Cystic Fibrosis for palliative care physicians

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Current situation

• 125 deaths in 2015 in UK
  – Median age at death: 28 years
  – Place of death usually hospital (rarely hospice)
  – If on transplant list
    • 33% die while waiting for transplant
    • More likely to die in ICU
    • Less likely to participate in end of life decisions
  – Early palliative care involvement is recommended but significant barriers exist

Benefits of palliative care input

• Bourke et al (2016) Palliative Medicine 30(7) 698–702
• Palliative care team became core members of CF MDT
• Reviewed all patients with advanced progressive disease or being considered for transplant
• Identify unmet/unidentified needs
  – Holistic symptom management
  – Bereavement support to family including young siblings and peers
• Benefit to CF team
  – CF teams rated early palliative input highly
  – Education and emotional support

Barriers to palliative care input

• Palliative teams may lack clinical expertise or confidence
• Model and location of service requires flexibility
• Difficult to identify who would benefit and when
• First contact may be too late to build trust
• Limited understanding of role of palliative care
• Parallel care planning is needed to hope for transplant with preparation for death

Perceptions of health among young adults with CF

Interviews with 31 adults with CF (age 18–40)


Health perceived as “normal”

• Compare life, activity, function and development to people without condition
• Achieving independence
  – leaving home
  – risk taking and testing boundaries
• Beginning to move into inter-dependance
  – increasingly mature relationships
  – Finding a social role and occupation
• Ongoing cognitive development
  – Evolving ability to plan or consider consequences
  – Still developing self-identity and beliefs
Challenges for palliative care when health perceived as “normal”

- Focus on getting on with life
  - Don’t see value in spending time in health care settings when well
  - Difficult to introduce concept of advance care planning or palliative care
- Risk-taking behaviour and leaving home may impact health
  - May lack experience in managing own affairs
  - Smoking, drugs, nutrition etc
- Non-Adherence to treatment

Adherence in young adults

- Interviews with 49 asthma patients age 14-20
- Reasons for non-adherence to treatment
  - Forgetfulness
  - Belief that the medication is ineffective
  - Denial that one is asthmatic
  - Difficulty using inhalers
  - Inconvenience
  - Fear of side effects
  - Embarrassment
  - Laziness
  

Health perceived as “controllable”

- Starting to experience complications of CF
- Focus on maintaining positive attitude
- Compare self to peers with CF who are less well
- Adapting to loss of spontaneity
- Decisions around disclosing condition
- Ongoing cognitive and psychological development
  - Self identity interpreted alongside illness
  - Emotional maturity in some areas but fluctuates and may be socially isolated and lack confidence

Health perceived as “controllable”

- Difficult to identify the best point to get involved
  - Focus is on treatment and getting back to normal
- May have physical symptoms but assume it is “to be expected” or related to treatment
- Denial may prohibit advance care planning or acceptance of palliative care
- Hope for transplant may cause fear of opioids/infection
- Distress around deterioration and death of peers with CF
- Need to acknowledge complex role of parents

A note on parents!

- Parents
  - Source of practical and emotional support
  - Act as advocates for the patient
  - May struggle with changing roles
  - Can become anxious or over-involved
- Young adults
  - May not feel ready to make decisions about healthcare alone
  - Need time and support to develop independence
Health perceived as distressing

- Overwhelmed by condition
- Compare self unfavourably to peers with CF
- Physical deterioration with increasing symptom burden
  - Breathlessness, cough, pain, nausea, vomiting and fatigue
- Loss of psychological resilience
  - Low mood, anxiety, isolation, emotional burden of being on transplant list
- Adoption of sick role – self-identify as “not normal”

Challenges for palliative care when health perceived as distressing

- Evidence on symptom management specifically for CF is currently limited
- Multi-organ disease has an impact
  - Diabetes, malabsorption, osteoporosis, liver impairment, fertility etc
- Psychological symptoms may exacerbate dyspnoea
- Benefit of intensive physiotherapy and medications often outweighs burden even in last days/hours

Practical solutions

- Integrated working with CF teams (and transplant team if needed)
- Agree in advance decision-making process around ventilator parameters and management of deteriorating condition
- Flexibility/creativity around location of input
  - Hospital/home/ITU
  - Ways to make hospice attractive and safe

Making hospice accessible

- Aim to meet the needs that the patient identifies
  - Enablement/goal-setting/bucket lists
  - Tools for living – finances/benefits/mindfulness etc
  - Respite?
  - Peer support (with infection risk management)
- Adapting environment
  - Options to individualise bed area
  - Flexible routines/visiting rules
  - Appropriate day space
  - Internet access!
Making space

- Our goal is to give the whole team confidence to focus on the things they always do well
  - Holistic, proactive, patient-centred care
  - Great communication skills, shared decision-making
  - Opportunities to regain control, achieve goals and do some of the "work of dying"
  - Support for family and friends pre and post bereavement
  - Confident symptom assessment and management
  - Open-minded: what makes a "good death" for this individual

CF references


Young adult references

- Buston K.M and Wood S.F. Non-Compliance Amongst Adolescents with Asthma; Listening to What They Tell us About Self-Management. Family Practice 2000, 17; 134-8
- Clark (2015) Young Adult Palliative Care: Challenges and Opportunities. American Journal of Hospice & Palliative Medicine 32(1) 101-111