

section 2.6.5: Cystic Fibrosis (CF)

General description of disease and progression

Cystic fibrosis is an inherited disease that affects breathing and digestion usually presenting in childhood. Advances in medical treatment have improved the outlook for affected children and adults, however no cure available. Most affected individuals survive into their 30s,¹ though some die in childhood and others live to age 40 or beyond. Major systems affected are the respiratory and gastro-intestinal tract, while chronic pulmonary infection with progressive lung impairment are the major clinical features.²

Potential problems

- nutritional concerns are often high and enzyme replacement is used for control of malabsorption
- intravenous antibiotics, oxygen therapy and long term physiotherapy are required
- complications of disease can be debilitating and include diabetes mellitus, liver disease and arthropathy
- pain relief for chest pain, back pain and headache is frequently required
- heart/lung transplantation may be considered – however not all patients will be suitable and waiting times can be long and uncertain

Specific considerations for end stage disease

The course of illness is particularly unpredictable³ – last stages of disease are usually punctuated with increasing frequency and severity of pulmonary exacerbations and frequent admissions to hospital, declining lung function, increasing oxygen requirements and decrease in quality of life.⁴ The major cause of death is of a respiratory nature with chronic bronchial sepsis and the presence of multi-resistant organisms.

Management strategies

- the focus of symptom management is on relief of breathlessness, sputum retention and cough
- active management of dyspnoea and chest pain in the late stages of disease is often required
- treatment options may include short term intravenous antibiotics and short term assisted ventilation (BIPAP)
- continued use of formal physiotherapy is reviewed if it causes distress or discomfort to the patient
- fluids and food are directed to prevention of dehydration rather than promotion of weight gain
- emotional support for patient and family is essential with emphasis on a familiar environment whether this be an acute hospital ward, home or hospice and staff who are known and trusted
- spiritual needs of patients including mental health and well being requires assessment
- establishing a relationship with the specialist palliative care team can provide access to support for the patient, family and staff and facilitate exploration of advance care wishes

references

¹ Woolcock Institute of Medical Research, Camperdown, NSW. see www.woolcock.org.au/

² Mitchell M, Nakielna M, Tullis E, Adair C (2000) Cystic Fibrosis- End stage Care in Canada. *Chest* 118:80-84.

³ Therapeutic Guidelines- Palliative Care Version 2, (2005). Therapeutic Guidelines Ltd, Victoria, Australia.

⁴ Etherington C (2001) Terminal Care(on line) Seacroft and St James University Hospitals, Leeds, UK.