NATIONAL INSTITUTE FOR HEALTH AND CARE EXCELLENCE

Health and social care directorate

Quality standards and indicators

Briefing paper

Quality standard topic: Cystic fibrosis

Output: Prioritised quality improvement areas for development.

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Contents

1 Introduction

This briefing paper presents a structured overview of potential quality improvement areas for cystic fibrosis. It provides the committee with a basis for discussing and prioritising quality improvement areas for development into draft quality statements and measures for public consultation.

1.1 Structure

This briefing paper includes a brief description of the topic, a summary of each of the suggested quality improvement areas and supporting information.

If relevant, recommendations selected from the key development source below are included to help the committee in considering potential statements and measures.

1.2 Development source

The key development source referenced in this briefing paper is:

Cystic fibrosis: diagnosis and management. NICE clinical guideline in development (draft for consultation). Expected publication date is 25 October 2017.

2 Overview[1]

2.1 Focus of quality standard

This quality standard will cover the diagnosis and management of cystic fibrosis in infants, children, young people and adults.

2.2 Definition

Cystic fibrosis is a multi-system genetic disorder affecting the lungs, pancreas, liver and intestine.

It results from mutations affecting a gene that encodes for a chloride channel called the cystic fibrosis transmembrane conductance regulator (CFTR), which is essential for the regulation of salt and water movements across cell membranes (Cystic Fibrosis Trust's Standards for the clinical care of children and adults with cystic fibrosis in the UK). Absent or reduced function of CFTR results in thickened secretions in the lungs, digestive system and other organs.

[1] Information in this section is from Cystic fibrosis: diagnosis and management NICE clinical guideline in development, unless referenced as from another source.

2.3 Incidence and prevalence

The UK Cystic Fibrosis Registry Annual Data Report 2016 shows that 10,461 people in the UK have cystic fibrosis, and 247 people were newly diagnosed with cystic fibrosis in the last year.

Diagnosis is primarily made during newborn screening. The annual data report shows the median age at diagnosis is 2 months, and 1 in every 2500 babies born in the UK has cystic fibrosis.

Cystic fibrosis can have a significant impact on life expectancy and quality of life. The annual data reports shows the current median age at death is 31 years and the median predicted survival is 47 years. More than 60% of people on the UK cystic fibrosis registry are aged over 16 years.

Lung function is often reduced in cystic fibrosis, and lung infections are a cause of significant morbidity. For example, 44.2% of adults with cystic fibrosis have chronic Pseudomonas aeruginosa infection and 19.9% have chronic Staphylococcus aureus infection. Such chronic infection may need long-term use of antibiotics.

In about 85% of cases the pancreatic exocrine ducts become sufficiently blocked to cause maldigestion and intestinal malabsorption (Cystic Fibrosis Trust's Standards for the clinical care of children and adults with cystic fibrosis in the UK). Infants may fail to thrive and older children and adults may become under-nourished.

2.4 Management

Most people with cystic fibrosis are diagnosed at birth through the newborn blood spot screening programme. However, cystic fibrosis may also be diagnosed later in life. The screening programme was introduced UK wide in 2006, so there is a cohort of people who have not been screened; parents can also choose not to have their babies screened; and the programme is unable to screen for all cystic fibrosis genetic variants.

Care for adults with cystic fibrosis is mainly provided by specialist cystic fibrosis centres. Outpatient care is the basis of management. Children with cystic fibrosis may be cared for solely by a specialist cystic fibrosis centre, but there are circumstances where they may also be seen in conjunction with a team in a local hospital as part of a paediatric network (shared care clinic). In addition there are other models of care and service that people with cystic fibrosis may use, including telemedicine, clinical review and treatment at home.

NHS service specifications for cystic fibrosis state that care will be delivered using a multidisciplinary team approach. Such teams include physicians or paediatricians with sufficient time in their job plans allocated to the disease, supported by specialist

nurses, dietitians, physiotherapists, pharmacists, social workers, clinical microbiologists and clinical psychologists.

2.5 National outcome frameworks

Tables 1–3 show the outcomes, overarching indicators and improvement areas from the frameworks that the quality standard could contribute to achieving.

Table 1 NHS outcomes framework 2016–17

Domain	Overarching indicators and improvement areas
2 Enhancing quality of life for people with long-term conditions	***Overarching indicator*** 2 Health-related quality of life for people with long-term conditions** ***Improvement areas*** **Ensuring people feel supported to manage their condition** 2.1 Proportion of people feeling supported to manage their condition
3 Helping people to recover from episodes of ill health or following injury	***Overarching indicators*** 3a Emergency admissions for acute conditions that should not usually require hospital admission 3b Emergency readmissions within 30 days of discharge from hospital* ***Improvement areas*** **Preventing lower respiratory tract infections (LRTI) in children from becoming serious** 3.2 Emergency admissions for children with LRTI
4 Ensuring that people have a positive experience of care	***Overarching indicators*** 4b Patient experience of hospital care ***Improvement areas*** **Improving people's experience of outpatient care** 4.1 Patient experience of outpatient services **Improving hospitals' responsiveness to personal needs** 4.2 Responsiveness to inpatients' personal needs **Improving the experience of care for people at the end of their lives** 4.6 Bereaved carers' views on the quality of care in the last 3 months of life **Improving children and young people's experience of healthcare** *4.8 Children and young people's experience of inpatient services*
5 Treating and caring for people in a safe environment and protecting them from avoidable harm	***Overarching indicators*** *5a Deaths attributable to problems in healthcare* *5b Severe harm attributable to problems in healthcare* ***Improvement areas***

	Reducing the incidence of avoidable harm 5.2 Incidence of healthcare associated infection (HCAI) i MRSA ii C. difficile
Alignment with Adult Social Care Outcomes Framework and/or Public Health Outcomes Framework * Indicator is shared ** Indicator is complementary Indicators in italics in development	

Table 2 Public health outcomes framework for England, 2016–2019

Domain	Objectives and indicators
2 Health improvement	***Objective*** People are helped to live healthy lifestyles, make healthy choices and reduce health inequalities ***Indicators*** 2.13 Proportion of physically active and inactive adults 2.20 National screening programmes 2.23 Self-reported well-being
3 Health protection	***Objective*** The population's health is protected from major incidents and other threats, whilst reducing health inequalities ***Indicators*** 3.08 Antimicrobial resistance
4 Healthcare public health and preventing premature mortality	***Objective*** Reduced numbers of people living with preventable ill health and people dying prematurely, whilst reducing the gap between communities ***Indicators*** 4.08 Mortality rate from a range of specified communicable diseases, including influenza 4.11 Emergency readmissions within 30 days of discharge from hospital*
Alignment with Adult Social Care Outcomes Framework and/or NHS Outcomes Framework * Indicator is shared ** Indicator is complementary Indicators in italics in development	

Table 3 Adult social care outcomes framework 2016–17

Domain	Overarching and outcome measures
1 Enhancing quality of life for people with care and support needs	***Overarching measure*** 1A Social care-related quality of life** ***Outcome measures***

	People manage their own support as much as they wish, so they are in control of what, how and when support is delivered to match their needs 1B Proportion of people who use services who have control over their daily life **Carers can balance their caring roles and maintain their desired quality of life** 1D Carer-reported quality of life** **People are able to find employment when they want, maintain a family and social life and contribute to community life, and avoid loneliness or isolation**
Alignment with NHS Outcomes Framework and/or Public Health Outcomes Framework ** Indicator is complementary	

3 Summary of suggestions

3.1 Responses

In total 20 stakeholders (including specialist committee members) responded to the 2-week engagement exercise 17/08/17–01/09/17.

Stakeholders were asked to suggest up to 5 areas for quality improvement. Specialist committee members were also invited to provide suggestions. The responses have been merged and summarised in table 4 for further consideration by the Committee.

Full details of all the suggestions provided are given in appendix 2 for information.

Table 4 Summary of suggested quality improvement areas

Suggested area for improvement	Stakeholders
Diagnosis • Sweat testing and gene testing	CFT, NHSE, PHE / UK NSC
Monitoring, assessment and management • Annual and routine reviews • Nutritional assessment and interventions • Exercise • Psychological assessment • Liver disease • Cystic fibrosis related diabetes	Abbott, ACPCF, BTS, CFDG UK, NHSE, SCM1, SCM2, SCM 3, SCM 4, SCM5
Pulmonary management • Airway clearance (access to adjuncts, nebulisers, airway clearance devices) • Treatment of infection (antibiotic therapy) • Inhaled therapies	ACPCF, SCM3, SCM5
Preventing cross-infection	ACPCF, CFT, NHSE, PH, SCM2, SCM3, SCM4
Service delivery • Access to care • Transition to adult services	ACPCF , BTS, NHSE, RCN, SCM1, SCM2, SCM3, SCM4, SCM5, VP
Additional / developmental areas • Lung transplant • Management of CF-SPID [Screen Positive, Inconclusive Diagnosis] group • Management of the upper airway • Monitoring adherence to treatments • Palliative care in cystic fibrosis • Physiotherapist's role relating to CFTR channel modulators • Registers and audit • Technology in airway clearance • Time to diagnostic assessment	ACPCF, BTS, CFT, NHSE, PHE / UK NSC, RCN, SCM1, SCM5
Abbott, Abbott ACPCF, Association of Chartered Physiotherapists in Cystic Fibrosis CFDG UK, Cystic Fibrosis Dietitians Group UK CFT, Cystic Fibrosis Trust BTS, British Thoracic Society NHSE, NHS England Clinical Reference Group PH, Papworth Hospital NHS Foundation Trust PHE / UK NSC, Public Health England / UK National Screening Committee RCN, Royal College of Nursing SCM x, Specialist Committee Member VP, Vertex Pharmaceuticals	
Notes 1. The following respondents have no comments at this stage • British Infection Association • Royal College of Paediatrics and Child Health	

- SCM 6, SCM7
- UK Psychosocial Professionals in Cystic Fibrosis Group

2. The Royal College of Physicians endorse the response of the British Thoracic Society

3.2 Identification of current practice evidence

Bibliographic databases were searched to identify examples of current practice in UK health and social care settings; 413 papers were identified for cystic fibrosis. In addition, 35 papers were suggested by stakeholders at topic engagement and 5 papers internally at project scoping.

Of these papers, 8 have been included in this report and are included in the current practice sections where relevant. Appendix 1 outlines the search process.

4 Suggested improvement areas

4.1 Diagnosis

4.1.1 Summary of suggestions

Sweat testing and gene testing

A stakeholder suggested the use of the sweat test for all babies that have screened positive for cystic fibrosis (as part of the newborn blood spot screening programme), adding that this does not happen in all cases. The sweat test is a key test used to confirm or exclude the diagnosis of cystic fibrosis.

Cystic fibrosis transmembrane conductance regulator (CFTR) mutation genotyping of people with cystic fibrosis was also suggested by stakeholders. Both CFTR mutations for all patients should be recorded, so they can benefit from emerging genotype-targeted therapies. One stakeholder said the results should be made available to patients.

4.1.2 Selected recommendations from development source

Table 5 below highlights recommendations that have been provisionally selected from the development source that may support potential statement development. These are presented in full after table 5 to help inform the committee's discussion.

Table 5 Specific areas for quality improvement

Suggested quality improvement area	Suggested source guidance recommendations
Sweat testing and gene testing	**Diagnosis of cystic fibrosis** Cystic fibrosis. NICE clinical guideline in development. Recommendations 1.1.2 and 1.1.3

Diagnosis of cystic fibrosis

Cystic fibrosis. NICE clinical guideline in development. Recommendation 1.1.2

Assess for cystic fibrosis and, when clinically appropriate, perform a sweat test (for children and young people) or a cystic fibrosis gene test (for adults) in people with any of the following:

- congenital intestinal atresia
- meconium ileus
- symptoms and signs that suggest distal intestinal obstruction syndrome

- faltering growth (in infants and young children)
- undernutrition
- recurrent and chronic pulmonary disease, such as:
  - recurrent lower respiratory tract infections
  - clinical or radiological lung disease (in particular bronchiectasis)
  - persistent chest X-ray changes
  - chronic wet or productive cough
- chronic sinus disease
- obstructive azoospermia (in young people and adults)
- acute or chronic pancreatitis
- malabsorption
- rectal prolapse (in children)
- pseudo-Bartter syndrome.

Cystic fibrosis. NICE clinical guideline in development. Recommendation 1.1.3

Refer people with suspected cystic fibrosis to a specialist cystic fibrosis centre if:

- they have a positive or equivocal sweat test result
- their assessment suggests they have cystic fibrosis but their test results are normal
- gene testing reveals 1 or 2 cystic fibrosis mutations

4.1.3 Current UK practice

Sweat testing and gene testing

No current practice data was identified for sweat testing. The UK Cystic Fibrosis Registry Annual Data Report 2016 shows that 98.4% of patients with complete data have genotypes recorded for both cystic fibrosis mutations (95% of cystic fibrosis patients on the register have complete data).

4.1.4 Resource impact

No significant resource impact is expected from implementing recommendations in the draft guidance.

4.2 Monitoring, assessment and management

4.2.1 Summary of suggestions

Annual and routine reviews

Regular monitoring and review were suggested as a key area for quality improvement by stakeholders. One emphasised the importance of monitoring the progression of the disease as new cystic fibrosis-related conditions emerge over time which require specialist management.

Nutritional assessment and interventions

Stakeholders proposed nutritional assessment and interventions as a key area for quality improvement, recognising that malnutrition can affect lung function, morbidity and mortality. Nutritional assessment was suggested for all people with cystic fibrosis, and some stakeholders commented that they should be seen by a specialist dietician. Pancreatic Enzyme Replacement Therapy (PERT) was suggested as a specific intervention for newborns with pancreatic insufficiency.

Exercise

Exercise was suggested by stakeholders as it can improve lung function, quality of life, and life expectancy. Comments indicated that exercise was important from diagnosis onwards. One stakeholder emphasised the importance of exercise programmes being individualised, and said there was variation in assessment and provision across cystic fibrosis centres.

Psychological assessment

Psychology input was suggested by one stakeholder, especially for newly diagnosed patients. They added that whilst psychology review is considered best practice for cystic fibrosis centres, there is a lack of information on the nature and types of reviews.

Liver disease

Liver disease was suggested by a stakeholder who described it as the third most common cause of mortality in adults with cystic fibrosis. They considered effective monitoring in childhood as paramount and said there is wide variation in assessment and management.

Cystic fibrosis related diabetes

Management of cystic fibrosis related diabetes was suggested by stakeholders who described it as the most common co-morbidity in cystic fibrosis. Comments noted that annual testing for cystic fibrosis related diabetes should begin from 10 years of

age, and that insulin and individualised dietary education is the recommended treatment. However, it was acknowledged that there is a lack of evidence in some areas.

4.2.2 Selected recommendations from development source

Table 6 below highlights recommendations that have been provisionally selected from the development source that may support potential statement development. These are presented in full after table 6 to help inform the committee's discussion.

Table 6 Specific areas for quality improvement

Suggested quality improvement area	Suggested source guidance recommendations
Annual and routine reviews	**Annual and routine reviews** Cystic fibrosis. NICE clinical guideline in development. Recommendations 1.5.2 and 1.5.3
Nutritional assessment and interventions	**Multidisciplinary team** Cystic fibrosis. NICE clinical guideline in development. Recommendation 1.3.12 **Annual and routine reviews** Cystic fibrosis. NICE clinical guideline in development. Recommendation 1.5.2 **Nutritional interventions and exocrine pancreatic insufficiency** Cystic fibrosis. NICE clinical guideline in development. Recommendations 1.7.1, 1.72 and 1.76
Exercise	**Annual and routine reviews** Cystic fibrosis. NICE clinical guideline in development. Recommendation 1.5.2 **Exercise** Cystic fibrosis. NICE clinical guideline in development. Recommendations 1.7.31 to 1.7.32
Psychological assessment	**Multidisciplinary team** Cystic fibrosis. NICE clinical guideline in development. Recommendation 1.3.14 **Annual and routine reviews** Cystic fibrosis. NICE clinical guideline in development. Recommendation 1.5.2

	Psychological assessment Cystic fibrosis. NICE clinical guideline in development. Recommendations 1.7.34 and 1.7.36
Liver disease	**Annual and routine reviews** Cystic fibrosis. NICE clinical guideline in development. Recommendation 1.5.2 **Liver disease** Cystic fibrosis. NICE clinical guideline in development. Recommendations 1.7.17 and 1.7.18
Cystic fibrosis related diabetes	**Annual and routine reviews** Cystic fibrosis. NICE clinical guideline in development. Recommendation 1.5.2 **Cystic-fibrosis-related diabetes** Cystic fibrosis. NICE clinical guideline in development. Recommendations 1.7.24

Note: Recommendations that follow are arranged by the sub-groups of suggested areas (left-hand column in table 6), then by sub-heading within the guideline (right-hand column in table 6). Recommendation 1.5.2 applies to all sub-groups, but for the sake of brevity it is shown only under 'Annual and routine reviews'.

Annual and routine reviews

Cystic fibrosis. NICE clinical guideline in development. Recommendation 1.5.2

Offer people with cystic fibrosis a comprehensive annual review that includes the following:

- a pulmonary assessment (see Pulmonary monitoring)
- an assessment of nutrition and intestinal absorption (see Nutritional interventions and exocrine pancreatic insufficiency 1.7.4)
- as assessment for liver disease (see Liver disease)
- testing for cystic fibrosis-related diabetes, from 10 years of age (see Cystic-fibrosis-related diabetes)
- an assessment for other potential or existing cystic fibrosis complications (see Complications of cystic fibrosis)
- assessments by a specialist physiotherapist, dietician, pharmacist and clinical psychologist (see Service delivery)

- a review of the exercise programme (see Exercise).

Cystic fibrosis. NICE clinical guideline in development. Recommendation 1.5.3

Provide regular routine reviews for people with cystic fibrosis, and do these more frequently immediately after diagnosis and in early life. For example:

- weekly in their first month of life
- every 4 weeks when they are between 1 and 12 months old
- every 6 or 8 weeks when they are between 1 and 5 years old
- every 8 or 12 weeks when they are over 5 years old
- every 3 or 6 months as adults.

Nutritional assessment and interventions: Multidisciplinary team

Cystic fibrosis. NICE clinical guideline in development. Recommendation 1.3.12

The specialist dietitian should assess and advise people with cystic fibrosis about nutrition at outpatient clinic visits, during inpatient admissions and at their annual review.

Nutritional assessment and interventions: Nutritional interventions and exocrine pancreatic insufficiency

Cystic fibrosis. NICE clinical guideline in development. Recommendation 1.7.1

The cystic fibrosis specialist dietitian should offer advice on the benefits of optimal nutrition, and at the annual assessment review the person's:

- total nutritional intake, including energy intake (calories)
- estimated nutritional needs
- pancreatic exocrine replacement therapy, if appropriate (see Exocrine pancreatic insufficiency)

Cystic fibrosis. NICE clinical guideline in development. Recommendation 1.7.2

Encourage people to increase calorie intake by increasing portion size and eating high-energy foods if there is concern about their nutrition (including weight loss and inadequate weight gain).

Cystic fibrosis. NICE clinical guideline in development. Recommendation 1.7.6

Offer oral pancreatic enzyme replacement therapy to people with exocrine pancreatic insufficiency. Adjust the dose as needed to minimise any symptoms or signs of malabsorption.

Exercise: Exercise

Cystic fibrosis. NICE clinical guideline in development. Recommendation 1.7.31

Offer people with cystic fibrosis an individualised exercise programme, taking into account their capability and preferences.

Cystic fibrosis. NICE clinical guideline in development. Recommendation 1.7.32

Regularly review exercise programmes to monitor the person's progress and ensure that the programme continues to be appropriate for their needs.

Psychological assessment: Multidisciplinary team

Cystic fibrosis. NICE clinical guideline in development. Recommendation 1.3.14

The specialist clinical psychologist should assess and advise people with cystic fibrosis at their annual review and when needed at outpatient and inpatient clinics.

Psychological assessment: Psychological assessment

Cystic fibrosis. NICE clinical guideline in development. Recommendation 1.7.34

A specialist cystic fibrosis clinical psychologist (see Multidisciplinary team) should be available to see people with cystic fibrosis at outpatient clinic visits and during inpatient admissions.

Cystic fibrosis. NICE clinical guideline in development. Recommendation 1.7.36

At the annual review, the specialist clinical psychologist should include assessments of:

- general mental health and wellbeing
- quality of life
- any factors that are making treatment adherence difficult
- psychosocial indicators
- behaviours that affect health outcomes.

Liver disease: Liver disease

Cystic fibrosis. NICE clinical guideline in development. Recommendation 1.7.17

Perform a clinical assessment and liver function blood tests at the annual review for people with cystic fibrosis.

Cystic fibrosis. NICE clinical guideline in development. Recommendation 1.7.18

If liver function blood tests are abnormal, perform a liver ultrasound scan and consider ursodeoxycholic acid treatment.

Cystic-fibrosis-related diabetes: Cystic-fibrosis-related diabetes

Cystic fibrosis. NICE clinical guideline in development. Recommendation 1.7.24

Test for cystic-fibrosis-related diabetes (as detailed in recommendation 1.7.23) in people with cystic fibrosis annually from 10 years of age.

4.2.3 Current UK practice

Annual and routine reviews

The UK Cystic Fibrosis Registry Annual Data Report 2016 shows that 95% of patients with cystic fibrosis have complete data based on their annual review (so this proportion must have had an annual review).

The Cystic Fibrosis Trust, British Thoracic Society and British Paediatric Society in conjunction with the national service commissioner previously facilitated peer review of UK cystic fibrosis specialist centres and their network clinics. Peer review reports were produced which include assessment against Cystic Fibrosis Trust standards of care, delivery against professional standards and a patient survey. For this briefing paper, a selection of the reports available for specific centres and networks have been used to present information on variation in practice. The reports chosen are:

- Birmingham Children's Hospital and Paediatric Shared Care Network (2014)
- Glenfield Hospital Adult Cystic Fibrosis Centre (2015)
- Great North Children's Hospital, Newcastle Paediatric Cystic Fibrosis Peer Review (2015 report)
- Royal Brompton Hospital Adult Centre (2013)
- University Hospital of South Manchester Adult Centre (2014)

There are limitations in using this selection; the reports were produced at different points in time, may not be representative of all the centres, and are unlikely to

capture all the variation that exists. However, they give some insight into variation in practice.

The Peer Review reports show that there is some variation across the country in relation to reviews. For example, the Royal Brompton Hospital Adult Centre did not meet a target of 90% of patients seen at least once per year for an annual review. At the University Hospital of South Manchester Adult Centre, 73% of patients were offered an annual review and only 66% attended the appointment. The reports for the other selected centres showed that the 90% target was met.

Nutritional assessment and interventions

The UK Cystic Fibrosis Registry Annual Data Report 2016 shows that 31.9% of people with cystic fibrosis received a form of supplementary feeding. The report adds that supplementary feeding is considered when a person has poor weight gain, or progressive weight loss, despite efforts to increase oral intake.

Peer Review reports include information on the availability of CF specialist dietitians at outpatient clinics, and reviews conducted during inpatient stays. There is a target of having a CF specialist dietitian available at all outpatient clinics. Only 2 of the 5 reports reviewed show the centres and networks are achieving this target across all outpatient clinics. There is also a target of ensuring that at least 60% of inpatients are reviewed 2 or more times during an inpatient stay. Of the selected providers, Birmingham Children's Hospital and shared care network was not achieving this target at all its sites.

Exercise

Exercise formed part of physiotherapy for 57.2% of people with cystic fibrosis (UK Cystic Fibrosis Registry Annual Data Report 2016).

Results from a survey of the provision of exercise testing and training in UK CF clinics[2] were published in 2010. 96 clinics responded to the survey. The results showed that exercise is underused as either an assessment tool or therapeutic intervention. For example, 38.9% of paediatric and 27.8% of adult patients had performed an exercise test in the preceding 12 months (most as part of an annual review process); and only 26.0% of clinics provided exercise training programmes.

Psychological assessment

Comments in the executive summaries of the Peer Review reports suggest that psychology services do not have an appropriate level of resource. All of the selected reports reviewed had comments to this effect. In most cases there was no

[2] D. Stevens, P.J. Oades, N. Armstrong, C.A. Williams, A survey of exercise testing and training in UK cystic fibrosis clinics, Journal of Cystic Fibrosis, Volume 9, Issue 5, 2010, Pages 302-306

psychologist, or an insufficient number of whole time equivalents, in the multidisciplinary team.

Liver disease

The UK Cystic Fibrosis Registry Annual Data Report 2016 shows that 13.7% of people on the register have liver disease. Peer Review reports show an expected compliance level of 100% of patients aged over 5 having a recoded abdominal ultrasound scan in the last 3 years. Most of the selected centres and networks reviewed were not achieving this level.

Cystic fibrosis related diabetes

29.5% of people with cystic fibrosis aged 10 or over are treated for cystic fibrosis related diabetes (UK Cystic Fibrosis Registry Annual Data Report 2016). The report shows that 1,363 (18.9% of those aged 10 or over) not known to have cystic fibrosis related diabetes were not screened for cystic fibrosis related diabetes.

Peer Review reports show that out of the selected centres and networks, only Glenfield Hospital Adult cystic fibrosis centre screened 100% of patients aged 12 years and older annually for cystic fibrosis related diabetes.

4.2.4 Resource impact

No significant resource impact is expected from implementing recommendations in the draft guidance.

4.3 Pulmonary management

4.3.1 Summary of suggestions

Airway clearance (access to adjuncts, nebulisers, airway clearance devices)

Access to therapy adjuncts and modern nebulisers was suggested. The stakeholder noted that financial constraints associated with airway clearance devices can lead to variable provision.

Treatment of infection (antibiotic therapy)

Aggressive and early treatment of infection was suggested as it is important to prevent lung deterioration in people with cystic fibrosis. The stakeholder said that there are different rates of use of preventative antibiotic therapy across centres, and variable access to inpatient and outpatient facilities for the treatment of acute infections.

Inhaled therapies

One stakeholder suggested use of inhaled therapies as they are key in maintaining and improving the clinical status of people with cystic fibrosis. They added that their use is recognised as a measure of the quality of service provision, and that national reports show there is variation.

4.3.2 Selected recommendations from development source

Table 7 below highlights recommendations that have been provisionally selected from the development source(s) that may support potential statement development. These are presented in full after table 7 to help inform the committee's discussion.

Table 7 Specific areas for quality improvement

Suggested quality improvement area	Suggested source guidance recommendations
Airway clearance (access to adjuncts, nebulisers, airway clearance devices)	**Airway clearance techniques** Cystic fibrosis. NICE clinical guideline in development. Recommendations 1.6.13, 1.6.14
Treatment of infection (antibiotic therapy)	There are numerous recommendations about different infections and antibiotic therapy in section 1.6 (Pulmonary infection section) of Cystic fibrosis, NICE clinical guideline in development. These recommendations are not shown in full below.

Inhaled therapies	There are numerous recommendations covering inhaled mucoactive agents and inhaled therapies for specific infections in section 1.6 of Cystic fibrosis, NICE clinical guideline in development. These recommendations are not shown in full below.

Airway clearance (access to adjuncts, nebulisers, airway clearance devices): Airway clearance techniques

Cystic fibrosis. NICE clinical guideline in development. Recommendation 1.6.13

When choosing an airway clearance technique for people with cystic fibrosis:

- assess their ability to clear mucus from their lungs, and offer an individualised plan to optimise this
- take account of the preferences of the person and (if appropriate) their parents and carers
- take account of any factors that may influence adherence.

Cystic fibrosis. NICE clinical guideline in development. Recommendation 1.6.14

Regularly assess the effectiveness of airway clearance techniques, and modify the technique or use a different one if needed.

4.3.3 Current UK practice

Airway clearance (access to adjuncts, nebulisers, airway clearance devices)

The UK Cystic Fibrosis Registry Annual Data Report 2016 contains information on the use of some airway clearance techniques. Specifically, it reports that 57.5% of people on the register received a form of positive expiratory pressure (this involves breathing out through a mask or mouthpiece against a resistance); and 1.9% a VEST technique (using an electric air pulse generator connected to an inflatable jacket to vibrate the chest).

Treatment of infection (antibiotic therapy)

The UK Cystic Fibrosis Registry Annual Data Report 2016 shows that 45.8% of people with cystic fibrosis received intravenous antibiotics. The report also identifies that 3,833 people with cystic fibrosis were on long-term azithromycin use. This equates to 39.5% of people with complete data on the register.

Inhaled therapies

The UK Cystic Fibrosis Registry Annual Data Report 2016 shows that 87.6% of people with chronic Pseudomonas aeruginosa (defined on the registry as 3 or more growths in the last 12 months) were receiving an inhaled antibiotic. The report also states that the consensus view in the UK is that 90% of people chronically infected with Pseudomonas aeruginosa should be prescribed at least one inhaled antibiotic.

Peer Review reports show there is variation between the centres in terms of the proportion of patients chronically infected with Pseudomonas aeruginosa on inhaled antibiotics. From the reports selected, the proportion ranged from 60% to 96%.

In relation to mucoactive agents, the UK Cystic Fibrosis Registry Annual Data Report 2016 shows that 59.7% of patients with cystic fibrosis are on DNase; 29.4% take hypertonic saline and 3.1% are on Mannitol. The 2016 report shows these proportions vary across centres and clinics. For example 49.0% of patients at South Manchester receive DNase treatment compared to 86.8% of adults at Royal Brompton.

4.3.4 Resource impact

No significant resource impact is expected from implementing recommendations in the draft guidance.

4.4 Preventing cross-infection

4.4.1 Summary of suggestions

Prevention of cross-infection was suggested by stakeholders. This included preventing transmission between people with cystic fibrosis, and also between patients and clinicians, across different settings. Comments stated that bacterial infection, including lung infection, can result in deterioration in clinical status; preclude people from lung transplant; and reduce life expectancy. Providing care through specialist centres results in grouping of people with cystic fibrosis and increases the potential for spread of infection. The importance of infection control policies and measures was recognised, but several stakeholders said these varied between cystic fibrosis centres. The emergence of new (and resistant pathogens) was flagged as a specific concern.

4.4.2 Selected recommendations from development source

Table 8 below highlights recommendations that have been provisionally selected from the development source that may support potential statement development. These are presented in full after table 8 to help inform the committee's discussion.

Table 8 Specific areas for quality improvement

Suggested quality improvement area	Suggested source guidance recommendations
Preventing cross-infection	**Information and support** Cystic fibrosis. NICE clinical guideline in development. Recommendation 1.2.7 **Service configuration** Cystic fibrosis. NICE clinical guideline in development. Recommendation 1.3.2 **Preventing cross-infection** Cystic fibrosis. NICE clinical guideline in development. Recommendations 1.8.2 to 1.8.4, and 1.8.6 to 1.8.8

Information and support

Cystic fibrosis. NICE clinical guideline in development. Recommendation 1.2.7

Provide people with cystic fibrosis with information about how to contact other people with cystic fibrosis without risking cross-infection (see Preventing cross-infection), for example, by directing them to online support groups.

Service configuration

Cystic fibrosis. NICE clinical guideline in development. Recommendation 1.3.2

Specialist cystic fibrosis centres should:

- plan patient care (including outpatient and inpatient care), taking into account the risk of cross-infection (see Preventing cross-infection)
- maintain local and national registers of patients that include information about their clinical condition, treatment and outcomes
- audit practice and outcomes.

Preventing cross-infection

Cystic fibrosis. NICE clinical guideline in development. Recommendation 1.8.2

To prevent cross-infection among people with cystic fibrosis, use microbiological surveillance and a local infection control strategy that covers outpatient and inpatient care.

Cystic fibrosis. NICE clinical guideline in development. Recommendation 1.8.3

Inform people with cystic fibrosis, their family members or carers (as appropriate) and staff involved in their care about the risk of cross-infection and how to avoid it.

Cystic fibrosis. NICE clinical guideline in development. Recommendation 1.8.4

Separate people with cystic fibrosis who have transmissible or chronic pseudomonas aeruginosa or Burkholderia cepacia complex infection, for example during outpatient clinics.

Cystic fibrosis. NICE clinical guideline in development. Recommendation 1.8.6

All specialist cystic fibrosis clinics should be organised to prevent cross-infection. Separate people at outpatient clinics, for example by managing:

- use of communal waiting areas
- attendance at diagnostic and treatment facilities.

Cystic fibrosis. NICE clinical guideline in development. Recommendation 1.8.7

Help people with cystic fibrosis plan their inpatient attendance to avoid contact with each other, for example when they use:

- hospital restaurants, schools and recreation areas

- diagnostic and treatment facilities.

Cystic fibrosis. NICE clinical guideline in development. Recommendation 1.8.8

During inpatient care, give people with cystic fibrosis individual rooms with en-suite facilities.

4.4.3 Current UK practice

Peer Review reports present two measures of infection control. One is the proportion of patients cared for in single, en-suite rooms during hospital admission. The target for compliance is 100%. From the reports selected, one centre (Birmingham Children's Hospital and shared care network) did not meet this target.

The second measure shows whether a target of 100% of patients cohorted to outpatient clinics according to microbiological status was being met. Of the selected reports, Glenfield Hospital Adult Cystic Fibrosis Centre, and two of the network clinics forming part of the Birmingham Children's Hospital and shared care network, were not achieving this target. The reports noted that the 2 network clinics were small so achieving the target may not be feasible.

4.4.4 Resource impact

No significant resource impact is expected from implementing recommendations in the draft guidance.

4.5 Service delivery

4.5.1 Summary of suggestions

Access to care

Access to appropriate treatment was suggested by one stakeholder who stated there are inequalities in access, and gave an example of disadvantaged patients being more likely to receive antibiotic and nutritional treatments but less likely to receive inhaled therapies. A stakeholder suggested equality of access between regions to expensive drugs.

Other stakeholders emphasised the importance of ensuring that the quality of care and outcomes for those in shared care are as good as those cared for by the centre alone. Shared care is a model of care where a local hospital cares for people with cystic fibrosis with oversight, support and direct involvement from a specialist cystic fibrosis centre. Recording the number of times shared care patients are reviewed by the centre was suggested as a measure.

Several stakeholders suggested access to homecare as it has numerous benefits, such as reducing the burden of care for people with cystic fibrosis, but commented that there is widespread variation in its provision. Some suggested what homecare should involve, such as reviews, routine monitoring, intravenous (IV) antibiotic therapy etc.

Timeliness of inpatient admission, and having sufficient inpatient beds, were suggested by stakeholders, and one commented that there are increasing patient numbers.

Stakeholders highlighted the need for cystic fibrosis care to be provided by a multidisciplinary team made up of specialists. Several stakeholders said there is variation in access to such teams across the country, and in some cases teams may not be complete or have the appropriate level of experience.

Transition to adult services

Stakeholders recognised the transition to adult services as a difficult time for young people with cystic fibrosis; that it could affect their health and clinical status; and that there is national variation in the process. A common theme in the comments was that transition needed to be timely and coordinated.

4.5.2 Selected recommendations from development source

Table 9 below highlights recommendations that have been provisionally selected from the development source(s) that may support potential statement development. These are presented in full after table 9 to help inform the committee's discussion.

Table 9 Specific areas for quality improvement

Suggested quality improvement area	Selected source guidance recommendations
Access to care	**Service configuration** Cystic fibrosis. NICE clinical guideline in development. Recommendations 1.3.1, 1,33, 1.3.6 and 1.3.7 **Multidisciplinary team** Cystic fibrosis. NICE clinical guideline in development Recommendation 1.3.8 **Pulmonary monitoring** Cystic fibrosis. NICE clinical guideline in development. Recommendation 1.6.6
Transition to adult services	**Transition to adult services** Cystic fibrosis. NICE clinical guideline in development. Recommendations 1.3.16, 1.3.17 and 1.3.19

Access to care: Service configuration

Cystic fibrosis. NICE clinical guideline in development. Recommendation 1.3.1

Care for people with cystic fibrosis should be provided by a specialist cystic fibrosis multidisciplinary team based at a specialist cystic fibrosis centre (see Multidisciplinary teams).

Cystic fibrosis. NICE clinical guideline in development. Recommendation 1.3.3

When a shared-care model is used for children and young people, it should include:

- formal arrangements between the local paediatric team at the shared-care centre and multidisciplinary team at the specialist cystic fibrosis centre
- direct involvement of specialist cystic fibrosis multidisciplinary team members
- an annual assessment and at least one other review per year by the specialist cystic fibrosis multidisciplinary team in addition to reviews by the local paediatric team (see Annual and routine reviews).

Cystic fibrosis. NICE clinical guideline in development. Recommendation 1.3.6

Consider telemedicine or home visits for routine monitoring where they are more appropriate than outpatient visits and if the person with cystic fibrosis prefers it.

Cystic fibrosis. NICE clinical guideline in development. Recommendation 1.3.7

Make arrangements (including providing equipment and expert support) for people to have intravenous antibiotic therapy at home, when this is appropriate.

Access to care: Multidisciplinary team

Cystic fibrosis. NICE clinical guideline in development. Recommendation 1.3.8

The specialist cystic fibrosis multidisciplinary team should include the following professionals who have specialist expertise in the condition:

- specialist paediatrician or adult physician
- specialist nurse
- specialist physiotherapist
- specialist dietitian
- specialist pharmacist
- specialist clinical psychologist
- social worker.

Access to care: Pulmonary monitoring

Cystic fibrosis. NICE clinical guideline in development. Recommendation 1.6.6

For people with cystic fibrosis with lung disease who have symptoms that are concerning them, their family members or carers (as appropriate) or a healthcare professional, decide whether a remote telemedicine (see telemedicine) or face-to-face assessment is needed and consider which of the following may be useful:

- review of the past history
- physical examination, including measurement of weight and length (if less than 2 years old) or height
- measurement of oxygen saturation
- collection of respiratory secretion samples for microbiological investigations, using sputum samples if possible, or a cough swab or NPA if not
- for adults, blood tests to measure white cell count and inflammatory markers such as C-reactive protein
- lung function testing, for example with spirometry (including FEV1, FVC, and FEF 25–75%) in adults, and in children and young people who can do this
- lung clearance index for people with normal spirometry results.

Transition to adult services: Transition to adult services

Cystic fibrosis. NICE clinical guideline in development. Recommendation 1.3.16

Begin the transition process to adult services with young people with cystic fibrosis when they are 12 years old, and with their family members or carers (as appropriate).

Cystic fibrosis. NICE clinical guideline in development. Recommendation 1.3.17

Ensure that young people with cystic fibrosis move to adults' services between the ages of 16 and 18.

Cystic fibrosis. NICE clinical guideline in development. Recommendation 1.3.19

Provide a named worker to lead the transition for each young person with cystic fibrosis who is moving to adults' services, and if possible ensure that the named worker is someone the young person already knows. For more guidance on named workers see the section on named workers in the NICE guideline on transition for young people using health or social care services

4.5.3 Current UK practice

Access to care

In relation to homecare, the UK Cystic Fibrosis Registry Annual Data Report 2016 provides information on people with cystic fibrosis who received intravenous (IV) antibiotics. This treatment can take place as an inpatient or at home. The report shows that 53.3% of 4,444 people receiving IV antibiotics had the treatment at home.

Cystic Fibrosis Trust Peer Review reports patient surveys asked patients to rate their cystic fibrosis team in relation to 'homecare / community'. The results for the selected centres and networks show that 70%-100% of patients rated their CF team good or excellent for this aspect of their care. However, the results do not identify what proportion of patients were unable to access homecare.

Peer Review reports also cover shared care networks. The Birmingham Children's Hospital and shared care network report shows that two-thirds of patients are treated on a shared care basis, and one third on a full care basis by the regional centre. Overall the centre and network care is described as being of 'good quality'. However, there was a key recommendation for joint network MDT clinics (comprising local CF MDT and centre CF MDT seeing patients jointly) to be established at each network clinic.

The reports also review the proportion of patients admitted within seven days of a 'decision to admit and treat' against an expected target of 100%. At the time of the

reports, 2 cystic fibrosis centres were not meeting this target, and waits of several weeks for admission were experienced in some instances.

Turning multidisciplinary teams (MDTs), comments in the executive summaries of the selected reports generally describe the MDTs as expert and experienced. However, every centre and network had low numbers of MDT staff compared to recommended levels.

Transition to adult services

Comments in the Cystic Fibrosis Trust Peer Review reports suggest there was variation in practice at the time the reports were produced. At Birmingham Children's Hospital and shared care network, the quality of the transition process is described as 'patchy across the network', mainly due to 'severe pressure on adult services'. A key recommendation was to support development of a new adult centre. In contrast, Glenfield Hospital Adult Cystic Fibrosis Centre is described as having a 'strong approach' to transition with bi-monthly joint transition clinics with paediatric colleagues, and the transition service is described later in the report as exemplary.

4.5.4 Resource impact

No significant resource impact is expected from implementing recommendations in the draft guidance.

4.6 Additional areas

Summary of suggestions

The improvement areas below were suggested as part of the stakeholder engagement exercise. However they were felt to be either unsuitable for development as quality statements, outside the remit of this particular quality standard referral or require further discussion by the committee to establish potential for statement development.

There will be an opportunity for the committee to discuss these areas at the end of the session on 12 October.

Lung transplant

A stakeholder suggested lung transplant. They added that improved understanding of the timing of referral, evaluation process, follow-up and outcomes can drive quality improvement. There are no recommendations in the source guidance that cover referral for, and management of, transplantation. NICE has published Living-donor lung transplantation for end-stage lung disease (NICE interventional procedure guidance 170) but this covers only the efficacy and safety of the procedure.

Management of CF-SPID [Screen Positive, Inconclusive Diagnosis] group

This was suggested both as an area for quality improvement and as an additional developmental area of emergent practice. There are no recommendations in the development source which cover this. There are recommendations on diagnosis of cystic fibrosis, but the main focus of the management recommendations is people diagnosed with cystic fibrosis.

Management of the upper airway in cystic fibrosis patients

This was suggested by a stakeholder as an additional developmental area of emergent practice. Management of the upper airway is not covered in the source guidance. The only coverage in the cystic fibrosis guideline in development is recommendation 1.4.1 which says to be aware that people with cystic fibrosis are at risk of upper airway complications.

Monitoring adherence to treatments

A stakeholder suggested adherence as a known issue for people with cystic fibrosis, but noted there is no national guidance focusing on measurement of adherence to cystic fibrosis treatments. Adherence is referenced in the source guidance, but there are no recommendations setting out how adherence should be measured. Medicines

adherence (NICE clinical guideline CG76) is a potential source, but it only covers adults and is not specific to cystic fibrosis.

Palliative care in cystic fibrosis

A stakeholder described this as an emerging area, and suggested development of a pathway for joint management between cystic fibrosis and palliative care teams. End of life care for adults (NICE quality standard QS13) quality statement 8 covers this by saying that people approaching the end of life receive care that is coordinated across all relevant settings and services.

Physiotherapist's role relating to CFTR [Cystic Fibrosis Transmembrane Conductance Regulator] channel modulators

This was suggested by a stakeholder as an additional developmental area of emergent practice. This is not covered within the development source.

Registers and audit

The quality and completeness of data collected for the UK cystic fibrosis registry was suggested. A stakeholder also highlighted the importance of capturing information on patient experience as this can be used to drive up standards.

These suggestions have not been progressed. Participation in audit and collation of experience information are methods by which quality improvement can be evidenced. Quality statements focus on actions that demonstrate high quality care or support, not the methods by which evidence is collated. However, audits and suggested methods of data collection may be referred to in the data sources for quality measures.

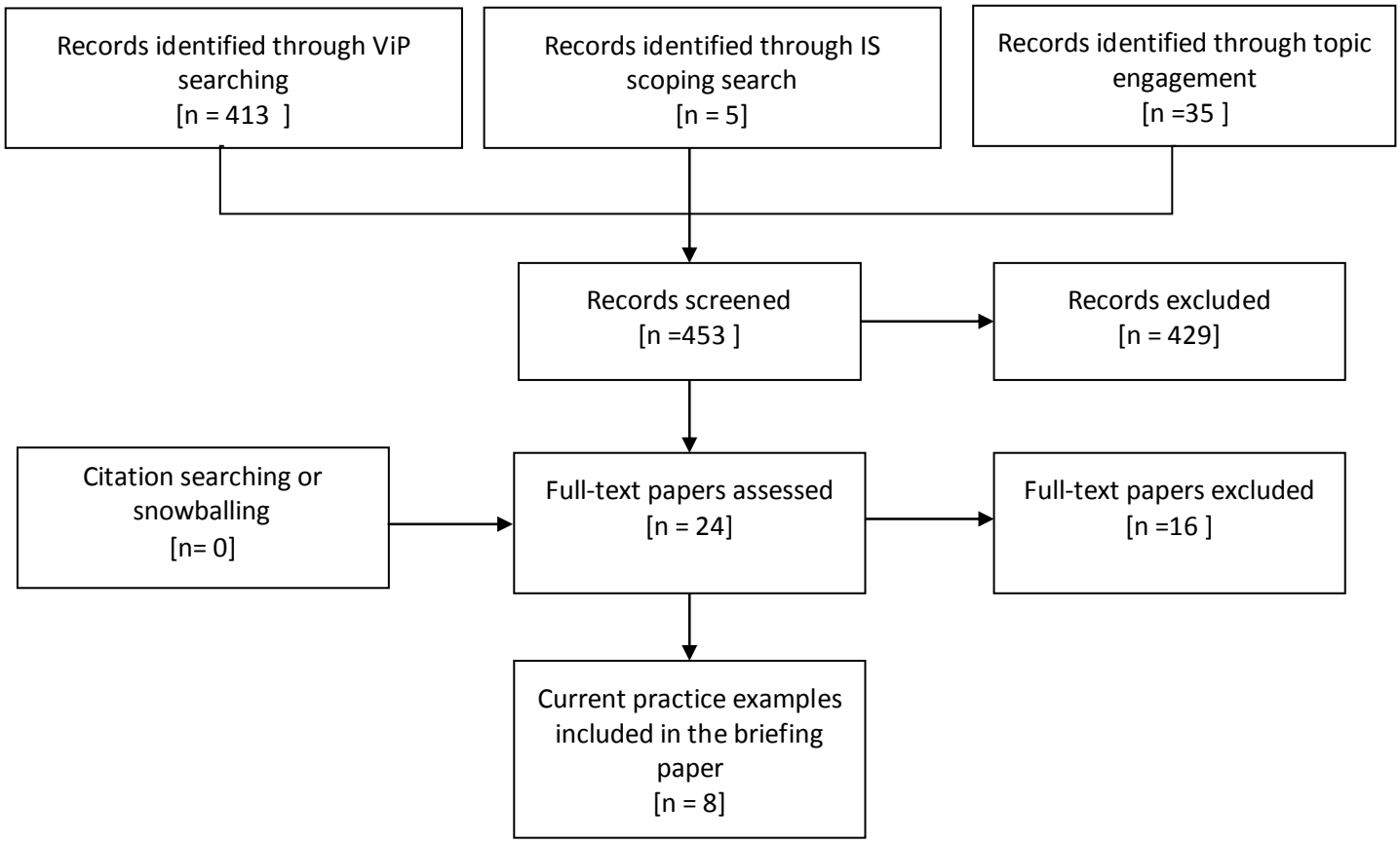
Technology in airway clearance

This was suggested by a stakeholder as an additional developmental area of emergent practice. No further details were provided.

Time to diagnostic assessment

Timeliness of referral and of the diagnostic assessment were identified by stakeholders. The time taken to the first clinical appointment following a positive screen of a baby was specifically suggested by a stakeholder as the area for improvement. There are no recommendations in the source guidance that cover timescales for diagnostic assessment. In addition, timely receipt into clinical care of babies who screen positive is covered by the UK National Screening Committee and NHS Screening Programmes standards.

Appendix 1: Review flowchart



Appendix 2: Suggestions from stakeholder engagement exercise – registered stakeholders

Diagnosis					
ID	**Stakeholder**	**Suggested key area for quality improvement**	**Why is this important?**	**Why is this a key area for quality improvement?**	**Supporting information**
1	Public Health England / UK National Screening Committee	Sweat test for diagnosis of cystic fibrosis(CF) following screen positive from newborn blood spot screening	A positive sweat test is a key component of making a CF diagnosis. https://www.gov.uk/government/uploads/system/uploads/attachment_data/file/411444/CF_clinical_referral_guidelines.pdf	The UK Newborn Blood Spot Screening annual data collection provides the evidence that not all cystic fibrosis screen positive babies receive a sweat test	Recent international consensus guidelines reiterate the need for a sweat test result on all infants with a positive newborn bloodspot screening result for CF (ref Farrell et al. 2016, pubmed ID 28129811)
2	Cystic Fibrosis Trust	CFTR mutation genotyping of people with CF	As genotype-targeted therapies are becoming available, it is critically important that individuals have access to this information, to benefit from emerging treatments.	The CF Trust's and UK CF Registry's 'Genotype Matters' campaign has driven numbers of patients with complete genotyping of their CFTR mutations to record levels. Clinics should aim for 100% coverage and accuracy.	The UK CF Registry has records for 98.4% of patients with complete annual data about both CFTR mutations. Please see: https://www.cysticfibrosis.org.uk/the-work-we-do/uk-cf-registry/reporting-and-resources
3	NHSE	[Comments from 2 respondents] Check ALL patients have been genotyped, - very important in case they have a rare gating mutation that means they should receive ivacaftor.			Relevant national audits – CF Registry, and Quality dashboards.

Diagnosis					
ID	**Stakeholder**	**Suggested key area for quality improvement**	**Why is this important?**	**Why is this a key area for quality improvement?**	**Supporting information**
		This is particularly important with genotype specific therapies now becoming available			

Monitoring, assessment and management					
ID	**Stakeholder**	**Suggested key area for quality improvement**	**Why is this important?**	**Why is this a key area for quality improvement?**	**Supporting information**
4	SCM 3	Monitoring of disease progression in people with cystic fibrosis	As people with cystic fibrosis grow older, new cystic fibrosis-related conditions emerge which require specialist management	There is variable assessment of these cystic fibrosis-related conditions.	See NICE CF guideline indicating monitoring of CF-related multisystem conditions at: https://www.nice.org.uk/guidance/indevelopment/gid-cgwave0736
5	SCM 4	People with CF should have regular reviews by an expert multidisciplinary team	CF is a complex condition. Professionals with relevant experience should provide tailored care for people with CF in order to improve outcomes for this group of patients.	Ensure improved quality of life for all people with CF.	NICE guidance CF Trust Standards of Care Specialised commissioning specifications
6	NHSE	Number of patients who have an annual assessment			Relevant national audits – CF Registry, and Quality dashboards.

Monitoring, assessment and management					
ID	**Stakeholder**	**Suggested key area for quality improvement**	**Why is this important?**	**Why is this a key area for quality improvement?**	**Supporting information**
7	Abbott	Regular nutritional screening and management of malnutrition	It is well known that nutritional depletion is a common problem in patients with severe chronic lung diseases such as cystic fibrosis (CF). Hypermetabolism, malabsorption and depletion of fat free mass are associated with increased morbidity and significant impairment of health status.[1] Recent European guidelines[3] recommend the following: Monitoring of growth and/or nutritional status at regular intervals to determine the adequacy of pancreatic enzyme replacement therapy; monitor at every clinic visit for infants, every 3 months for older children and adolescents, and every 6 months for adults. Clinicians consider the use of oral nutritional supplements for treating children and adults who fail to achieve optimal growth rates and nutritional status with oral dietary intake and pancreatic enzyme replacement therapy (PERT) alone. Clinicians regularly review and re-evaluate patients who are taking	Koen, 2008, [2] reports that malnutrition is still prevalent in this patient group, with rates of between 8.3% - 42% being reported in children with CF.	Please see the ESPEN-ESPGHAN-ECFS guidelines on nutrition care for infants, children, and adults with cystic fibrosis: http://www.clinicalnutritionjournal.com/article/S0261-5614(16)00095-9/fulltext

Monitoring, assessment and management					
ID	**Stakeholder**	**Suggested key area for quality improvement**	**Why is this important?**	**Why is this a key area for quality improvement?**	**Supporting information**
			oral nutritional supplements to determine whether the patient should continue taking them. Clinicians consider the use of polymeric enteral tube feeding when oral interventions have failed to achieve acceptable rates of growth and nutritional status. (If this is not well tolerated, an elemental or semi-elemental feed may be beneficial). Basing the route, formula, and timing for enteral feeding selection on individual needs and preferences. Advising patients on macronutrient balance in the diet, with attention to protein and fat intake that is sufficient to prevent or delay loss of muscle mass and function. There is a clear link between good nutritional status and better lung function, which improves clinical outcomes and survival. Nutritional care and support should be an integral part of management of CF. Obtaining a normal growth pattern in children and maintaining an adequate nutritional status in adults are		

Monitoring, assessment and management					
ID	**Stakeholder**	**Suggested key area for quality improvement**	**Why is this important?**	**Why is this a key area for quality improvement?**	**Supporting information**
			major goals of multidisciplinary cystic fibrosis centres.[3] Soto-Martínez ME., 2017. Pediatric Pulmonology, suppl. Supplement 46 52: S65-S67. Koen FM et al., 2008. Current Opinion in Pediatrics. 20: p590-596 ESPEN-ESPGHAN-ECFS, 2016: https://www.ncbi.nlm.nih.gov/pubmed/27068495		
8	Cystic Fibrosis Dietitians Group UK	Nutritional assessment and management strategies for cystic fibrosis across increasingly diverse population	Poor growth and malnutrition contribute to increased mortality and morbidity in CF and have a long-term impact on lung function. Factors affecting energy requirements include age, gender, nutritional status, chronic and acute infection, gastro-oesophageal reflux and control of malabsorption and maldigestion. (in those who are pancreatic insufficient) Reliable and are an essential part of the clinical assessment of health status in CF	All people with CF should have sequential measurements of growth and nutritional status and be seen regularly by a Specialist CF Dietitian, to assess and address all factors which may compromise appetite, malabsorption and maldigestion and dietary intake. In children aged 2–18 years nutritional advice and interventions should be aimed at maintaining BMI on or above the 50th percentile and in adults, the aim should be achieving a BMI of 22kg/m2 in women and 23kg/m2 in men, as these are associated with better lung function. However 1) Emerging targeted medical therapies are now focusing on Class of mutation. Nutritional and	Stallings VA, Stark LJ, Robinson KA, Feranchak AP, Quinton H. Evidence-based practice recommendations for nutrition-related management of children and adults with cystic fibrosis and pancreatic insufficiency: results of a systematic review. J Am Diet Assoc. 2008;108(5):832–9. Yen EH, Quinton H, Borowitz D. Better nutritional status in early childhood is associated with improved clinical outcomes and survival in

Monitoring, assessment and management					
ID	**Stakeholder**	**Suggested key area for quality improvement**	**Why is this important?**	**Why is this a key area for quality improvement?**	**Supporting information**
				management strategies according to Class of mutation remain unexplored and may require differing nutritional management strategies 2) Outcomes for nutritional status have been traditionally measured by percentile position and BMI. Increased longevity and emerging research suggests that lean body mass may be a more useful indicator of prognosis and outcome. Nutritional strategies to better promote increased LBM are unexplored	patients with cystic fibrosis. J Pediatr. 2013;162(3):530–5. Somaraju UR, Solis-Moya A. Pancreatic enzyme replacement therapy for people with cystic fibrosis. Cochrane database Syst Rev. 2014;10:CD008227 https://www.cysticfibrosis.org.uk/~/media/documents/the-work-we-do/care/consensus-documents-with-old-address/nutritional-management-of-cystic-fibrosis-sep-16.ashx?la=en
9	SCM 1	Nutritional assessment to include body composition, and management strategies for cystic fibrosis across an increasingly diverse population.	It is accepted that poor growth and malnutrition contribute to increased mortality and morbidity in CF and have a long-term impact on lung function. Factors affecting energy requirements are multi-factorial including age, gender, nutritional status, chronic and acute infection, gastro-oesophageal reflux, and control of malabsorption and maldigestion in patients with pancreatic insufficiency.	It is current good practice well known that all people with CF should have sequential measurements of growth and nutritional status and be seen regularly by a Specialist CF Dietitian, to assess and address all factors which may compromise appetite, malabsorption and maldigestion and dietary intake. In children aged 2–18 years nutritional advice and interventions should be aimed at maintaining BMI on or above the 50th percentile and in adults, the aim should be achieving a BMI of 22kg/m2 in women and 23kg/m2 in men,	Please see: Stallings VA, Stark LJ, Robinson KA, Feranchak AP, Quinton H. Evidence-based practice recommendations for nutrition-related management of children and adults with cystic fibrosis and pancreatic insufficiency: results of a systematic

Monitoring, assessment and management					
ID	**Stakeholder**	**Suggested key area for quality improvement**	**Why is this important?**	**Why is this a key area for quality improvement?**	**Supporting information**
			Reliable and up to date evidence based nutritional care are an essential part of the clinical treatment for optimising health status and outcomes for patients with CF	as these are associated with better lung function. However, advances in our knowledge of CF and the treatment challenges and options, requires new evidence and improvement for the nutritional approach and outcomes. For example: 1) Emerging targeted medical therapies are now focusing on Class of mutation. Nutritional and management strategies according to Class of mutation remain unexplored and may require differing nutritional management strategies 2) Outcomes for nutritional status have been traditionally measured by percentile position and BMI. Increased longevity and emerging research suggests that lean body mass may be a more useful indicator of prognosis and outcome. Nutritional strategies to better promote increased LBM are unexplored. (This information would enhance and accompany any QI areas that target exercise programmes in CF, and the effect of exercise on body composition , nutritional requirements etc. And where additionally, another crucial nutritional unexplored / non- evidenced area is dietary salt requirements. CF is a salt loosing	review. J Am Diet Assoc. 2008; 108(5):832–9. Yen EH, Quinton H, Borowitz D. Better nutritional status in early childhood is associated with improved clinical outcomes and survival in patients with cystic fibrosis. J Pediatr. 2013; 162(3):530–5. Somaraju UR, Solis-Moya A. Pancreatic enzyme replacement therapy for people with cystic fibrosis. Cochrane database Syst Rev. 2014;10:CD008227 https://www.cysticfibrosis.org.uk/~/media/documents/the-work-we-do/care/consensus-documents-with-old-address/nutritional-management-of-cystic-fibrosis-sep-16.ashx?la=en

Monitoring, assessment and management					
ID	**Stakeholder**	**Suggested key area for quality improvement**	**Why is this important?**	**Why is this a key area for quality improvement?**	**Supporting information**
				condition, and exercise affects salt / electrolyte requirements.)	
10	SCM 2	Pancreatic Enzyme Replacement Therapy	Poor growth and malnutrition contribute to increased mortality and morbidity in CF .It is a fundamental requirement that pancreatic insufficient new-borns with CF have access to a specialist dietician and are supported to give PERT along with reliable and up to date evidence based nutritional care.	There is little evidence written about this subject. CF Nurses and Dieticians work closely with parents and carers in the first year following diagnosis as education with regard to feeding/ nutrition is key to continued growth.	https://www.cysticfibrosis.org.uk/~/media/documents/the-work-we-do/care/consensus-documents-with-old-address/nutritional-management-of-cystic-fibrosis-sep-16.ashx?la=en
11	Association of Chartered Physiotherapists in Cystic Fibrosis	Exercise opportunities in the CF population	There is good evidence that regular exercise is beneficial to CF patients and is recommended by NICE. It should be encouraged at all stages of the disease process	Exercise is recognised as having significant potential to improve lung function and quality of life and therefore increase life expectancy in patients with CF. It is also recognised that pre-transplant rehabilitation influences clinical outcomes. Establishment of a healthy activity based lifestyle from diagnosis through to ongoing participation in regular exercise throughout all stages of the disease is an extremely important aspect of care.	Standards of Care and Good Clinical Practice for The Physiotherapy Management of Cystic Fibrosis 2017 file:///C:/Users/user/AppData/Local/Packages/Microsoft.MicrosoftEdge_8wekyb3d8bbwe/TempState/Downloads/Consensus%20on%20physiotherapy%20management%20%20third%20edition%202017.pdf ECFS Statement on Exercise https://www.ecfs.eu/ecfs_exercise_wg/publications

Monitoring, assessment and management					
ID	**Stakeholder**	**Suggested key area for quality improvement**	**Why is this important?**	**Why is this a key area for quality improvement?**	**Supporting information**
12	SCM 1	Exercise opportunities in the CF population	There is good evidence that regular exercise is beneficial to CF patients and is recommended by NICE. It should be encouraged at all stages of the disease process	Exercise is recognised as having significant potential to improve lung function and quality of life and therefore increase life expectancy in patients with CF. It is also recognised that pre-transplant rehabilitation influences clinical outcomes. Establishment of a healthy activity based lifestyle from diagnosis through to on-going participation in regular exercise throughout all stages of the disease is an extremely important aspect of care. Additionally , the effect that exercise has on energy requirements and body composition is important to consider and remains unexplored in CF.	Please see: Standards of Care and Good Clinical Practice for The Physiotherapy Management of Cystic Fibrosis 2017 file:///C:/Users/user/AppData/Local/Packages/Microsoft.MicrosoftEdge_8wekyb3d8bbwe/TempState/Downloads/Consensus%20on%20physiotherapy%20management%20%20third%20edition%202017.pdf ECFS Statement on Exercise https://www.ecfs.eu/ecfs_exercise_wg/publications
13	SCM 5	Access to an individualised exercise programme	The importance of exercise is acknowledged by multiple recommendations in the draft NICE guideline for cystic fibrosis. The draft guideline acknowledges that regular exercise improves both lung function and overall fitness and so people with cystic	There is significant variation and underutilisation in exercise assessment and provision across CF centres in the UK as demonstrated by previous peer review reports and published papers.	See past peer review reports: https://www.cysticfibrosis.org.uk/the-work-we-do/clinical-care/peer-reviews Also see past publications which demonstrate variation

Monitoring, assessment and management					
ID	**Stakeholder**	**Suggested key area for quality improvement**	**Why is this important?**	**Why is this a key area for quality improvement?**	**Supporting information**
			fibrosis should be offered an individualised exercise programme which is regularly reviewed.		and under-utilisation of exercise/physical activity: D. Stevens, P.J. Oades, N. Armstrong, C.A. Williams, A survey of exercise testing and training in UK cystic fibrosis clinics, Journal of Cystic Fibrosis, Volume 9, Issue 5, 2010, Pages 302-306 Craig A. Williams, Daniel Stevens, Physical activity and exercise training in young people with cystic fibrosis: Current recommendations and evidence, Journal of Sport and Health Science, Volume 2, Issue 1, 2013, Pages 39-46
14	British Thoracic Society	Psychology input especially in newly diagnosed patients	Psychology review is seen as best practice in CF centres, but scope of reviews and detailed data on number and type of review, is not currently part of quality standard. Also newly diagnosed patients need input	Currently 1 review per year prior to annual review is what is incorporated into the quality standard Perhaps number and types of Psychology reviews (IP CF OP, separate psychology reviews, and annual review) should be recorded and be considered for incorporation into quality standards.	Specialised services quality dashboard: Cystic fibrosis – adults https://www.england.nhs.uk/commissioning/spec-services/npc-crg/spec-dashboards/

Monitoring, assessment and management					
ID	**Stakeholder**	**Suggested key area for quality improvement**	**Why is this important?**	**Why is this a key area for quality improvement?**	**Supporting information**
15	Cystic Fibrosis Dietitians Group UK	Management of Cystic Fibrosis Related Diabetes	Cystic fibrosis-related diabetes is the most common co-morbidity in CF, affecting 20 percent of adolescents and 40–50 percent of adults over the age of 30 years Cystic fibrosis-related diabetes is associated with deterioration in clinical status and contributes to poor nutritional status, decreased lung function, more frequent hospital admissions and increased mortality. Early diagnosis and treatment have been associated with a decline in mortality rates in CFRD.	Annual OGTT to screen for CFRD should begin by the age of 10 years in all people with CF who do not have CFRD. However there is continued debate owing to sensitivity of this test and the changing glycaemic state of patients as they slowly move towards CFRD Insulin and individualised dietary education is the recommended treatment for CFRD, but there is a lack of evidence a) supporting the type of educational programme used and how these should be implemented for CFRD and outcomes defined b) Optimal timing of insulin as individuals slowly move into CFRD c) The outcomes associated with best practice in receiving care from a Diabetologist in close liaison with the Cystic fibrosis MDT	Cystic Fibrosis Trust. Management of cystic fibrosis related diabetes mellitus. Bromley; 2004. Moran A, Dunitz J, Nathan B, Saeed A, Holme B, Thomas W. Cystic fibrosis-related diabetes: Current trends in prevalence, incidence, and mortality. Diabetes Care. 2009;32(9):1626–31. Kelly A, Moran A. Update on cystic fibrosis-related diabetes. J Cyst Fibros. 2013;12(4):318–31. Moran A, Pekow P, Grover P, Zorn M, Slovis B, Pilewski J, et al. Insulin therapy to improve BMI in cystic fibrosis-related diabetes without fasting hyperglycemia: Results of the cystic fibrosis related diabetes therapy trial. Diabetes Care. 2009;32(10):1783–8.

Monitoring, assessment and management					
ID	**Stakeholder**	**Suggested key area for quality improvement**	**Why is this important?**	**Why is this a key area for quality improvement?**	**Supporting information**
					https://www.cysticfibrosis.org.uk/~/media/documents/the-work-we-do/care/consensus-docs-with-new-address/cfrd-mellitus.ashx?la=en
16	SCM 1	Management of Cystic Fibrosis Related Diabetes	Cystic fibrosis-related diabetes is the most common co-morbidity in CF, affecting 20 percent of adolescents and 40–50 percent of adults over the age of 30 years Cystic fibrosis-related diabetes is associated with deterioration in clinical status and contributes to poor nutritional status, decreased lung function, more frequent hospital admissions and increased mortality. Early diagnosis and treatment have been associated with a decline in mortality rates in CFRD.	Annual OGTT to screen for CFRD should begin by the age of 10 years in all people with CF who do not have CFRD. However there is continued debate owing to sensitivity of this test and the changing glycaemic state of patients as they slowly move towards CFRD Insulin and individualised dietary education is the recommended treatment for CFRD, but there is a lack of evidence in the following areas: : a) Supporting the type of educational programme used and how these should be implemented for CFRD and outcomes defined b) Optimal timing of insulin as individuals slowly move into CFRD c) The outcomes associated with best practice in receiving care from a Diabetologist in close liaison with the Cystic fibrosis MDT	Please see: Cystic Fibrosis Trust. Management of cystic fibrosis related diabetes mellitus. Bromley; 2004. Kelly A, Moran A. Update on cystic fibrosis-related diabetes. J Cyst Fibrosis. 2013; 12(4):318–31. https://www.cysticfibrosis.org.uk/~/media/documents/the-work-we-do/care/consensus-docs-with-new-address/cfrd-mellitus.ashx?la=en
17	SCM 2	Liver Disease	Liver disease is the third most common cause of mortality in adults with CF and around 10-30% will develop CF related liver disease. Therefore effective	A widespread variation in assessment and management of CF liver disease. More research is needed looking at the effectiveness of Ursodeoxycholic acid in children.	CF related liver disease fact sheet Feb 2017. www.ncvinlm.nih.gov/pmc/articles/PMC4110359/ Liver disease in CF 2014.

Monitoring, assessment and management					
ID	**Stakeholder**	**Suggested key area for quality improvement**	**Why is this important?**	**Why is this a key area for quality improvement?**	**Supporting information**
			monitoring in childhood is paramount.		Cystic Fibrosis and liver disease a guide. Children's liver disease foundation.

Pulmonary management					
ID	**Stakeholder**	**Suggested key area for quality improvement**	**Why is this important?**	**Why is this a key area for quality improvement?**	**Supporting information**
18	Association of Chartered Physiotherapists in Cystic Fibrosis	Access to therapy adjuncts and modern nebulisers	NICE guidance recommends regular discussion regarding the use of Airway Clearance Techniques (ACT's) and training in an appropriate ACT for those with lung disease. It is important to offer an individualised plan to optimise mucous clearance. There are a variety of modern nebuliser systems available to enhance access and adherence to inhalation therapy. It is important that these are widely available.	All patients are individual with respect to preferences associated with airway clearance. Frequently there are financial constraints associated with their provision and this is variable nationally. We think it is important that there is easy access to all forms of airway clearance devices to enable holistic management and improvements in adherence and subsequent clinical outcomes. Modern nebulisers are costly to provide and to support by way of consumables. They allow access to inhaled therapies such as Cayston and Quinsair and improve adherence. They should be widely available to all patients with CF to enable optimal management of their microbiology	Standards of Care and Good Clinical Practice for The Physiotherapy Management of Cystic Fibrosis 2017 file:///C:/Users/user/AppData/Local/Packages/Microsoft.MicrosoftEdge_8wekyb3d8bbwe/TempState/Downloads/Consensus%20on%20physiotherapy%20management%20%20third%20edition%202017.pdf
19	SCM 3	Treatment of infection in people with cystic fibrosis	Aggressive and early treatment of infection is important to prevent	There are different rates of use of preventative antibiotic therapy in different centres, and	See NICE CF guideline which indicates standards for antibiotic treatment at:

Pulmonary management					
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			lung deterioration in people with cystic fibrosis	variable access to inpatient and outpatient facilities for the treatment of acute infections	https://www.nice.org.uk/guidance/indevelopment/gid-cgwave0736
20	SCM 5	Use of key inhaled therapies	There is good evidence that the use of inhaled therapies such as mucolytics, osmotics and nebulised antibiotics are key in maintaining and improving the clinical status of people with cystic fibrosis.	The use of key inhaled therapies such as mucolytics, osmotics and nebulised antibiotics are recognised as key process outcomes to indicate quality of cystic fibrosis service provision. They are so given prominence in annual cystic fibrosis registry reports. There is very specific guidance about the use of inhaled therapies in people with CF with a clinical commissioning policy and NICE technology appraisals regarding specific inhaled treatments. Despite this registry reports still demonstrate variation.	See the CF trust registry reports.

Preventing cross-infection					
ID	**Stakeholder**	**Suggested key area for quality improvement**	**Why is this important?**	**Why is this a key area for quality improvement?**	**Supporting information**
21	Association of Chartered Physiotherapists in Cystic Fibrosis	Cross Infection	Acquisition of particular bacteria can lead to deterioration in clinical status and potentially preclude patients from lung transplantation	There is variation nationally in infection control practises There is also lack of evidence for emerging pathogens	NTM interim guidelines October 2013 - CF Trust US Cystic Fibrosis Foundation and European Cystic Fibrosis Society consensus

Preventing cross-infection					
ID	**Stakeholder**	**Suggested key area for quality improvement**	**Why is this important?**	**Why is this a key area for quality improvement?**	**Supporting information**
				Patients may need psychology support in light of stringent segregation policies and in cases where transplantation is no longer an option	recommendations for the management of non-tuberculous mycobacteria in individuals with cystic fibrosis. Thorax 2016; 71:i1 - i22
22	Papworth Hospital NHS Foundation Trust	Universal and standardised infection control measures for all CF patient contacts – use of gowns and gloves for all patient groups	With the emergence of new and resistant pathogens it is imperative that we are making all possible effort to minimise cross infection and transmission between patients via every possible route, including clinicians going from patient to patient. Strict infection control practice has already been successful in reducing transmission of resistant organism such as Burkholderia and MRSA. With the emergence of organisms such as Mycobacterium abscessus and Carbepenemase-producing enterobacteriaceae the potential threat to CF patients is high.	Infection control measures in CF centres are variable for pathogens other than those that have already been identified as serious threats (NICE Infection prevention and control guideline for cystic fibrosis) With new pathogens emerging the complexity of these infection control measures is rising, and therefore adopting a universal approach for all patients may avoid unnecessary risks and exposures.	Emergence and spread of a human-transmissible multi-drug resistant nontuberculous mycobacterium Bryant et al **Science 2016** This publication demonstrates that Mycobacterium abscessus is transmissible, potentially via fomites and aerosols supporting the argument that personal protective equipment for patient contacts may help to minimise this risk.
23	SCM 2	Cross Infection	Acquisition of bacterial lung infections may lead to	The control of infection and the policies that CF centres use differ nationally.	US Cystic Fibrosis Foundation and European

Preventing cross-infection					
ID	**Stakeholder**	**Suggested key area for quality improvement**	**Why is this important?**	**Why is this a key area for quality improvement?**	**Supporting information**
			deterioration in health with long term consequence. Acquisition of certain organisms may preclude patients from Lung transplantation.	There is a lack of evidence around emerging pathogens.	Cystic Fibrosis Society consensus recommendations for the management of non-tuberculous mycobacteria in individuals with cystic fibrosis. Thorax 2016; 71:i1 - i22 open Guideline NTM
24	SCM 3	Prevention of cross-infection between people with CF	Acquisition of lung infections is the most important cause of deterioration in the cystic fibrosis condition, and certain types can spread between people with cystic fibrosis	Increasing numbers of people with cystic fibrosis grouped into specialist centres means there is a greater potential for contact and spread of infections	See NICE CF guideline which suggests strategies to prevent cross infection in clinics at: https://www.nice.org.uk/guidance/indevelopment/gid-cgwave0736
25	Cystic Fibrosis Trust	Cross infection prevention	Infection and infection-related inflammation are key drivers of morbidity and mortality in cystic fibrosis. Minimising risk is critical in clinical settings.	Transparency relating to cross infection prevention measures and outcomes will further develop best practice models and help drive quality improvement initiatives.	NHS England's Specialised Services Quality Dashboard (SS-QD) for cystic fibrosis
26	SCM 4	Organisation of CF clinics to reduce risk cross infection between patients	It is well established that people with Cystic Fibrosis have a reduced life-expectancy if they chronically colonise certain organisms.	Practice differs across the UK Organisms such as NTMs now recognised as an area of specific concern	NICE guidance CF Trust Standards of Care ECFS Guidelines BTS guidelines

Preventing cross-infection					
ID	**Stakeholder**	**Suggested key area for quality improvement**	**Why is this important?**	**Why is this a key area for quality improvement?**	**Supporting information**
27	NHSE	Cross infection prevention – clinics and in-patients. Segregation clinics. Staff hygiene.			Relevant national audits – CF Registry, and Quality dashboards.

Service delivery					
ID	**Stakeholder**	**Suggested key area for quality improvement**	**Why is this important?**	**Why is this a key area for quality improvement?**	**Supporting information**
28	Vertex Pharmaceuticals	Ensuring that patients with cystic fibrosis get access to appropriate treatment is an area that requires improvement. The Quality Standard provides an opportunity to ensure appropriate consideration of all treatment options for each person with cystic fibrosis.	There are inequalities in access to treatment for people with cystic fibrosis which need to be addressed. Taylor-Robinson et al highlighted that “In terms of use and access to treatments in the British National Health Service (NHS), more disadvantaged CF patients are more likely to receive antibiotic and nutritional treatments, after adjusting for disease severity, but less likely to receive inhaled therapies such as DNase. This inequality becomes more evident after transition to adult care”. (Taylor-Robinson D, 2015).	Reference to the forthcoming Clinical Guideline and relevant NHS England documents will be important to inform an indicator on treatment. It is important that the Quality Standard takes into account all relevant policy documents on treatment. At present, the following NHS England commissioning documents for cystic fibrosis are missing: Ivacaftor for children aged 2-5 years with cystic fibrosis (named mutations) Ivacaftor for cystic fibrosis (named mutations) Both policies recommend ivacaftor for routine commissioning. The treatment is an established part of the care pathway and should be considered as such in any Quality statements relating to treatment.	

Service delivery					
ID	**Stakeholder**	**Suggested key area for quality improvement**	**Why is this important?**	**Why is this a key area for quality improvement?**	**Supporting information**
29	Association of Chartered Physiotherapists in Cystic Fibrosis	Access to Homecare for the CF population	Home reviews by members of the MDT has significant advantages to CF patients. It reduces the burden of care for patients as they don't have to travel to the centre, it reduces the risk of cross infection, enhances quality of life and for the NHS, it reduces pressure on hospital beds.	There is large regional variation in provision of community support to CF patients. There is a lack of existing evidence in this area	NHS outcomes framework – domain 2
30	British Thoracic Society	How often patients in shared care were seen by the centre	It is important to monitor outcomes of CF patients in shared care, to ensure that these are as good as patients receiving centre care only	Monitoring number of Centre visits for shared care patients would help to monitor quality of care in this area	CF registry https://www.cysticfibrosis.org.uk/the-work-we-do/uk-cf-registry
31	British Thoracic Society	Timeliness of inpatient admission	With increasing patient numbers, providing timely access to Inpatient care can be difficult, particularly during winter period	Measure of waiting times for accessing hospital bed for acute CF admissions	CF registry- already collect numbers of IV days and whether IP or OP https://www.cysticfibrosis.org.uk/the-work-we-do/uk-cf-registry
32	SCM 1	Access to Homecare for the CF population	Home reviews by all members of the MDT has significant advantages to patients with CF. It a) reduces burden of care for patients for example by reducing travel to the CF centre with time , cost and experience implications b) reduces the risk of cross infection c) enhances quality of	There is currently significant widespread regional variation in provision of community support to patients with CF. Evidence is currently lacking in this area. To improve the patient experience and journey for patients and their families when living with a	Please see: NHS outcomes framework – domain 2 NHS Outcomes Framework 2016 to 2017 - GOV.UK

Service delivery					
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			life . It also has advantages to the acute NHS services and CF centre by reducing pressure on acute hospital beds.	chronic life limiting disease such as CF that requires such regular MDT review and care.	1. McCabe, H & Curran, R (2011) Funding Helps Lift the Barriers to Dietetics as the "Cinderella" Profession in CF Care. Abstract: Pediatric Pulmology 34th European CF Conference. 2. Lake, E. Food for thought: Patient's and Carer's views on dietetic care in Cystic Fibrosis. Cystic Fibrosis Trust (2010) 3. Barnes, R. Why home healthcare is so important for patients with Cystic Fibrosis? British J Home Healthcare (2005);1;1;14-16 4. Byrne, N. Community physiotherapy for children with Cystic Fibrosis: A family satisfaction survey. J of Cystic Fibrosis (2005); 4; 123-127
33	SCM 2	Access to Home care to include Port access/ I/V antibiotic therapy. Nebuliser care, Enteral feeding / nutritional management Pulmonary monitoring	Compliments inpatient care. Enhance Quality of Life. Reduces burden of care for patient. Allows patient and family to continue normal daily activities i.e. School, work etc.	Existing evidence is lacking that looks at clinical outcomes Quality of life. Regional variation as some centres offer this service and others do not.	NHS Outcomes Framework 2016 to 2017 - GOV.UK https://www.cysticfibrosis.org.uk/~/media/documents/the-work-we-do/care/consensus-

Service delivery					
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		assessment and management.	Reduces potential risk of cross infection.	Significant differences between provision by paediatric and adult clinics.	documents- Role of the CF Nurse Specialist
34	SCM 5	Choice around ways to access routine monitoring; i.e access to telemedicine or home visits versus outpatient visits	Both the draft NICE guideline for cystic fibrosis and the CF trust standards of care suggest routine monitoring of people with cystic fibrosis at least every 8 weeks for children and at least every 12 weeks for adults. This is a significant burden for people with cystic fibrosis and their families. As per the draft NICE guideline for cystic fibrosis, telemedicine or home visits should be considered for routine monitoring where they are more appropriate than outpatient visits and if the person with cystic fibrosis prefers it.	Some centres offer telemedicine or home visits for some reviews and/or have been part of recent trials looking at remote monitoring but access to routine monitoring away from the clinic is not established or national practice and so variation in access exists. There is currently no national record of where routine monitoring takes place either through registry reports or through the quality dashboard.	See the CF trust registry reports and dashboard which both demonstrate no recording of location of monitoring visit.
35	NHSE	Number of patients reviewed at least once (twice?) by tertiary centre for shared care patients (shared care mostly confined to paediatrics)			Relevant national audits – CF Registry, and Quality dashboards.
36	Association of Chartered Physiotherapists in Cystic Fibrosis	Transitional care	Transition is a difficult time for young people and their parents/carers. It needs to be a timely and co-ordinated process.	Transition occurs during teenage years, which is already a difficult time for young people with CF. If transition is not a smooth process, there is	Transition from childrens' to adults' services for young people using health or social care services (NG43)

Service delivery					
ID	**Stakeholder**	**Suggested key area for quality improvement**	**Why is this important?**	**Why is this a key area for quality improvement?**	**Supporting information**
			Many young people with CF struggle to maintain their health in the early stages following transition	potential for irreversible deterioration in clinical status. There are currently national variations	https://www.nice.org.uk/guidance/ng43 Ready Steady Go
37	British Thoracic Society	CF transition from paediatrics to adult care – outcome measures	All CF centres are involved in Transition from Paediatrics to adults, and there has been work in this area showing that smooth transition improves onward care	Development of outcomes measures Evidence of number and staffing at joint clinics Patient visits to new site Development of Patient satisfaction questionnaires	CF registry would be best placed to help in this area https://www.cysticfibrosis.org.uk/the-work-we-do/uk-cf-registry
38	Royal College of Nursing	Appropriate and timely transition to adult services			
39	SCM 1	Transitional care	Transition is a difficult time for young people and their parents/carers. It needs to be a timely and co-ordinated process. Many young people with CF struggle to maintain their health in the early stages following transition	Transition occurs during teenage years, which is already a difficult time for young people with CF. If transition is not a smooth process, there is potential for irreversible deterioration in clinical status. There are currently national variations	Please see: Transition from children's' to adults' services for young people using health or social care services (NG43) https://www.nice.org.uk/guidance/ng43 Ready Steady Go- National Programme http://ep.bmj.com/content/edpract/100/6/313.full.pdf

Service delivery					
ID	**Stakeholder**	**Suggested key area for quality improvement**	**Why is this important?**	**Why is this a key area for quality improvement?**	**Supporting information**
40	SCM 2	Transitional care	Effective transition from Paediatric to Adult services is a key point in patient's life. It comes at a time when there are lots of changes happening and it needs a coordinated progressive approach,	National variation in the way transition is carried out. Collaboration within the Paediatric and Adult CF services is not well defined. Completion of transition to adult care is not well defined... There needs to be a consistent approach.	Transition from children's' to adults' services for young people using health or social care services (NG43) https://www.nice.org.uk/guidance/ng43 Ready Steady Go- National Programme http://ep.bmj.com/content/edpract/100/6/313.full.pdf
41	SCM 5	Quality of the transition process	Transition occurs around adolescence and this period marks an increase in the risk of reduced clinical status in people with cystic fibrosis. It is accepted that a strong transition process is needed to support people during this challenging period of their life. The draft NICE guidance for cystic fibrosis states that people with cystic fibrosis and their family members or carers (as appropriate) should be asked for feedback on the quality of the transition service.	The NICE guideline for transition emphasised that young people should be involved in service design, delivery and evaluation. Although there are a number of single centre studies reviewing the quality of the transition process for people with cystic fibrosis, there has been no national review of the transition process and the transition process hasn't been a particular focus within peer review as peer review has focused on each CF centre rather than the bridging between them. It is likely that national variation exists and that this variation may lead to variation in outcome.	See the NICE draft guidance for cystic fibrosis and Transition from children's to adults' services for young people using health or social care services. NICE guideline No 43. London: NICE, 2016.

Service delivery					
ID	**Stakeholder**	**Suggested key area for quality improvement**	**Why is this important?**	**Why is this a key area for quality improvement?**	**Supporting information**
42	SCM 4	Children's and adult services should have a coordinated and documented pathway for transition	Transition is a particularly stressful life event for those with chronic illnesses and comes at a time when individuals are coping with multiple life events. Poor outcomes in this group (16 – 18 year olds) can be avoided with collaboration and good communication	Practice differs across the UK Need to raise profile of transition and encourage collaboration between paediatric and adult services	NICE guidance CF Trust Standards of Care
43	NHSE	Transition. Timing, methods of transition clinics. Young/adult and parent preparedness and satisfaction.			Relevant national audits – CF Registry, and Quality dashboards.
44	Royal College of Nursing	Access to specialist teams that includes a specialist nurse			
45	Royal College of Nursing	Access to appropriate therapy, i.e. physiotherapy, dietician etc.			
46	Royal College of Nursing	Access to a range of (HCP's) Health Care practitioners including school nurse			
47	SCM 3	Delivery of specialist healthcare for people with cystic fibrosis	People with cystic fibrosis have multisystem disease which requires regular monitoring and management by a team of specialists.	There is variable access to multidisciplinary specialist teams for people with cystic fibrosis	See NICE CF guideline indicating membership of CF healthcare teams and frequency of monitoring at: https://www.nice.org.uk/guid

Service delivery					
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					ance/indevelopment/gid-cgwave0736 See also UK CF Trust consensus document Standards for the Clinical Care of Children and Adults with Cystic Fibrosis in the UK. Second edition. December 2011 at: https://www.cysticfibrosis.org.uk/the-work-we-do/clinical-care/consensus-documents
48	SCM 5	Access to a full and appropriately experienced CF specialist multidisciplinary team	As per the draft NICE guideline for cystic fibrosis, care for people with cystic fibrosis should be provided by a specialist cystic fibrosis multidisciplinary team based at a specialist cystic fibrosis centre. The delivery of the complex care needed by people with CF requires clinicians with the appropriate knowledge and skills to deliver it (CF trust standards of care 2011).	Past peer review reports have demonstrated variation in access to a full specialist cystic fibrosis multidisciplinary team. It is shown from these peer review reports that where the CF team is not complete or appropriately trained/experienced, then care fully compliant with the national service specification and CF trust standards of care is not achieved.	See past CF Trust peer review reports which highlight variation in staffing and with compliance to standards of care: https://www.cysticfibrosis.org.uk/the-work-we-do/clinical-care/peer-reviews

Service delivery					
ID	**Stakeholder**	**Suggested key area for quality improvement**	**Why is this important?**	**Why is this a key area for quality improvement?**	**Supporting information**
49	SCM 4	The CF MDT should consist of specialist: Physicians Nurses Pharmacist Physiotherapists Dieticians Psychologists Social workers / welfare advisors	Evidence suggests that the make-up of the MDT is a key factor in improving clinical and quality of life outcomes for patients with CF	While the need for an expert MDT is accepted, the provision and expertise provided differs across the UK	NICE guidance CF Trust Standards of Care Specialised commissioning specifications
50	NHSE	Access to specialist CF care. Includes acute issues ie if patient goes into their local DGH. Bed availability in CF units. Access to full CF MDT in clinics and in-patients. Access to other specialists eg endocrine, gastro etc.			Relevant national audits – CF Registry, and Quality dashboards.
51	NHSE	Other possible issues – equality of access to expensive drugs by region			Relevant national audits – CF Registry, and Quality dashboards.

Additional / developmental areas					
ID	**Stakeholder**	**Suggested key area for quality improvement**	**Why is this important?**	**Why is this a key area for quality improvement?**	**Supporting information**
52	Association of Chartered Physiotherapists in Cystic Fibrosis	Additional developmental areas of emergent practice	Alterations in the role of the physiotherapist relating to CFTR channel modulators Management of the upper airway in CF patients Management of CF-SPID group Technology in airway clearance		
53	British Thoracic Society	Palliative care in Cystic fibrosis	The development of effective palliative care in CF is an emerging area, and no unified strategy has been developed so far	Joint working with CF and palliative care teams. The development of standard care pathway for joint management is a suggested area for quality improvement, and perhaps adding to quality standard	Specialised services quality dashboard: Cystic fibrosis – adults https://www.england.nhs.uk/commissioning/spec-services/npc-crg/spec-dashboards/
54	SCM 5	Monitoring adherence to treatments	Adherence to therapy, including medication and other treatments such as exercise, nutritional recommendations and airway clearance, is a known issue for people with cystic fibrosis. With median adherence being less than 50% for many aspects of treatment.	Despite multiple mentions of adherence in the draft NICE guidance for cystic fibrosis there is no specific national guidance focusing on measurement of adherence to cystic fibrosis treatments. It is known that adherence to different treatments are variable in people with CF and that different methods of measurement will yield different results.	A large multicentre trial is underway with a focus on adherence measurement and intervention; Development and evaluation of an intervention to support Adherence to treatment in adults with Cystic Fibrosis (ACtiF).

Additional / developmental areas					
ID	**Stakeholder**	**Suggested key area for quality improvement**	**Why is this important?**	**Why is this a key area for quality improvement?**	**Supporting information**
			Increasing adherence rates are likely to have significant impacts on clinical status.	There is currently no guideline about methods of monitoring adherence to cystic fibrosis treatments and no national monitoring of adherence levels takes place either through registry reports or through the quality dashboard.	
55	Cystic Fibrosis Trust	Quality of annual collection of key clinical data	The purpose of the UK CF Registry is to improve the health of people with cystic fibrosis. This is done in several ways: helping people with CF and their families understand CF, and make informed decisions; Providing data for research to find out the best ways of treating, and beating cystic fibrosis; Giving clinical teams the evidence they need to improve the quality of care; Helping commissioners provide funding to NHS CF centres that is proportionate to their patients' disease severity; Monitoring the safety and effectiveness of new treatments for cystic fibrosis.	Data quality is a key area for quality improvement as accuracy and completeness of data are vital for ensuring the robustness of other indicators, which we rely on to evaluate clinical care. Data transparency and engagement with service audit procedures may also be a proxy indicator of a well-functioning unit.	The UK CF Registry collects demographic, treatment and health outcomes data. You can find a full list of the data items collected at www.cysticfibrosis.org.uk/registry.
56	Cystic Fibrosis Trust	Lung transplantation	Many people with CF face the possibility of a lung transplant. Lung transplantation can extend and improve quality of life, but it	Available information about referral, acceptance and outcomes is limited and dispersed between local and national datasets.	Datasets available at NHSBT ODT and UK CF Registry.

Additional / developmental areas					
ID	**Stakeholder**	**Suggested key area for quality improvement**	**Why is this important?**	**Why is this a key area for quality improvement?**	**Supporting information**
			involves an extensive evaluation process and requires extensive support new lungs healthy.	Improved understanding of timing of referral, with relation to clinical indicators, evaluation process, follow-up and outcomes can help understand any variability and drive quality improvement.	
57	Cystic Fibrosis Trust	Patient experience	Good quality care is best judged by the impact it has on the lived experience of patients.	Capturing indicators of this experience that can meaningfully influence care delivery is critically important to drive up standards.	No nationally standardised process. However, CFT's Peer Review process has collected patient experience data locally since 2007. It is envisaged that – as part of NHS England's QST function – patient experience indicators will be built into the QST's QSIS.
58	NHSE	[Comments from 2 respondents] Management of CFSPID patients (diagnosed after newborn screening). In CF clinic or other specialist clinic? Length of follow up etc. Should they go on Registry? Management of CFSPID patients would be for paediatric centres not adult centres, adult centres could have referrals for lung transplant for patients with			Relevant national audits – CF Registry, and Quality dashboards.

Additional / developmental areas					
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		end stage lung disease, and/or screening and management of CF diabetes eg number screened annually, HbA1C levels.			
59	SCM 1	Pancreatic enzyme replacement therapy – what are patients really taking? What is the treatment burden? How can treatment be improved and how can this inform new PERT therapies?			
60	SCM 1	Impact of living with the dietary implications required for optimal health and outcomes in Cystic Fibrosis against a backdrop of healthy eating and government agendas that are aimed at tackling an obesity epidemic within an obesogenic environment.			
61	Public Health England / UK National Screening Committee	Timely entry into clinical care of all cystic fibrosis screen positive babies to ensure that health benefits are achieved by reducing morbidity/mortality	The national UK standards state; CF (2 mutations detected) to attend first clinical appointment by 28 days of age Acceptable: ≥ 95.0% Achievable: 100%	The annual data and performance analysis report shows the standard for timely entry into clinical care for cystic fibrosis is not met for all babies. https://www.gov.uk/government/uploads/system/uploads/attachment_data/file/638857/Data_Coll	Consensus exercises have repeatedly identified timeliness as a key metric for the performance of a newborn blood spot screening programme.

Additional / developmental areas					
ID	**Stakeholder**	**Suggested key area for quality improvement**	**Why is this important?**	**Why is this a key area for quality improvement?**	**Supporting information**
			CF (1 or no mutation detected) to attend first clinical appointment by 35 days of age Acceptable: ≥ 80.0% Achievable: 100%	ection and Performance Analysis Report 2015-16.pdf	Guidelines and the ECFS standards state that screened infants have nutritional and respiratory advantages. Regional differences in processing a CF NBS sample mean that this is a key parameter in assessing performance, as it reflects a number of distinct phases in the process. A deficit in any of these components will adversely impact on timeliness, which will have a cumulative impact of the well-being of these infants. (ref Smyth et al. 2014 pubmed ID 24856775)
62	Royal College of Nursing	Timely assessment referral for diagnosis			