

6.9 CYSTIC FIBROSIS

This is an inherited disorder. One in twenty of the population carries the gene for cystic fibrosis. If two carriers of the gene have children they have a one in four chance of having a child who develops cystic fibrosis.

There are currently 420 patients with cystic fibrosis diagnosed in Northern Ireland and all attend the adult and paediatric centres in Belfast.

The underlying disorder in cystic fibrosis is one of very sticky secretions. This leads to clogging of the internal organs. Major problems occur in the digestive system with the gut blocking. The pancreas can also fail leading to diabetes and poor absorption of food.

Lung problems are a prominent feature of cystic fibrosis. Repeated heavy chest infections are the hallmark of symptomatic cystic fibrosis. These infections often require antibiotics by injection. Specialist multidisciplinary teams are required to care for patients as they have the expertise to carry out necessary investigations and treatment.

Early diagnosis (by screening all newborn babies and carrying out the correct tests at the specialist centre) means that better management can be started at an earlier age. People with cystic fibrosis are living longer. However this means that their physical and psychological needs are increasing. It is important that people/carers are supported and that, in particular, young people are supported in their move from children's services to adult services.

The repeated courses of antibiotics, which are needed to treat infections, do mean that there is emergence of resistant bacteria. Specialist treatment centres are needed with facilities for control of infection such as isolation in single rooms.

Overarching Standard 34:

Newborn Screening

All babies born in Northern Ireland should be screened for cystic fibrosis.

Rationale:

Early diagnosis and appropriate referral improves survival and reduces morbidity in cystic fibrosis.

Evidence:

UK Newborn Screening Programme (CF Programme)

<http://newbornbloodspot.screening.nhs.uk/cf>

CF Trust Standards of Care (2001)

http://www.cftrust.org.uk/aboutcf/publications/consensusdoc/C_3000Standards_of_Care.pdf

Standards for newborn blood spot screening (HSS [MD] 16/2005)

http://www.ich.ucl.ac.uk/clinical_information/clinical_guidelines/cpg_guideline_0053

Castellani et al. Consensus on the use and interpretation of cystic fibrosis mutation analysis in the clinical setting. J Cyst Fibros, Vol 7, Issue 3, Pages 179-196 (May 2008) [http://www.cysticfibrosisjournal.com/article/S1569-1993\(08\)00032-5/abstract](http://www.cysticfibrosisjournal.com/article/S1569-1993(08)00032-5/abstract)

Responsibility for delivery / implementation

HSC Board

Public Health Agency

HSC Trust

Primary Care

Regional Reference Laboratory

Quality Dimension

1. All children born in Northern Ireland should be included in the national neonatal CF screening programme.
2. Positively screened babies should be referred within seven days to a specialist CF clinician.
3. All parents should be offered the opportunity to receive appropriate information at the point of diagnosis.
4. All children and adults diagnosed with CF/or related disorders should be referred to the appropriate specialist centre.
5. Patients referred for assessment should have access to all appropriate diagnostic tests to enable an accurate diagnosis to be made.

Performance Indicator	Data Source	Anticipated Performance Level	Date to be achieved by
Percentage of babies born in Northern Ireland who have had an IRT test by day 10 of life	Reference laboratory for Northern Ireland Clinical biochemistry Laboratory Belfast Health and Social Care Trust	99.5%	March 2011
Percentage of babies with a raised IRT who have had DNA analysis by day 21 of life	Neonatal screening audit Reference laboratory for Northern Ireland Clinical biochemistry Laboratory Belfast Health and Social Care Trust	99.5%	March 2011

Overarching Standard 35:

Diagnostic testing

All people suspected of having cystic fibrosis should have appropriate diagnostic testing at a specialist centre.

Rationale:

Early diagnosis and appropriate referral improves survival and reduces morbidity in cystic fibrosis.

Evidence:

UK Newborn Screening Programme (CF Programme)

<http://newbornbloodspot.screening.nhs.uk/cf>

CF Trust Standards of Care (2001)

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Responsibility for delivery / implementation

HSC Trusts

Paediatric and Adult Cystic Fibrosis Centres

Regional Reference Laboratory

Quality Dimension

1. Positively screened babies should be referred within seven days to a specialist CF clinician
2. All children and adults suspected of having CF or related disorders should be referred to the appropriate specialist centre
3. Patients referred for assessment should have access to all appropriate diagnostic tests to enable an accurate diagnosis to be made.

Performance Indicator	Data Source	Anticipated Performance Level	Date to be achieved by
Percentage of screened positive babies who are referred to a specialist centre	Paediatric CF Centre, Belfast Health and Social Care Trust	100%	March 2011
Percentage of patients referred for a clinical diagnosis who receive this within 8 weeks of seeing a consultant	Paediatric / Adult CF Centres, Belfast Health and Social Care Trust	100%	March 2011

Overarching Standard 36:**Inpatient / Outpatient Care**

All patients with cystic fibrosis should receive care as per guidelines via specialist multidisciplinary teams.

Rationale:

Specialist centre care improves quality of life and survival of people with cystic fibrosis.

Evidence:

Kerem E, Conway S, Elborn S, Heijerman H; Standards of care for patients with cystic fibrosis: a European consensus. J Cyst Fibros. 2005 Mar;4(1):7-26

http://www.elsevier.com/framework_products/promis_misc/2005.pdf

Clinical guidelines for the physiotherapy management of cystic fibrosis (January 2002)

http://www.cftrust.org.uk/aboutcf/publications/consensusdoc/C_3400Physiotherapy.pdf

CF Trust Standards of Care (2001)

http://www.cftrust.org.uk/aboutcf/publications/consensusdoc/C_3000Standards_of_Care.pdf

Responsibility for delivery / implementation

HSC Board

Public Health Agency

HSC Trusts

Paediatric and Adult Cystic Fibrosis Centres

Quality Dimension

1. All people with CF should have access to specialist multidisciplinary care for in-patient and outpatient care.
2. Specialist CF Centres should have adequate staffing levels to provide effective multidisciplinary care as set out in the standard of care document.
3. There should be effective open communication and involvement between people /families with CF and the CF centre teams.

Performance Indicator	Data Source	Anticipated Performance Level	Date to be achieved by
Percentage of patients who have an annual recorded: FEV1 BMI Sputum or cough swab microbiology and median age of death (averaged) Survival time	UK CF Registry Annual reviews	Comparable to upper quartile of UK centre performances for each dimension	March 2011
Percentage of patients and parents reporting a high satisfaction with communication / information received from the specialist team	CF Trust Patient Advocate UK CF Registry Annual reviews	For Adults 75% 90% For Children and Young People 75% 90%	March 2011 March 2012 March 2011 March 2012

Overarching Standard 37:

Infection Control

All patients with cystic fibrosis should have their care provided in a safe environment consistent with infection control policies.

Rationale:

Effective infection control reduces the number of people with CF infected with bacteria associated with poor outcomes.

Evidence:

The Burkholderia cepacia complex – Suggestions for prevention and infection control. Second edition (September 2004)

http://www.cftrust.org.uk/aboutcf/publications/consensusdoc/C_Burkholderia_cepacia_Sep_2004.pdf

Pseudomonas aeruginosa infection in people with cystic fibrosis. Suggestions for prevention and infection control. Second edition (November 2004)

http://www.cftrust.org.uk/aboutcf/publications/consensusdoc/C_Pseudomonas_aeruginosa_Nov_04.pdf

Clinical guidelines for the physiotherapy management of cystic fibrosis (January 2002)

http://www.cftrust.org.uk/aboutcf/publications/consensusdoc/C_3400Physiotherapy.pdf

Kerem E, Conway S, Elborn S, Heijerman H; Standards of care for patients with cystic fibrosis: a European consensus. J Cyst Fibros. 2005 Mar;4(1):7-26

http://www.elsevier.com/framework_products/promis_misc/2005.pdf

Responsibility for delivery / implementation

HSC Trusts

Quality Dimension

1. All people with CF should be cared for in an environment which minimises the risk of cross infection.
2. All inpatients should be cared for in single room accommodation.
3. Outpatient clinics should be segregated according to infecting organism.

Performance Indicator	Data Source	Anticipated Performance Level	Date to be achieved by
Percentage of patients receiving microbiological surveillance of at least 4 sputum samples per year	Cystic Fibrosis Centre Belfast Health and Social Care Trust	90% 95%	March 2011 March 2012
Percentage of inpatients in single room accommodation	Cystic Fibrosis Centre Belfast Health and Social Care Trust	95%	March 2012