care Federation of Massachusetts [18] with permission

Drug Name/Class	Suggested starting dose	Usual maintenance dose	Drug interactions	Side effects or adverse reaction	Incremental dose for titration	Issues to consider/ Incompatibilities
Midazolam (Versed)/ benzodiazepine	Infusion IV or SQ at 0.5–1.5 mg/hr after a loading dose of 1–5 mg	30–100 mg/day	CNS depressant so use cautiously with opiates or other CNS depressants. Diltiazem and verapamil increase midazolam levels.	Hiccups, decreased respiratory rate, nausea and vomiting, variations in blood pressure and pulse rates, paradoxical behaviour or excitement	• Hourly maintenance dose should be 25–33% of the required induction dose • Bolus is equal to the hourly rate every 2 hours • Adjust the maintenance dose every 2 hours based on numbers of rescue doses needed	Drug of choice for "respite sedation" or whenever reversal of sedation is desired. Drug has a short half-life. Drug may be mixed with morphine, Demerol, atropine or scopolamine. IV drug is diluted with D5W or normal saline. Drug has minimal cardiovascular effects at sedating doses.
Lorazepam (Ativan)/ benzodiazepine	Start infusion at 0.5–1 mg/hr 1–4 mg PO or buccal q 2–6 hr	4–40 mg/day Start at 1 mg for drug naïve patient	CNS depressants, may increase digoxin levels and risk of toxicity.	Paradoxical agitation, hypotension, abdominal discomfort, nausea	Titrate dose in increments of 0.5–1 mg q 15 minutes times three SQ or IV push, titrate by 1 mg q 2 hr	For bolus dosing, dilute with equal volume of sterile water for injection, normal saline for injection, or D5W. Give slowly at no more than 2 mg/minute.
Pentobarbital (Nembutal)/ Long-acting barbiturate	60–200 mg PR q 4 hr 1–3 mg/kg IV loading dose followed by 1 mg/hr	50–150 mg	CNS depression potentiated by narcotics.	N/V, respiratory depression, rash, Stevens-Johnson syndrome, angioedema, hypotension, syncope, bradycardia laryngospasm, paradoxical excitement in the elderly	Increase in 1 mg/kg increments per hour to maintain sedation	Difficult to obtain from community pharmacies. Drug has a long half life and sedation reversal is difficult. DO NOT mix with other drugs in syringe or IV. Diluted solution is stable for 12 hours only. Parenteral solution is alkaline which can cause local tissue reactions and injection site pain with extravasation.
Phenobarbital/ Long-acting barbiturate	60–120 mg PR, PO, SQ Loading dose is 200 mg bolus 1–3 mg/kg SQ or IV bolus dose, followed by starting infusion of 0.5 mg/kg/hr	Approx. 50 mg/hr	CNS depression potentiated by narcotics, Valproic acid can increase Phenobarbital levels.	Paradoxical excitement in the elderly, hypotension, nausea and vomiting, Steven's Johnson syndrome, angioedema, rash agranulocytosis, thrombocytopenia	Increase in increments of 30 mg Increase in 1 mg/kg/hr increments to maintain sedation	Drug has long half life and reversal of sedation is difficult. Drug has no analgesic effect; minimal effect on salivation; respiratory and cardiac depressant effects are dose dependent. Don't mix parenteral drug with any acidic solution. Dilute drug with half normal saline, normal saline, D5W, lactated Ringer's or Ringer's solution.

Table 2 (continued)

Drug Name/Class	Suggested starting dose	Usual maintenance dose	Drug interactions	Side effects or adverse reaction	Incremental dose for titration	Issues to consider/ Incompatibilities
Chlorpromazine (Thorazine)/ phenothiazine	25–100 mg q 4–12 hr PR 12.5 mg q 4–12 hr or 3–5 mg/hr IV	12.5–50 mg every 4–12 hr	Anticonvulsants can lower seizure threshold. Barbiturates may decrease phenothiazine effect.	May cause hypotension and extrapyramidal reactions, neuroleptic malignant syndrome, urinary retention	Good choice for patient with mild dementia Additional 25–50 mg in 1 hour as needed	Drug has no analgesic effect; minimal respiratory effect; dose-dependent vasodilation with resultant reduction in blood pressure. For IV solutions mix drug with D5W, Ringer's injection, lactated Ringer's injection or normal saline for injection.
Haloperidol (Haldol)/ butyrophene	1.0–2.0 mg PO 0.5–1 mg SQ or IV – For continuous infusion: 1.0 mg initial dose followed by infusion of 0.5 mg to 1.0 mg/hr	5 to 15 mg per day	Increased CNS depression when used with other CNS depressants. Anticholergenics are potentiated when combined with Haldol causing increased anticholigernic effect.	May cause extrapyramidal reactions, seizures, neuroleptic malignant syndrome, urinary retention, dyaphoresis, N/V	Generally do not exceed 20 mg/day to minimize the risk of neuroleptic malignant syndrome Increase infusion rate by 0.5 mg/hr	Drug is beneficial for patients with dementia.

Note: Dose ranges are highly variable, determined by patient weight, renal and hepatic function, state of hydration, concurrent medication use and other variables. Start low and titrate the dose to the desired clinical end point. Doses should be increased by approximately 30% every hour until sedation is achieved. Once the desired sedation is achieved the dose is usually maintained at that level as long as the patient seems comfortable. Previous doses of opioids and other symptom relieving medications should be continued.
Abbreviations: IV, intravenous; SQ, subcutaneous; CNS, central nervous system; D5W, Dextrose 5% in water; PO, per os; PR, per rectum.

Table 3
Clinical risk factors for death in CF patients [28,29]

Patients with CF not listed for transplant
- Pulmonary function and exercise capacity
- Microbiology (e.g. *Pseudomonas aeruginosa*, *Staphylococcus aureus*, *Burkholderia cepacia* complex)
- Nutritional status (weight)
- Age
- Female sex
- Pancreatic insufficiency
- CF-related diabetes
- Lower socioeconomic status
- Pulmonary hypertension
- Number of acute exacerbations
- Geographic location
- CF centre care

Patients with CF listed for transplant
- Yearly rate of decline in percent predicted FEV1
- Exercise test results (distance, SaO_2)
- Pulmonary hypertension
- CF-related diabetes
- Nutritional status/intervention
- PCO_2 and PO_2

Table 4
Guideline for referral and listing for lung transplantation [30]

Guidelines for referral
- FEV_1 below 30% predicted or a rapid decline in FEV_1 (particularly in young female patients)
- Exacerbation of pulmonary disease requiring ICU admission
- Increasing frequency of exacerbations requiring antibiotic therapy
- Refractory and/or recurrent pneumothorax
- Recurrent haemoptysis not controlled by embolisation

Guidelines for transplantation
- Oxygen-dependent respiratory failure
- Hypercapnia
- Pulmonary hypertension

the lung transplantation waiting list were more likely to die in the ICU receiving assisted ventilation. Ford and Flume [19] argue that the provision of more aggressive care to those awaiting lung transplantation is appropriate and surmise that clinicians are opting for aggressive care without considering the realistic odds of survival to transplantation. There are however differing management styles around the world with many adult CF centres in both the UK and Europe successfully using NIV without the need to transfer patients to an ICU. This practice has the advantage of keeping patients involved in their treatment decisions and end of life planning right up until the last day of life.

There appears to be reluctance on the part of CF clinicians to discuss advanced care planning, even though adults with CF may wish to do so. In a national survey of 234 adults with CF, 79% reported feeling comfortable talking to their clinician about advanced directives or advanced care planning, but only 28% said that their CF clinicians had asked about these issues [11]. A more comprehensive understanding of the factors that promote or hinder effective decision-making about lung transplantation and a more detailed directive on the use of medical interventions as death approaches should help clinicians support patients and families through this complex set of decisions.

3.2.3. Management of advanced lung disease prior to transplantation

Patients with CF should be cared for by a multidisciplinary team in a specialist CF centre. Treatment options for patients with advanced lung disease should be maximised by optimising all relevant aspects of care, for example, airway clearance techniques should be adapted to the patient's needs and circumstances and aerobic exercise should be performed, whenever possible. Nebulised rhDNAse can be used even in severely ill patients and nebulised hypertonic saline may have additional benefits. In patients infected by *Pseudomonas aeruginosa* (PA), maintenance inhaled antibiotics are standard care, although treatment options include monthly or alternate monthly nebulisers often with two different antibiotics. Acute exacerbation of chronic bacterial infections should be treated promptly with at least two different antimicrobial agents. In very sick patients with CF, the optimal duration of treatment is unknown, but might well be beyond the empirical 14–21 days. Respiratory complications, such as pneumothorax and massive haemoptysis need adequate intervention and may prompt consideration for lung transplantation [20]. There are no data supporting steroid use in end stage CF. However, steroids have been found to be helpful particularly in patients with an element of small airways disease. In patients with weight loss or low BMI, nutritional supplements should be offered. CF related diabetes (CFRD) should be always excluded, if found, vigorous insulin treatment with continuous glucose monitoring should be introduced according to guidelines [32].

3.2.4. Management of respiratory failure

When end-stage CF lung disease leads to hypoxemic and/or hypercapnic respiratory failure, long-term oxygen therapy with or without assisted ventilation may become necessary. As there are no specific guidelines for the use of long-term oxygen supplementation therapy [21] and non-invasive ventilation (NIV) in patients with CF, published guidelines for other chronic obstructive lung diseases may be considered [33].

A French survey on the use of NIV in 4,416 patients with CF showed that it is used as a first-line treatment for severe hypercapnic respiratory exacerbation and for stable diurnal hypercapnia, especially when associated with sleep disturbances. Bi-level, pressure-targeted ventilation is a suggested ventilation mode. Settings are adjusted to arterial blood gas values, patient comfort and sometimes sleep studies. The surveyed centres reported a number of expected benefits from NIV, but few of those benefits have been formally proven [34]. Long-term NIV with biphasic positive airway pressure (BiPAP) or intermittent nasal positive pressure may also feature as a "bridge" to transplantation [35]. In other patients, these techniques can be used in the short term as a moderately effective therapy for dyspnoea or to facilitate sputum

expectoration during physiotherapy. However, they require a period of adaptation and it must be remembered that there is a point at which NIV fails to control hypercapnia. It could be argued that the use of NIV may complicate end-of-life care and may appear for some patients to be only marginally less "invasive" than endotracheal intubation. However, patients using NIV, particularly older children and adults, are able to communicate about the helpfulness or otherwise of this treatment thereby being involved in decisions about whether to continue or stop at any time.

The use of IMV at the end of life is increasing in CF and it appears that the consensus against the use of aggressive care at the end of life in CF may be eroding. Prior to the widespread availability of lung transplantation, placing an older patient with CF on IMV was considered to be futile because it could not reverse the underlying lung disease and was associated with high mortality rates. Neither physicians nor patients wanted the patient to be transferred to the ICU if death on the ventilator was the likely outcome. Early studies demonstrated that survival in adults requiring intubation and IMV due to acute respiratory failure did not improve [36]. Recently, in the context of improved overall survival and the availability of lung transplantation, several studies have reassessed indications for intensive care and IMV in CF. Patients with acute respiratory failure due to an acute reversible condition (haemoptysis, pneumothorax or antibiotic allergy) have been reported as having a one year survival indicating that ICU treatment, including IMV, may be both appropriate and effective for those patients [4,6,37,38]. Survival among patients with acute-on-chronic respiratory failure without any identifiable triggering event necessitating IMV is less effective, but in a study by Sood et al. [4] 55% survived to ICU discharge; 40% received lung transplants and 33% were alive one year later. Intensive care, including IMV, might also be appropriate in the selected CF lung transplant candidate as a bridge to lung transplantation. The likelihood that a patient will be transplanted in due time should however, be considered when deciding to proceed with this strategy, as waiting times will vary in different countries. Several groups have reported similar transplant outcomes in ventilated CF patients compared with non-ventilated candidates [5].

Despite these promising results, clinicians and patients should carefully consider the decision to use non-invasive or invasive ventilation, as end of life care for patients with CF may be made more difficult in the ICU setting, in that the withdrawal of ventilatory support will be the common mode of death. In a sense, these methods may introduce an unsettling sort of "choice" into the decisions surrounding terminal care. Many patients with CF have grown up with a certain image of how their last weeks and days will be spent, and the introduction of an alternative is likely to challenge the expectations of both patients and caregivers.

4. Psychological aspects

End of life care in CF is very different to models used in other disease specialties such as oncology. In CF, palliative care is introduced in parallel to active treatment as both therapeutic models help to improve and relieve symptoms. Patients and families need support and involvement in treatment decision making at this time as it can be confusing. Many families and patients may have little or no experience of death or of being with someone at the end of life and can feel frightened by the unknown, therefore constant explanation must be provided. In addition, CF teams must keep in mind that both parents or partners will have been actively involved in providing care for many years and may need encouragement to continue, both for themselves and for their loved one.

It is necessary to remember the following points:

- Good communication between the CF team, the patient and their family is paramount during all aspects of the terminal stages. The CF team must take into account the patient and their family's level of understanding, concern, and fear of the unknown when discussing any treatment changes and be prepared to answer questions and re-discuss issues throughout this time [13].
- Treatment choices must allow for continued good communication between the patient and his/her family.
- It is important to provide support to friends and family about the issue of visiting and staying with the patient.
- It is important for the CF team to understand the possibilities and limitations of the care they are able to provide [39].
- The CF team, the patient and the family must work in partnership when planning end of life care, including providing comfort and the use of analgesia and anxiolytics.
- Bereavement support for friends and family often starts before death and continues afterwards. Provision should be made for support to be offered whenever necessary [40].

Attention to all these points can assist in creating a communication pathway with the patient and their family so that their needs can be met fully. Families who have been bereaved will continue to struggle with their loss. However, if they feel that they were fully involved in the process and received ongoing and appropriate support and understanding, then the bereavement process can become easier.

5. Organisation of terminal care

Terminal care should be organised in the place chosen by the patient and their family (hospital, hospice where possible, or home). Whatever option is chosen it should not restrict cooperation between the CF team and palliative care team (doctor, nurse, physiotherapist, psychologist, social worker, member of the multi-faith chaplaincy).

Ideally terminal care should not end when the patient dies, instead psychological and spiritual support should continue to the bereaved families. There are many methods to provide this service and families should be offered one that meets their needs, these include individual or group counselling provided by the CF Centre, individual or group counselling provided by the family doctor or individual or group counselling provided by associated agencies.

6. Conclusion

Although therapeutic innovations may continue to enhance survival and quality of life, families, patients, and physicians will still have to confront the premature mortality that accompanies the essentially fatal diagnosis of CF. Even in a period of advancing survival, compassionate and skilful end of life care will remain an essential part of comprehensive care for patients and their families, requiring specialist skills and good cooperation of the CF and palliative care teams.

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Conflict of interest

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