



End-of-Life Care for People with Cystic Fibrosis

*A Discussion Paper prepared by the Cystic Fibrosis Association of
Ireland in conjunction with the Irish Hospice Foundation*

CONTENTS

1. FOREWORD

2. INTRODUCTION and CONTEXT in broaching end-of-life care for people with CF

- 2.1 Irish Demographics
- 2.2 Palliative Care in Ireland

3. INTERNATIONAL PERSPECTIVE

4. SURVEY of Irish CF teams, PWCF and input from families

- 4.1 Introduction of discussion
- 4.2 Expressed Wishes/Advanced Care Planning
- 4.3 Specialist Palliative care involvement
- 4.4 Bereavement

5. CONCLUSIONS

6. RECOMMENDATIONS

- 6.1 The Next Steps

7. RESEARCH, RESOURCES and REFERENCES

8. APPENDIX

-CONTRIBUTORS TO MEETINGS

-INITIAL QUESTIONNAIRE

1. Foreword

In 2009, the Forum on End of Life Care was launched with the aim of identifying what matters most to the public on end of life from a wide range of perspectives: social, health, economic, legislative, administrative, educational, cultural and religious (IHF 2009). The Forum aimed to engage with and encourage input from all sectors of society.

In preparation for their input to this Forum, the Cystic Fibrosis Association of Ireland (CFAI) began to consider the area of end of life care for people with cystic fibrosis (PWCF). Following the presentation of their submission, and arising out of the work of the joint HSE and IHF 'Palliative Care for All' report (Irish Hospice Foundation 2008), which examined how all levels of palliative care could apply to people with diseases other than cancer, the IHF and the CFAI agreed to pursue how end of life issues for PWCF in Ireland could be addressed. A meeting was held in the CFAI in June 2009 to further address this issue, with attendance from a cross-section of staff from multidisciplinary CF teams, CF Patient Advocates and representatives from Specialist Palliative Care teams. The meeting highlighted a wide difference of opinion in how the end of life needs of PWCF should be incorporated into the routine care of PWCF. It was agreed that it would be helpful to more formally seek views on this issue. A questionnaire was subsequently devised and circulated to CF teams to capture their feedback; the views of PWCF were sought via the CF Patient Advocate.

This targeted consultation document summarises the responses of the questionnaires returned from the CF teams and the views of PWCF, as well as setting out international perspectives in this area. This document has been prepared by the CFAI in conjunction with the IHF in order to create more awareness of the issues that are present in this area, to promote discussion within the community, and to seek consensus regarding the potential mechanisms that would enhance service responses in supporting and addressing the end of life needs of PWCF. It concludes with a summary of the *general* issues raised and a number of recommendations relative to PWCF, their families and the CF teams for how to tackle these issues identified.

The document will be circulated to CF teams and the CF Adults Representation Group. Parent's views will also be sought in regard to the needs of children with CF, and the views of Specialist Palliative Care teams will also be invited. The feedback received from these avenues of communication will shape and determine the outputs of this document. Both the CFAI and IHF have committed to provide some initial financial support to assist in the implementation of the actions arising from the final report.

2. Introduction and context in broaching end-of-life care for people with CF

From the outset, it is clear from the feedback received from PWCF, parents, and healthcare professionals, that difficulties can arise when people talk about ‘end of life’ care in cystic fibrosis. This may largely be due to the variance in the age range of PWCF at time of death as well as various factors preceding death. Consequently, the need for an individualised approach is continually emphasised.

In addition, and in contrast to other degenerative conditions, in CF care there are measures that can be taken to extend life span; namely avoidance/treatment of infection and the possibility of lung transplantation. Therefore, in an aggressive medical model, these are the measures that PWCF and medical teams have tended to focus on. As a result, ‘end-stage’ CF in itself is difficult to define (severe respiratory insufficiency is generally understood to relate to this term as, at present, respiratory and related cardio problems are the final cause of death in almost 75% of PWCF, but liver disease or failure, transplant or related, or other/unknown issues have been named as the cause of death in the other 25% of cases)¹. This has meant that it is extremely difficult to develop a standardised approach to end of life care for PWCF, and the situation as it stands is that introducing palliative care interventions for PWCF are seriously underdeveloped in Ireland. In other illnesses, it may be possible to cease all curative treatment other than pain and symptom control at end of life. The very nature of CF as an illness, with its recurrent cycles of infection–treatment–improvement means that it is often difficult to recognise what can ultimately become ‘end stage’. In CF care, the provision of oxygen, IV antibiotics and physiotherapy are conventionally continued right up until the patient’s final days. Although often unlikely to combat infection at this late stage, medications and antibiotics may continue to be administered to help the PWCF clear or lighten their sputum, which is one of the most debilitating aspects of the illness. Active and palliative care treatments may need to run concurrently and it would appear that currently, in some cases, active care aimed at combating the disease overshadows palliative care options.

In the Palliative Care for All document², palliative care has been described as ‘*an approach that improves the quality of life of patients and their families facing the problems associated with life-threatening illness, through the prevention and relief of suffering by means of early identification and impeccable assessment and treatment of pain and other problems, physical, psychosocial and spiritual*’. In the current CF pathway when physical treatments are being tailored to meet the end stage needs of PWCF and their families, it appears that in some cases the psychosocial and emotional/spiritual needs are not always acknowledged or addressed.

¹ Source, CFRI data, 2002–2009

² HSE/IHF Joint Project, 2008

2.1 Irish Demographics

The prevalence of CF in the Irish population is higher than in any other country. One in nineteen people here is a carrier of the CF gene, and there are 1130 people living with CF in Ireland at present. For the first time, this population is made up of predominantly adults (51%) as opposed to children (49%). This shift reflects the major advances that have taken place in the care and management of CF, so that it is no longer a 'childhood disease', but one with which people are successfully living well into adulthood.

At present, the projected survival age for a person with CF born in 1980 is 30 years.³ Although this is still lower than international standards (34 years in the UK, 37 years in the USA), we are seeing a growing number of PWCF live into their forties and even fifties.

While this is very encouraging, it means that care provision available must adapt to the growing needs of an adult patient group. A total of 21 Irish people died from the illness in 2008; in 2009 the final toll was 17. So far in 2010, the figure stands at 7. It is hoped that together with effective treatment of the condition, attention would also be given to the needs of PWCF for end-stage management and palliative care, and subsequent bereavement counselling for their families.

2.2 Palliative Care in Ireland

Within the palliative care arena, there has been increasing acknowledgment of the need for good end of life care for all people with life-limiting diseases (Irish Hospice Foundation 2008), and specialist palliative care services (which have traditionally responded to those dying with cancer) are beginning to evolve to respond to people with non-malignant disease (O'Leary and Tiernan 2008). This approach is in line with Irish policy since 2001 (Department of Health and Children 2001). This report outlines 3 ascending levels of palliative care as follows:

Level 1 – The Palliative Care Approach

This should be a core skill of every clinician at hospital and community level. Many patients with progressive and advanced disease will have their care needs met comprehensively and satisfactorily without referral to specialist palliative care units or personnel.

Level 2 – General Palliative Care

At an intermediate level, a proportion of families will benefit from the expertise of healthcare professionals who, although not engaged full-time in palliative care, have had some additional training and experience in palliative care. Such intermediate level expertise may be available in hospital or community settings.

Level 3 – Specialist Palliative Care

The core activity of these services is limited to the provision of palliative care. These services are involved in the care of patients with more complex and demanding care needs and consequently, require a greater degree of training, staff and other resources.

³ Source, CFRI Registry 2009

In May 2010 the Irish Hospice Foundation launched ‘The Quality Standards for End of Life Care in Hospitals. These standards set out a shared vision of end-of-life care each person should have and what each hospital should aim to provide, and how each hospital can ensure that the essential elements of a consistent quality approach to end-of-life care are in place in all parts of the hospital, whether death is unexpected or expected.

The report of the National Audit of End of Life Care in Hospitals was also published in May 2010. This report notes that the quality of care for people who die in an Irish hospital compares favourably with other hospitals in the US, the UK, and France, the main countries for which we have comparative data. At the same time, the audit identified significant weaknesses in how the hospital system responds at each stage of the patient’s journey from admission through to death. These weaknesses also contain the seeds for improvement since the substantial variation in quality of care offered to patients and relatives at the end of life – not only between hospitals, specialties and wards but within them as well – demonstrates the scope for improvement that already exists, and that better and best are possible and within reach.

The publication of both these reports is very timely and pertinent with regard to the issues highlighted in this discussion document. Both these documents will be a useful guidance tool for CF teams when considering how to approach and respond to end-of-life needs of people with CF.

3. International Perspective

The treatment of CF has changed dramatically over the past ten years, with increasing life expectancy and decreasing burden of disease. There has been a shift in perception from what was previously known as a universally fatal disease, to a chronic life-limiting illness, and the preparation for end-of-life care now requires a more considered approach (Robinson 2009). Whilst recognising this shift in the disease trajectory, it has to be acknowledged that PWCF are still aware that that they may die young whilst at the same time, retain hope that medical advances will prolong their life. This tension can hinder the introduction of conversations about end of life issues and death (E Chapman 2005).

A report of the experience of palliative care treatment by the Adult Cystic Fibrosis Centre in the Newcastle upon Tyne reviewed 41 deaths over a ten year period. The transition to palliative care was gradual for the 35 who declined or were unsuitable for transplantation. It was found that the discussion of potential transplantation provided an opportunity to discuss palliative care and end of life issues. Although 46% of the patients reviewed participated in a long term palliative care programme, 37% had an abrupt change from active to palliative therapy in the last 2 days, which reflects the sudden change associated with this disease. This study noted that an inadequate level of palliative care in the final stages of the disease could lead to patient discomfort and add to the distress and bereavement of family and other patients with CF, and concluded that there are ongoing challenges in introducing the concept of palliative care when a patient is on active lung transplantation (Bourke, Doe et al. 2009).

A survey of adults with CF in the US with regard to their advance care plans reported that 79% reported feeling comfortable talking to their clinician about this issue, however, only 28% of their CF clinicians had asked them about their wishes for the

care they would want if they became too ill to make decisions for themselves. The study recommended that efforts to improve clinician communications with CF adults about advance care planning are required to ensure that such a discussion becomes an integral component of adult CF care (Sawicki, Dill et al. 2008). Additional supports required for staff was also identified in the CF centre in Newcastle upon Tyne, and they recommended that regular psychological input at team meetings, staff supervision and advice regarding specific patients has been integral to changing the culture of death, dying and bereavement in their centre. This study also observed that the introduction of end of life issues should not be regarded as a negative view of CF or indeed a failure of the CF team. (E Chapman 2005).

Robinson reports that introducing palliative care for people with CF will assist the management of difficult treatments and support planning for complex decisions, and he concludes that it is possible to use a palliative approach to manage and control symptoms whilst retaining the optimistic strategy required for clinical improvement (Robinson 2009). The study carried out by the CF centre in Newcastle upon Tyne agreed for the need to integrate palliative care and active treatment as this will allow flexibility of treatment approaches. This study also reported the following symptoms as prominent during the end stage of the disease; breathlessness, difficulty expectorating sputum, chest pain, severe malaise. It noted that the symptom of shortness of breath can involve a complex interplay of physical, psychological, emotional and functional factors. Whilst most of the treatment was given by the CF team, referrals were made to specialist palliative care personnel for assistance with symptoms relating to fear and anxiety, as well as with difficulty sleeping. (E Chapman 2005).

4. Survey of Irish CF teams, PWCF and input from families

A questionnaire was formulated based on issues that presented at a meeting in June 2009 in CF house on this topic (see appendix). This was circulated to the eight major centres involved in CF care in ROI, and it was requested that the survey be completed following discussion at the regular multi-disciplinary team meetings. Eight questionnaires were returned, five from adult and three from paediatric centres. Representation from the CF Adults group was made through their Patient Advocate; two teleconference meetings were held in May/June 2009 to discuss the issues raised in the questionnaire and to document PWCFs own priorities for the management of care at end stage. Eight to ten PWCFs actively participated in these teleconference meetings. Anecdotal information received by staff at CF House from families is also referred to.

The key areas examined include introduction of discussions, expressed wishes, palliative care involvement and bereavement. The below are summaries of responses from the surveys.

4.1 Introduction of discussion

When people talk about 'end of life' issues, these can encompass a huge range of physical, psychological, emotional, spiritual and social needs. Indeed, as the PWCF group pointed out to their Patient Advocate, any discussion about general life decisions for PWCF can raise end of life issues, e.g., difficulties when applying for travel insurance, mortgage, pension plans etc; these issues force a PWCF to confront his/her reduced life expectancy and as such, his/her own mortality from an early

stage. And yet it would appear that there still exists amongst both professionals and families a major reticence to raise and openly talk about end of life issues. Clearly a major distinction has to be drawn between paediatric and adult CF care, but it is important for us to give credence to the needs of both children and adults in this aspect of care.

Based on the data from CFRI, the vast majority of PWCF die of respiratory failure. This terminal phase in CF is usually accompanied by increased frequency and severity of respiratory exacerbations, oxygen dependence and poor lung function. Generally at this stage the PWCF will be experiencing shortness of breath, tightness and pain in the chest area, exaggerated inhalation, fear and exhaustion. So one can imagine that when a person reaches end-stage CF and is encountering these difficulties, the situation would be quite traumatic for the PWCF and their family to deal with; clearly this is not the most appropriate time to be having the first conversation about death and dying. However, feedback from medical staff in the various hospitals has revealed that, at present, discussion around the wishes of the PWCF/their family in relation to resuscitation and other end of life issues takes place on average approximately 3 days before the date of death. Even taking into consideration the uncertainty in disease trajectory for each PWCF, the question has to be asked: why is this discussion only taking place at this late stage, when everyone is aware that CF is a life-limiting disease?

The answer emerges clearly from the data returned. None of the CF teams have a protocol/policy in place to support team members to respond to these queries. Less than half reported that individual team members feel competent and confident to engage in end of life discussions with patients. The majority of CF teams have had no specific training in this area (some exceptions noted for individual team members such as social workers/psychologists). Education of primary care teams is therefore of vital importance in the future delivery of palliative care for PWCF.

Only a minority of teams have a nominated time to raise and discuss issues around deterioration of condition/end of life. Therefore, it is not surprising that discussion around end of life issues in the vast majority of cases only takes place when there is a significant deterioration in the PWCF's condition or when transplant is the next option. The formal discussion is usually introduced by the Consultant and to a lesser extent by the CF Nurse. In fact, the CF Nurses and Physiotherapists are the team members most frequently approached on an informal basis in relation to concerns that PWCFs may have on end of life issues. When a PWCF raises a concern in this area, most commonly the CF staff member refers to another nominated team member, rather than dealing with the matter themselves, or raises the issue at the next MDT for discussion. This again highlights the need for formal training and ongoing education and support of team members so that they are equipped to deal/respond to such difficult discussions. It would appear that PWCF and their families are not being afforded a suitable time and environment to raise and discuss their end of life concerns.

Key points

- *Clarity is required with regard to timing of prompts for introduction of discussion re. end of life issues*
- *An in-house protocol and procedure should be implemented in hospitals to support team members to respond to these issues*
- *Training appropriate to skill level of staff is required so that team members feel competent when end of life issues are raised*

4.2 Expressed Wishes/Advanced Care Planning

Having become ‘managers of their own care’ throughout their lifetimes, it is often the case that the majority of PWCF will not get the opportunity to make their wishes known for how their care is to be managed at the final stage. Less than half of the teams responded that the issue of resuscitation is discussed with PWCF. In cases where it is discussed, it is without variation at end stage of illness when death is anticipated – this is at a time when the PWCF is feeling physically vulnerable, and their family are emotionally stressed. The point was made by CF care professionals at the meeting in June 2009 that raising issues around death and dying when a PWCF is healthy and functioning well could cause undue anxiety and even anger, but when the PWCF group themselves were consulted, they stated the following:

“We feel it is important that end of life discussions should happen when the PWCF:

- a) can discuss the issues rationally,*
- b) can absorb the information being presented to them,*
- c) is in reasonably good health as it is easier to think the process through and make decisions suitable for each individual; when sick this thought process is much more difficult.*

*We have a preference for discussions to take place when receiving annual assessment results as this is the time when there is the possibility of change in treatments, medications etc...a yearly discussion would also give PWCF an opportunity to change their minds if they wanted to. Speaking about end-of-life care each year would also help to lift some of the negativity attached to it”.*⁴

The PWCF Adult Group also noted that if end of life issues were to be discussed at Annual Assessment stage, it would be their preference to discuss these issues with the same member of the CF team, rather than repeating information of such a sensitive nature. At present, discussions about expressed wishes remain somewhat taboo and are not broached in any structured manner. Most frequently it is the Consultant who is involved in this discussion, followed by CF Nurse and occasionally other MDT members. If a PWCF wishes to discuss palliative care and other related issues at annual review stage, it is of vital importance that the CF teams are educated and equipped to offer this very sensitive information and care. Only one team has a formal document relating to resuscitation. Other teams reported that there might be a note put on the chart or a DNR signed by the Consultant. One team is currently working on a document specifically for PWCF, but on the whole it appears that there is a major dearth of opportunities for PWCF and their families to speak to their care team about

⁴ PWCF feedback document following teleconference

their advanced care plan, and to discuss certain treatment that might arise at a future period when they no longer have the capacity to express their wishes.

The need for Advanced Care Planning is also emphasised through feedback received from bereaved families. One member recalled how distressing it was that the family were not adequately prepared or facilitated to prepare for end of life issues; in this particular situation, making the PWCF as comfortable as possible was overshadowed by the hope of transplantation. This culminated in the family experiencing severe anxiety up to the last moments with their family member, whereas in retrospect the family felt that the focus should have been on giving the family the space and privacy to say goodbye.

With regards to discussion of end of life issues for paediatrics, some feedback has suggested that these issues be addressed with parents when the child's clinical condition merits transplant assessment. In general, an individual is not sent for transplant assessment unless they are quite ill; this may be an appropriate time to introduce the palliative care team to the family so that they can assist with more difficult symptoms that may be present, or that could arise in the future.

As CF treatment is generally maintained at an intensive level right to the last, most PWCF do not have the choice of returning home to die. Many parents have also expressed that if they could change one thing about their child's death, it would be that they were afforded the opportunity to bring their child home to die. This isn't the case nationwide, for example, in Our Lady's Children's Hospital in Crumlin over 50% of children with CF die at home with the help of a palliative care team.

Key points

- *The CF care pathway must incorporate opportunities for PWCF and their families to discuss treatment options and make decisions regarding end-of-life care in the event that their health deteriorates*
- *The opportunity or 'prompt' for this discussion should take place at a time when the PWCF is in reasonably good health*
- *Concerns that CF staff have can be addressed in training and education sessions to raise awareness on this issue*

4.3 Specialist Palliative care involvement

Responses to the survey suggest that it is *generally* not standard practice for CF teams to liaise with specialist palliative care teams. The Hospital Outreach Nurse liaises with the team in some instances, however, end-of-life care is primarily managed by the CF team themselves as opposed to referring to the hospital palliative care team; only two teams reported that it was standard practice to refer CF patients to the hospital palliative care team. Again, the time of such a referral was at the point of severe health deterioration. Some of the reasons cited for non-referral included the lack of palliative care input available or that the CF Consultant would manage this aspect of care. It would appear that in most, but not all, instances that the CF team are not fully aware of the nature of input available from the specialist palliative care team, or the expertise they can bring to the complex symptoms that can emerge for PWCF at end of life.

Six of the eight CF teams stated in their questionnaires that a CF team member with a community liaison role would assist the Specialist Palliative Care and Primary Care teams in the community in supporting end-of-life care for PWCF in their own home. Five of eight CF teams felt that some education/training in the field of caring for the CF patient at home would be helpful.

Key points

- *CF team members need more information about the benefits of introducing a palliative approach within the routine care of PWCF*
- *Specialist palliative care teams in the community could play a vital role in supporting PWCF be cared for in their own home as they near the end of life*
- *CF team members need to recognise when aggressive treatments are of no further benefit to the patient, and this needs to be communicated sensitively with the family members in a timely manner*

4.4 Bereavement

Bereavement support was one of the most pressing needs identified in CF care, particularly in the paediatric teams. As part of our remit, the CFAI is currently involved in sourcing appropriate counselling for parents who contact the Association seeking assistance and support in dealing with their grief and loss. Five out of the eight CF centres reported that families are offered a bereavement service, but there is huge variation as to what this may entail – examples include a general hospital bereavement service, social work/psychology follow up with the family, the offer of a follow-up appointment with the Consultant if they wish to accept, a home visit by the CF Nurse, religious memorial services etc. However, seven of the eight teams felt that the bereavement needs of PWCF and their families are inadequately met, and this finding would match the experience of CFAI. The bereavement needs of siblings, particularly other siblings with CF, have been highlighted as an area that is often overlooked. Suggestions as to how families may be better supported post-bereavement primarily focused on greater need for access to appropriate counselling, which could be made available from the hospital itself or externally by other support services. It is important to acknowledge that many parents/partners that CFAI have been in contact with often find it very difficult to return to the place where they lost their loved one, and find it easier to attend an outside agency. Also, for many people it can take time to recover from the shock of a death (again possibly stemming from the lack of emotional preparation in the lead-up to the death) to reach the stage where they feel comfortable to discuss their loss with others, and some people contact the Association years after their PWCF has passed away. There is a clear need for any bereavement support on offer to be open-ended rather than restrictive in timeframe.

Another issue arising in this area is support for others affected by a PWCF death. Only three of the eight CF centres responded that a PWCF death is reported to other PWCF on the ward, and only then when directly asked by patients themselves, or if staff were aware that there was a relationship between the patients. Again, staff being equipped to broach these discussions with other PWCF might afford them the opportunity to share their grief and also allow them the opportunity to talk about any concerns they have in relation to their own end-of-life care. It is also important to acknowledge issues around confidentiality and sensitivity for the PWCF who passed away, as well as his/her family.

Fewer than half of the CF centres provide staff with in-house support following the death of a PWCF. The long-term nature of CF as an illness and the fact that PWCFs often require lengthy periods of inpatient treatment mean that staff members can build up close relationships with those in their care, and their grief following a bereavement is as valid as that of any other party. Dealing with continual bereavements without adequate support is an unfair demand to expect of any staff member, and it is important that any discussion about post-death support encompasses the needs of the care providers as much as the care recipients.

Some informal practices have developed at CF centres in order to help staff deal with bereavements, but in most instances these relate to personal volition as opposed to an agreed policy. While practices such as attending the PWCF's funeral may be helpful for some staff members, these practices do not compensate for the lack of a formal policy relating to staff support post-bereavement. It was noted also that some of these practices are not always feasible options for staff in the Adult CF units in particular, given staffing constraints and the unfortunate number of deaths each year. All eight centres felt that it would be helpful to have an in-house policy relating to team members and bereavement support.

Key points

- *Greater range of bereavement support is needed for family members, flexibility regarding location of service and the complexity of bereavement support required*
- *Clarity is required regarding the procedures in communicating about the death of a person with CF so that friends, and particularly those friends who have CF, are informed in a timely and sensitive manner*
- *Practices must be put in place to ensure that the bereavement needs of staff are supported appropriately*

5. Conclusions

The findings from recent international studies in the area of end of life issues for PWCF resonate in the main with the issues that were identified from the CF teams and CF Advocates in Ireland. The challenges for both PWCF and CF teams are to balance the optimism associated with the fact that people with CF are living longer, with the possibility that an acute exacerbation can potentially mean they will die unexpectedly. Experience from other countries has demonstrated that it is possible and practicable in the CF pathway to introduce a palliative care approach whilst still exploring and introducing life-prolonging treatments. The recommended time for introducing discussions regarding end of life is during the patient annual review, however training, guidance, supervision and support is required for staff so that they have the competence and confidence to deal with issues that may emerge from such a conversation. CF is a life-limiting illness – it is essential that staff in contact with PWCF are adequately educated, trained and prepared to recognise and meet their needs, as well as the needs of their families. The role of specialist palliative care for people with CF appears to be more established in the UK and the US, and it is helpful to see that CF teams in these countries avail of the specialist palliative care teams for advice regarding complex symptom management, particularly regarding breathlessness, fear and anxiety. The requirement for bereavement support for families did not surface in the international literature, compared with the need identified in Ireland; however, the bereavement and support interventions required for CF teams are equally recognised.

6. Recommendations

For Adult PWCF and their families

- End of life **discussions** with PWCF and their CF team should take place when the person is in reasonably good health, preferably with the same CF team member, at annual assessment stage
- General **information** leaflets and guidelines could be produced on what a PWCF may need to consider on end of life issues
- Create more **awareness** of end of life/palliative care options
- Greater information on **bereavement support** for CF families and siblings with CF, anticipated grief, ideas for remembrance ceremonies etc
- Highlight different avenues, e.g., through the CFAI forum, where PWCF can seek support and advice from fellow PWCF/CF Advocates on these sensitive issues

For Children with CF – paediatric setting

- Provision of education services to children with CF (could utilise arts and media) is an important paediatric palliative care issue
- **Information** leaflets and guidelines directed towards parents of children with CF to deal with anticipatory loss or end of life issues

CF Teams

- The CF care pathway should incorporate opportunities or ‘prompts’ into its clinical agenda for PWCF, so that physical care preferences and/or end of life issues can be discussed/organised in advance if so desired. Feedback from PWCF has highlighted a preference for this to take place at annual assessment stage.
- Encourage CF team members to undergo further **education** and **training** initiatives to give them the skills, knowledge and confidence to support PWCF (both adults and children) in dealing with aspects of their care relating to end of life issues. For example, highlight and circulate existing resources and training programmes currently available through the Irish Hospice Foundation (www.hospice-foundation.ie)
- Develop a **policy framework** in CF centres to support end of life issues and advanced care planning initiatives
- Establish procedures in communicating about the death of a PWCF so that friends, and particularly those friends who have CF, are informed in a timely and sensitive manner
- Additional **bereavement supports** are required to support CF staff from both a personal and professional perspective.
- Workshop on bereavement training for CF team members – would this be welcomed?
- Create more **awareness** to CF teams on the benefits on introducing palliative care into routine care
- Improve the lines of **communication** and develop relationships between CF teams and Specialist Palliative Care teams in the community
- Would a seminar be useful to discuss issues raised in this discussion document? At the seminar, speakers from Ireland and abroad could address various aspects of palliative care for people with CF

Specialist Palliative Care Services

- To promote and develop **relationships** with CF teams to enhance service development for PWCF
- Improve **communication** with CF teams, perhaps through collaboration and regular attendance of team meetings, to be available for advice re complex clinical issues.
- Highlight to the CF community any seminars/workshops applicable for further education and training
- Encourage the involvement of these teams in education/training programmes with CF teams.

General

- Development of a repository of reference material and resources on this area, which could be hosted by CFAI
- Encourage more formal research in this area

Education and Training and Information Awareness

Some contributors made the following recommendations for how education and training of CF healthcare professionals may develop and progress:

- Education should be delivered at an undergraduate level as well as post-graduate level to all disciplines through the various colleges
- We should be moving towards the ideal that all CF physicians would be able to continue to deliver care through the PWCF journey (Level 2) and know when to refer patients onto more specialist levels for difficult symptoms (physical, psychological etc).
- Refer people to The Database of Palliative Care, End-of-Life Care and Bereavement Courses (http://www.hospice-foundation.ie/index.php?option=com_content&task=view&id=397&Itemid=264). This database was developed by the Palliative Care Education Taskforce (PCET), which is a collaborative palliative care education forum of the Irish Hospice Foundation, the Irish Association for Palliative Care and the former National Council for Specialist Palliative Care. It provides information about palliative care, end-of-life care and bereavement courses from both the Republic of Ireland and Northern Ireland, and is updated every 24 hours.

6.1 The next steps...

This document has been circulated to the Specialist CF centres and CF Adults Representation Group for feedback. The views of the Specialist Palliative Care teams will also be invited. Specifically, we welcome feedback on the recommendations listed above. These recommendations are at the preliminary stage, and are only intended to be a platform on which we can build a more structured approach to addressing the issues outlined in this document.

The feedback and interest we receive from these resources will ultimately determine what happens next, e.g., formation of a National Interest Group of members from different disciplines within CF care who would be charged with the responsibility of progressing the recommendations (once finalised) as outlined in this document, implementation of protocols and procedures to equip CF team members to better cope with addressing end of life issues, etc.

7. Research, resources and references

Bourke, S. J., S. J. Doe, et al. (2009). "An integrated model of provision of palliative care to patients with cystic fibrosis." Palliative Medicine **23**(6): 512-7.

Conway et al (2008) Cystic Fibrosis in Children and Adults, the Leeds Method of Management, Revised Edition Number 7, St. James's & Seacroft University Hospitals Publication

Department of Health and Children (2001). Report of the National Advisory Committee on Palliative Care. Dublin Department of Health and Children: 157.

E Chapman, A. L. (2005). "End of life care for adult CF patients - Facilitating a good enough death." Journal of Cystic Fibrosis **4**. Volume 4, Issue 4, 249-257
E.Chapman, A.Landy, A.Lyon, C.Haworth, D.Bilton

Irish Hospice Foundation. (2009). "Forum on End of Life." Retrieved 4.12.2009, 2009, from <http://www.endoflife.ie/>.

Irish Hospice Foundation, (2008). Palliative Care for All, Integrating palliative care into disease management frameworks. Dublin: 154.

Irish Hospice Foundation, (2010). Quality Standards for End of Life Care in Hospitals. Dublin 142.

McKeown, K., Haase, T., Pratschke, J., Twomey, S., Donovan, H., and Engling, F., 2010. Dying in Hospital in Ireland: An Assessment of the Quality of Care in the Last Week of Life, Report 5, Final Synthesis Report, Dublin: Irish Hospice Foundation. Available at <http://www.hospicefriendlyhospitals.net>

O'Leary, N. and E. Tiernan (2008). "Survey of specialist palliative care services for noncancer patients in Ireland and perceived barriers " Palliative Medicine **22**: 77-83.

Robinson, W. M. (2009). "Palliative care and end of life care in cystic fibrosis: what we know and what we need to know." Current Opinion in Pulmonary Medicine **15**: 5.

Sawicki, G. S., E. J. Dill, et al. (2008). "Advance care planning in adults with cystic fibrosis." Journal of Palliative Medicine **11**(8): 1135-41.

8. Appendix 1 - Contributors to Meetings

List of attendees at Multidisciplinary Interest group meetings

Gerry Canny, Consultant in Paediatric Respiratory Medicine, Our Lady's Children's Hospital, Crumlin

Zai Edworthy, Psychologist in CF care, Temple Street Children's Hospital

Linda Groenewald, Senior Medical Social Worker in CF Care, St. Vincent's Hospital

Ruth Hannon, Senior Dietician in CF Care, Beaumont Hospital

Caroline Heffernan, PWCF Advocate, Cystic Fibrosis Association of Ireland

Geraldine Leen, Clinical Nurse Specialist CF Care, National Children's Hospital

Eleanor Leahy, Senior Physiotherapist in CF Care, Beaumont Hospital

Marie Lynch, Programme Development Manager, Irish Hospice Foundation

Ailín O'Dea, Senior Psychologist in CF Care, Beaumont Hospital

Caroline O'Brien, Medical Social Worker in CF Care, National Children's Hospital

Máirín O'Shea, Services Co-ordinator, Cystic Fibrosis Association of Ireland

Olga Price, Clinical Nurse Specialist Palliative Care, St. Vincent's University Hospital

Clare Reilly, Physiotherapist in CF care, St. Vincent's Hospital

Cathy Short, Clinical Nurse Specialist in CF Care, Cork University Hospital

Marie Twomey, Consultant in Palliative Care, Crumlin Children's Hospital

Annette Winston, Acting Principal Social Worker, Beaumont Hospital

Appendix 2

QUESTIONNAIRE

Aim: To determine existing practices of CF teams in response to end of life issues for people with Cystic Fibrosis.

Context: At an initial meeting to discuss end of life issues for people with CF on 17th June '09 held in the CF Association, it became clear that different practices are in operation in different CF centres, and indeed team involvement regarding end of life issues can vary depending on the particular patient. The need for an individualised approach was stressed and it was acknowledged that staff-patient relationships and family dynamics can have a major impact. At the meeting it was agreed that it would help to ascertain the range of existing practices, both formal and informal, that are in place. The following questionnaire has been prepared by the CF Association and the Irish Hospice Foundation for the purpose of gathering the various practices by CF teams in this area and this will inform the preparation of a discussion paper on this topic (one of the actions agreed at the above meeting).

We would ask that each CF team discuss the questions and fill out the questionnaire together. We have left space for 'comments' at the end of each section – this can be used to outline differing views between team members should these arise. Returned questionnaires will remain confidential and the discussion paper will not identify the individual contributions. We would ask that the questionnaire be returned to Máirín O'Shea by Friday 14th August.

Please state if your team deals with care of

Adults

Paediatrics

Address of CF team (optional)

I. Introduction of discussion

1. Is there a nominated time (eg. annual review or specific stage in illness) to raise and discuss issues around deterioration of condition / end of life?

Yes No

If yes, please state

a) when _____

b) whether there is a nominated person in the CF team to introduce this discussion _____

2. Who on the CF team is most frequently approached by a PWCF in relation to end of life concerns?

3. If a PWCF raises a concern in this area with a member of the CF team, which of the following takes place

a) the staff member deals with the matter themselves

b) the CF staff member refers to another nominated team member

c) other – please state _____

4. Is there a protocol or policy to support CF team members to respond to such queries? Yes No

If yes, please attach or describe

5. Do individual CF team members feel competent and confident to have this discussion with a patient? Yes No Some

If no, are individual CF team members able to acknowledge the issue raised and refer on appropriately? _____

II. Expressed Wishes

1. Is the issue of resuscitation discussed with all PWCF?

Yes No

If yes, when? _____

2. Who is involved in this discussion?

3. Is there a formal document relating to resuscitation, or a procedure that facilitates the signing of one? Yes No

If yes, please attach or describe

Comments on this Section

III. Palliative care involvement

1. What is your expectation of the role of the Specialist Palliative Care team in supporting the person with CF?

2. Is it standard practice to refer CF Patients to the hospital palliative care team? Yes No

If no, why not? _____

If yes, at what stage is referral made? _____

3. What would assist the Specialist Palliative Care and Primary Care team in the community to support end of life care for PWCF in their own home?

- Joint case conferences
- CF team member having community liaison role
- Education/training in Care of CF patient at home
- Access to patient files
- Other

Comments on this Section

IV. Bereavement

1. Are family provided with a bereavement service from the CF centre?

Yes No

If yes, please describe _____

2. Do you think the bereavement needs of people with CF and their families are adequately met? Yes No

If no, what alternatives would you suggest _____

3. Is the death communicated to other PWCF on the ward?

Yes No

If yes, how and when _____

If no, why not _____

4. We are aware that some families of people with CF appreciate when CF team members attend funeral and removal services, although this is not always possible or practical. Do you think it would be helpful for your department to have a policy in this area? Yes No

5. Are staff provided with any in-house support following the death of a PWCF?

Yes No

Does the CF team see this as a need? _____

Comments on this Section
