Cystic fibrosis

NICE quality standard

Draft for consultation

December 2017

This quality standard covers diagnosing and managing cystic fibrosis in infants, children, young people and adults. It describes high-quality care in priority areas for improvement.

It is for commissioners, service providers, health practitioners and the public.

This is the draft quality standard for consultation (from 15 December 2017 to 19 January 2018). The final quality standard is expected to publish in May 2018.

Quality statements

Statement 1 People with cystic fibrosis have a comprehensive annual review.

Statement 2 People with cystic fibrosis have individual rooms with en-suite facilities when admitted to hospital as inpatients.

Statement 3 People with cystic fibrosis who have chronic *Pseudomonas aeruginosa* infection have sustained treatment with an inhaled antibiotic.

Statement 4 People with cystic fibrosis who have clinical evidence of lung disease are prescribed rhDNase[1] as the first choice of mucoactive agent.

NICE has developed guidance and a quality standard on patient experience in adult NHS services (see the NICE pathway on patient experience in adult NHS services), which should be considered alongside these quality statements.

Other quality standards that should be considered when commissioning or providing cystic fibrosis services include:

- End of life care for infants, children and young people (2017) NICE quality standard 160
- Transition from children's to adults' services (2016) NICE quality standard 140
- Antimicrobial stewardship (2016) NICE quality standard 121
- Medicines optimisation (2016) NICE quality standard 120
- Healthcare-associated infections (2016) NICE quality standard 113
- Infection prevention and control (2014) NICE quality standard 61
- End of life care for adults (2011) NICE quality standard 13

A full list of NICE quality standards is available from the quality standards topic library.

[1] At the time of consultation (December 2017), rhDNase did not have a UK marketing authorisation for use in children with cystic fibrosis. The prescriber should follow relevant professional guidance, taking full responsibility for the decision. Informed consent should be obtained and documented. See the General Medical Council's Prescribing guidance: prescribing unlicensed medicines for further information.

Questions for consultation

Questions about the quality standard

Question 1 Does this draft quality standard accurately reflect the key areas for quality improvement?

Question 2 Are local systems and structures in place to collect data for the proposed quality measures? If not, how feasible would it be for these to be put in place?

Question 3 Do you think each of the statements in this draft quality standard would be achievable by local services given the net resources needed to deliver them? Please describe any resource requirements that you think would be necessary for any statement. Please describe any potential cost savings or opportunities for disinvestment.

Questions about the individual quality statements

Question 4 For draft quality statement 1: Do all people with cystic fibrosis currently have annual reviews? If so, do the annual reviews include all the components listed in the definition of statement 1?

Question 5 For draft quality statement 3: Although the proportion of people with cystic fibrosis and chronic *Pseudomonas aeruginosa* who are taking inhaled antibiotics is high nationally, it varies across specialist centres and network clinics. Is this variation caused by inhaled antibiotics not being offered, or offered but not taken, or some other reason?

Local practice case studies

Question 6 Do you have an example from practice of implementing the NICE guideline that underpins this quality standard? If so, please submit your example to NICE local practice case studies on the NICE website. Examples of using NICE quality standards can also be submitted.

Quality statement 1: Annual reviews

Quality statement

People with cystic fibrosis have a comprehensive annual review.

Rationale

Cystic fibrosis is a multisystem genetic disorder that needs regular monitoring and a range of assessments for effective management. Assessments are undertaken by different specialists and can take place at different times to reduce the burden on the person with cystic fibrosis. An annual review brings together the results of all the assessments. It enables the multidisciplinary team to understand the progression of the person's disease and make changes to their care to prevent or limit the symptoms and complications of cystic fibrosis.

Quality measures

Structure

a) Evidence that cystic fibrosis multidisciplinary teams have professionals with specialist expertise in the condition including a paediatrician or adult physician, nurse, physiotherapist, dietitian, pharmacist and a clinical psychologist.

Data source: Local data collection, for example service specifications. NHS England service specifications for cystic fibrosis cover multidisciplinary team composition and require providers to demonstrate they are meeting requirements.

b) Evidence of local systems to identify and invite people with cystic fibrosis to have comprehensive annual reviews.

Data source: Local data collection, for example service specifications. NHS England service specifications for cystic fibrosis cover annual reviews and require providers to demonstrate they are meeting requirements.

Process

Proportion of people with cystic fibrosis who have a comprehensive annual review.

Numerator – the number in the denominator who have had a comprehensive annual review in the last year.

Denominator – the number of people with cystic fibrosis.

Data source: Local data collection, for example local audit of patient records.

Outcome

a) Lung function (forced expiratory volume in 1 second [FEV_1]) of people with cystic fibrosis.

Data source: Local data collection, for example local audit of patient records. Appendix 2 in the UK Cystic Fibrosis Registry annual data report includes FEV_1 converted from absolute litres of air blown out into a predicted percentage. The predicted percentage is based on the FEV_1 expected for a person without cystic fibrosis of the same age, gender, height and ethnicity. NHS England's specialised services quality dashboard for cystic fibrosis reports the median FEV_1 as a predicted percentage for cystic fibrosis clinics.

b) Health-related quality of life scores of people with cystic fibrosis.

Data source: Local data collection, for example a survey of people with cystic fibrosis using a cystic fibrosis quality of life questionnaire.

c) Median BMI percentiles in children and young people with cystic fibrosis.

Data source: Local data collection, for example, local audit of patient records. The UK Cystic Fibrosis Registry annual data report presents median BMI percentiles for children and young people with cystic fibrosis (aged 2 to 19 years) in the UK. The percentiles are compared to a BMI percentile for a healthy person of the same age; the 50th percentile or the BMI percentile that half of the UK population people of that age has achieved.

d) Median BMI in adults with cystic fibrosis.

Data source: Local data collection, for example, local audit of patient records. The UK Cystic Fibrosis Registry annual data report includes median BMI for adults with

cystic fibrosis (aged 20 years and over) in the UK. The median BMI is compared to a target BMI of 23 for males and 22 for females.

What the quality statement means for different audiences

Service providers (such as cystic fibrosis centres) ensure that systems are in place to identify people with cystic fibrosis for annual review, that annual reviews are comprehensive and carried out by specialist multidisciplinary teams, and that the specialist multidisciplinary teams have access to equipment, testing and diagnostic services needed for assessments.

Healthcare professionals (members of cystic fibrosis multidisciplinary teams) meet annually to review assessment results and treatment for all people with cystic fibrosis. Team members undertake comprehensive assessments specific to their profession before the annual review. This may be on the same day as the review, or at an earlier time to reduce the burden on the person. The results of the assessments and outcomes are shared with the person.

Commissioners (NHS England) ensure that service specifications require comprehensive annual reviews to be carried out by cystic fibrosis multidisciplinary teams for all people with cystic fibrosis.

People with cystic fibrosis have the results of all the medical checks and tests they've had during the past year reviewed by a team of specialists called a cystic fibrosis multidisciplinary team. This review is done every year and means that the team of specialists can see how well the person's treatment is working and whether any changes are needed.

Source guidance

Cystic fibrosis: diagnosis and management (2017) NICE guideline NG78, recommendation 1.5.2

Definitions of terms used in this quality statement

Comprehensive annual review

An annual review carried out by a specialist cystic fibrosis multidisciplinary team that includes the following:

- pulmonary assessment
- an assessment of nutrition and intestinal absorption
- an assessment for liver disease
- testing for cystic-fibrosis-related diabetes, from 10 years of age
- an assessment for other potential or existing cystic fibrosis complications
- a psychological assessment
- assessments by a specialist nurse, physiotherapist, pharmacist and social worker
- a review of a the person's exercise programme.

The components of the annual review above do not all have to take place on the same day.

[NICE's guideline on cystic fibrosis: diagnosis and management, recommendation 1.5.2, full guideline and expert consensus]

Equality and diversity considerations

People living in isolated areas may find it harder to travel to specialist cystic fibrosis centres than people living in cities. Other models of care should be considered for such groups. Shared-care models with a local paediatric team can be used for children and young people, and outreach care for adults can be delivered by specialists at a local hospital. Routine reviews can also take place during home visits and using telemedicine in some circumstances.

Question for consultation

Do all people with cystic fibrosis currently have annual reviews? If so, do the annual reviews include all the components listed in the definition of statement 1?

Quality statement 2: Preventing cross-infection

Quality statement

People with cystic fibrosis have individual rooms with en-suite facilities when admitted to hospital as inpatients.

Rationale

People with cystic fibrosis are vulnerable to cross-infection. Bacteria that are usually harmless to people who don't have cystic fibrosis can be harmful to those who do. Infection can be passed from person to person through social contact, actions such as coughing, and sharing rooms, equipment, food or drink. The risk of cross-infection increases when people are in close proximity to one another for long periods of time, such as in hospital wards. Treating people with cystic fibrosis in individual rooms with en-suite facilities when they are admitted to hospital reduces the risk of cross-infection.

Quality measures

Structure

a) Evidence of local infection control strategies that cover inpatient settings for people with cystic fibrosis.

Data source: Local data collection, for example infection control policies or admission protocols. NHS England service specifications for cystic fibrosis state that services must have policies and procedures in place to protect patients from the risk of cross-infection.

b) Evidence of inpatient wards containing individual rooms with en-suite facilities.

Data source: Local data collection, for example service specifications or ward layout plans. NHS England service specifications for cystic fibrosis states that every person with cystic fibrosis admitted as an inpatient will be in their own room with en-suite facilities.

Process

Proportion of inpatient admissions for people with cystic fibrosis where they have individual rooms with en-suite facilities.

Numerator – the number in the denominator where admission was to an individual room with en-suite facilities.

Denominator – the number of inpatient admissions for people with cystic fibrosis.

Data source: Local data collection, for example local audit of patient records. NHS England's specialised services quality dashboard for cystic fibrosis reports the percentage of patients admitted who are admitted to a single room or cubicle.

Outcome

a) Incidence of cross infection in people with cystic fibrosis admitted as inpatients.

Data source: Local data collection, for example local audit of patient records.

b) Health-related quality of life scores of people with cystic fibrosis.

Data source: Local data collection, for example a survey of people with cystic fibrosis using a cystic fibrosis quality of life questionnaire.

What the quality statement means for different audiences

Service providers (such as cystic fibrosis centres) ensure that all cystic fibrosis inpatient wards consist of single rooms with en-suite facilities, that there are sufficient rooms to manage planned and emergency admissions, and that systems are in place to make sure that people with cystic fibrosis treated outside of the cystic fibrosis ward are allocated to individual rooms with en-suite facilities.

Healthcare professionals (such as bed managers and members of cystic fibrosis multidisciplinary teams) make sure that admission arrangements for people with cystic fibrosis who need an overnight stay include individual rooms with en-suite facilities. Members of the cystic fibrosis multidisciplinary team discuss the risk of cross-infection and the need for separate rooms with the person admitted. They work with bed managers to arrange planned admissions to help prevent people with cystic

fibrosis coming into contact with each other, such as when they use diagnostic facilities or communal areas.

Commissioners (NHS England) ensure that service specifications require people with cystic fibrosis to be treated in single rooms with en-suite facilities when admitted as inpatients; and require providers to have policy and procedures in place to protect people with cystic fibrosis from the risk of cross-infection.

People with cystic fibrosis have single rooms with en-suite facilities when they stay overnight in hospital. This makes it less likely that they will pick up an infection from another person.

Source guidance

Cystic fibrosis: diagnosis and management (2017) NICE guideline NG78, recommendation 1.8.7

Quality statement 3: Treating chronic lung infection

Quality statement

People with cystic fibrosis who have chronic *Pseudomonas aeruginosa* infection have sustained treatment with an inhaled antibiotic.

Rationale

Lung infection is the cause of much of the morbidity and mortality associated with cystic fibrosis. *Pseudomonas aeruginosa* is the most frequent cause of lung infection in people with cystic fibrosis. Chronic infection with *Pseudomonas aeruginosa* leads to worsening signs and symptoms and reduced lung function. Long-term treatment with an inhaled antibiotic suppresses *Pseudomonas aeruginosa* infection and helps to maintain lung function and quality of life.

Quality measures

Structure

a) Evidence of local arrangements to identify people with cystic fibrosis who have *Pseudomonas aeruginosa* infection.

Data source: Local data collection, for example protocols for microbiological surveillance of respiratory secretions.

b) Evidence of the availability of devices for people with cystic fibrosis to take inhaled antibiotics.

Data source: Local data collection, for example from service protocols. NHS England service specifications for cystic fibrosis require a comprehensive nebuliser service to provide devices that deliver drugs in a fast and efficient manner.

Process

a) Proportion of people with cystic fibrosis who have chronic *Pseudomonas aeruginosa* infection prescribed an inhaled antibiotic for sustained treatment.

Numerator – the number in the denominator who are prescribed an inhaled antibiotic for sustained treatment.

Denominator – the number of people with cystic fibrosis who have chronic *Pseudomonas aeruginosa* infection.

Data source: Local data collection, for example local audit of patient records. NHS England's specialised services quality dashboard for cystic fibrosis reports the percentage of people with chronic *Pseudomonas aeruginosa* infection receiving inhaled antibiotics (indicators CFS12-A and CFS12-P).

b) Proportion of people with cystic fibrosis who have chronic *Pseudomonas aeruginosa* infection taking an inhaled antibiotic for sustained treatment.

Numerator – the number in the denominator who are taking an inhaled antibiotic for sustained treatment.

Denominator – the number of people with cystic fibrosis who have chronic *Pseudomonas aeruginosa* infection.

Data source: Local data collection, for example local audit of patient records. The UK Cystic Fibrosis Registry annual data report includes inhaled antibiotic use among people with chronic *Pseudomonas aeruginosa* infection. NHS England's specialised services quality dashboard for cystic fibrosis reports the percentage of people with chronic *Pseudomonas aeruginosa* infection receiving inhaled antibiotics (indicators CFS12-A and CFS12-P).

Outcome

a) Lung function (forced expiratory volume in 1 second [FEV_1]) of people with cystic fibrosis.

Data source: Local data collection, for example local audit of patient records. Appendix 2 of the UK Cystic Fibrosis Registry annual data report includes FEV_1 converted from absolute litres of air blown out into a predicted percentage The predicted percentage is based on the FEV_1 expected for a person without cystic fibrosis of the same age, gender, height and ethnicity.

b) Number of people with cystic fibrosis who experience a pulmonary exacerbation.

Data source: Local data collection, for example local audit of patient records.

What the quality statement means for different audiences

Service providers (such as cystic fibrosis centres) have access to microbiology services and systems to identify people with cystic fibrosis who have chronic *Pseudomonas aeruginosa* infection. They have protocols to prescribe inhaled antibiotics for sustained use to people with cystic fibrosis when eradication treatment has not worked; and to monitor the effects of the treatment.

Healthcare professionals (such as cystic fibrosis specialist paediatricians and adult physicians) make sure that people with cystic fibrosis have frequent microbiological surveillance of respiratory secretions and that eradication is attempted for first and subsequent new *Pseudomonas aeruginosa* infections. They prescribe inhaled antibiotics for sustained use when eradication treatment has not worked, and discuss adherence and the benefits and harms of the treatment with the person.

Commissioners (NHS England) ensure that service specifications require sustained treatment with an inhaled antibiotic people for people with cystic fibrosis who have chronic *Pseudomonas aeruginosa* infection when eradication treatment has not worked.

People with cystic fibrosis who have a long-term lung infection called *Pseudomonas aeruginosa* take an antibiotic medicine that they inhale (breathe in) to benefit their lungs. The medicine will not get rid of the infection, but it will help to keep it under control.

Source guidance

Cystic fibrosis: diagnosis and management (2017) NICE guideline NG78, recommendation 1.6.34

Definitions of terms used in this quality statement

Pseudomonas aeruginosa

A bacterial infection which affects the lungs. [NICE's guideline on cystic fibrosis: diagnosis and management, glossary in full guideline]

Sustained treatment

Long-term treatment intended to supress and control an infection after attempts to eradicate it have not worked. [Expert opinion]

Question for consultation

Although the proportion of people with cystic fibrosis and chronic *Pseudomonas aeruginosa* who are taking inhaled antibiotics is high nationally, it varies across specialist centres and network clinics. Is this variation caused by inhaled antibiotics not being offered, or offered but not taken, or some other reason?

Quality statement 4: Choice of mucoactive agent

Quality statement

People with cystic fibrosis who have clinical evidence of lung disease are prescribed rhDNase[1] as the first choice of mucoactive agent.

Rationale

Sticky mucus accumulates in the lungs in people with cystic fibrosis, making them more prone to infection. Repeated infection can cause permanent damage to the lungs. Airway clearance techniques are used to loosen and remove excess sticky mucus. The mucoactive agent rhDNase thins the mucus, making it easier to clear from the lungs. This helps to maintain lung function and prevent infection.

Quality measures

Structure

a) Evidence of local arrangements to provide radiological imaging and lung function testing to identify people with cystic fibrosis who have lung disease.

Data source: Local data collection, for example service specifications. NHS England service specifications for cystic fibrosis state that cystic fibrosis service will have access to specialist radiology services and specialist lung function laboratories.

b) Evidence of the availability of devices for people with cystic fibrosis to take inhaled therapies.

Data source: Local data collection, for example from local protocols. NHS England service specifications for cystic fibrosis require a comprehensive nebuliser service to provide devices that deliver drugs in a fast and efficient manner.

Process

Proportion of people with cystic fibrosis who have clinical evidence of lung disease prescribed rhDNase as the first choice of mucoactive agent.

Numerator – the number in the denominator prescribed rhDNase as the first choice of mucoactive agent.

Denominator – the number of people with cystic fibrosis who have clinical evidence of lung disease.

Data source: Local data collection, for example, local audit of patient records. The UK Cystic Fibrosis Registry annual data report presents the proportion of patients with cystic fibrosis on DNase.

Outcome

a) Lung function (forced expiratory volume in 1 second [FEV_1]) of people with cystic fibrosis.

Data source: Local data collection, for example local audit of patient records. Appendix 2 in the UK Cystic Fibrosis Registry annual data report includes FEV_1 converted from absolute litres of air blown out into a predicted percentage. The predicted percentage is based on the FEV_1 expected for a person without cystic fibrosis of the same age, gender, height and ethnicity.

b) Number of people with cystic fibrosis who experience a pulmonary exacerbation.

Data source: Local data collection, for example local audit of patient records.

What the quality statement means for different audiences

Service providers (such as cystic fibrosis centres) ensure that specialist cystic fibrosis multidisciplinary teams have access to radiological services and lung function testing to identify people with cystic fibrosis who have lung disease. They have systems in place, such as prescribing protocols or prescribing decision support systems, to offer rhDNase as the first choice mucoactive agent and to monitor the effects of the treatment.

Healthcare professionals (such as cystic fibrosis specialist paediatricians or adult physicians) use radiological imaging or lung function testing to identify people with cystic fibrosis who have lung disease. They offer rhDNase as the first-choice mucoactive agent and identify an appropriate delivery device. They discuss the treatment and the importance of adherence with the person. They assess the effects of, and tolerance to, the treatment once it has started.

Commissioners (NHS England) ensure that service specifications require rhDNase to be prescribed as the first choice mucoactive agent to people with cystic fibrosis who have clinical evidence of lung disease.

People with cystic fibrosis who have lung disease take a medicine called rhDNase, which they inhale (breathe in). The medicine makes mucus in the lungs less thick and sticky, so it is easier to cough out. This helps with breathing and makes lung infections less likely.

Source guidance

Cystic fibrosis: diagnosis and management (2017) NICE guideline NG78, recommendations 1.6.17 and 1.6.18

Definitions of terms used in this quality statement

Clinical evidence of lung disease

Evidence of lung disease based on radiological imaging or lung function testing. [NICE's guideline on cystic fibrosis: diagnosis and management, full guideline]

rhDNase

Recombinant human deoxyribonuclease; dornase alfa[1]. A mucoactive agent. [NICE's guideline on cystic fibrosis: diagnosis and management, full guideline]

Mucoactive agent

A drug that affects the viscosity of mucus, usually given to make the removal of mucus through coughing easier. [NICE's guideline on cystic fibrosis: diagnosis and management, glossary in full guideline]

About this quality standard

NICE quality standards describe high-priority areas for quality improvement in a defined care or service area. Each standard consists of a prioritised set of specific, concise and measurable statements. NICE quality standards draw on existing NICE or NICE-accredited guidance that provides an underpinning, comprehensive set of recommendations, and are designed to support the measurement of improvement.

Expected levels of achievement for quality measures are not specified. Quality standards are intended to drive up the quality of care, and so achievement levels of 100% should be aspired to (or 0% if the quality statement states that something should not be done). However, this may not always be appropriate in practice. Taking account of safety, shared decision-making, choice and professional judgement, desired levels of achievement should be defined locally.

Information about how NICE quality standards are developed is available from the NICE website.

See quality standard advisory committees on the website for details of standing committee 2 members who advised on this quality standard. Information about the topic experts invited to join the standing members is available on the quality standard's webpage.

This quality standard has been included in the NICE Pathway on cystic fibrosis, which brings together everything we have said on cystic fibrosis in an interactive flowchart.

NICE has produced a quality standard service improvement template to help providers make an initial assessment of their service compared with a selection of quality statements. This tool is updated monthly to include new quality standards.

NICE produces guidance, standards and information on commissioning and providing high-quality healthcare, social care, and public health services. We have agreements to provide certain NICE services to Wales, Scotland and Northern Ireland. Decisions on how NICE guidance and other products apply in those countries are made by ministers in the Welsh government, Scottish government, and

Northern Ireland Executive. NICE guidance or other products may include references to organisations or people responsible for commissioning or providing care that may be relevant only to England.

Improving outcomes

This quality standard is expected to contribute to improvements in the following outcomes:

- lung function
- incidence and prevalence of infection
- health-related quality of life
- survival rates.

It is also expected to support delivery of the Department of Health's outcome frameworks:

- Adult social care outcomes framework 2016–17
- NHS outcomes framework 2016–17
- Public health outcomes framework for England, 2016–19.

Resource impact

NICE quality standards should be achievable by local services. The potential resource impact is considered by the quality standards advisory committee, drawing on resource impact work for the source guidance. Organisations are encouraged to use the resource impact statement for the NICE guideline on cystic fibrosis to help estimate local costs.

Diversity, equality and language

During the development of this quality standard, equality issues were considered and equality assessments are available. Any specific issues identified during development of the quality statements are highlighted in each statement.

Commissioners and providers should aim to achieve the quality standard in their local context, in light of their duties to have due regard to the need to eliminate unlawful discrimination, advance equality of opportunity and foster good relations.

Nothing in this quality standard should be interpreted in a way that would be inconsistent with compliance with those duties.

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