

Physiotherapy works ✓

Cystic Fibrosis

Physiotherapy is key to lifelong management of adults and children with cystic fibrosis

What is cystic fibrosis (CF)?

CF is one of the UK's most common, life-shortening, genetically inherited disease. It affects the internal organs, especially the lungs and digestive system and is characterised by chronic lung infection and inflammation and digestive problems. It is usually diagnosed in the first few weeks of life following heel prick and sweat tests, although people born before 2007 or abroad may be diagnosed later following development of symptoms.^(1, 2)

What is the role of physiotherapy?

Physiotherapists are vital members of the multidisciplinary team providing:

- **Comprehensive management** of people with CF from infants⁽³⁾ through to adulthood^(4, 5)
- **Individual treatment** primarily focussed on airway clearance and inhalation therapies to prevent airway damage and maintaining fitness, by actively encouraging people with CF to participate in regular physical activity and exercise⁽¹⁻³⁾
- **Education and training** for patients and their families to enable them to self manage their condition through essential daily home physiotherapy. Physiotherapists adapt and optimise treatment regimes throughout the person's life in line with age, disease severity and health and socio-economic status^(2, 5, 6)
- **Assessment and advice** for secondary complications such as musculoskeletal and postural problems, bone health, and continence issues.⁽⁵⁾ ▶▶

"My health dips if I don't do my physio twice a day!"

Tom (age 15)

Size of the problem

- One of the UK's most common, life-shortening, genetically inherited diseases affecting multiple organs in the body
- Affects over **10,000** individuals in the UK
- **2 million people** (1 in 25) carry the faulty gene that causes CF
- **90 per cent** of CF deaths result from related lung disease
- Each week **5 babies** are born with CF
- Each week **2 young lives** are lost to the disease; in 2010 median age at death was **29 years.**^(3, 11)

Prevention of airway damage caused by increased resistance and obstruction

Physiotherapists use and teach patients a variety of techniques to aid the removal of excess lung secretions (mucus) which cause airway obstruction and resistance.⁽⁶⁾ Used regularly in conjunction with inhaled therapies, these techniques help to clear the lungs by reducing mucus viscosity (stickiness), and stimulating cough. A systematic review has reported that individuals benefit from the short-term effects of airway clearance when compared to no airway clearance.⁽⁷⁾

Exercise is very important in the management of CF

Regular physical activity and physiotherapy led exercise help improve physical function, cardiovascular performance and muscle strength.^(5, 8, 9) Exercise may also aid airway clearance, slow the rate of lung function decline, and contribute to an improved quality of life.^(9, 10)

Case study

Two recent UK studies, in children (n=28) with mild-moderate CF lung disease, reported that comprehensive outreach physiotherapy programmes focussed on structured exercise as well as optimising inhalation and airway clearance therapy, significantly increased aerobic fitness and reduced the need for routine intravenous (IV) antibiotic treatment and hospital admissions. These 12-month intensive programmes resulted in important improvements in clinical status, lung health and quality of life, along with substantial savings to the NHS.^(13,14) Mean reduction in IV antibiotics-related costs per child ranged from £5,500 to £7,100 compared to prior year costs.

Cost of ill health



Annual expenditure on standard healthcare (excluding transplantation) for cystic fibrosis in England is around **£100m**, equivalent to **£13,700** per patient.⁽¹¹⁾ Costs of managing CF in specialist centres range from **£5,142-£40,919** per patient (excluding high cost CF drugs).⁽¹²⁾

Conclusion

- Daily lifelong physiotherapy is effective and essential in enabling people with CF achieve the best possible quality of life; decreasing complications, reducing hospital admissions and the need for antibiotic therapy and improving exercise tolerance
- Specialist physiotherapists provide expert ongoing assessment, adapting treatment plans to reflect changes in condition to maintain optimal health and wellbeing.

Further information

CSP Enquiry Handling Unit

Tel: 0207 306 6666

Email: enquiries@csp.org.uk

Web: www.csp.org.uk



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