NICE National Institute for Health and Care Excellence

NICE quality standard

Cystic fibrosis

Quality standard
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Contents

This standard is based on NG78.

This standard should be read in conjunction with QS13, QS61, QS113, QS120, QS121, QS140 and QS160.

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Quality statement 1: Annual reviews

Quality statement

People with cystic fibrosis have the results of all assessments they have had during the past year reviewed annually by a specialist multidisciplinary team.

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 child, young person or adult with cystic fibrosis and their family or carer. 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 the results of all assessments they have had during the past year reviewed by a specialist multidisciplinary team.

Numerator – the number in the denominator who have had the results of all assessments they have had during the past year reviewed by a specialist multidisciplinary team.

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.

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 with 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, and that annual reviews look at the results of all assessments the person has had during the past year and are carried out by specialist multidisciplinary teams. They ensure 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. They review disease progression and adjust care as needed to prevent or limit symptoms and complications. Team members undertake comprehensive assessments specific to their profession before the annual review. These assessments may take place on the same day as the review, or at an earlier date to reduce the burden on the person. The results of the assessments are shared with the child, young person or adult with cystic fibrosis and their family or carer.

Commissioners ensure that service specifications require the results of all assessments undertaken during the past year to be reviewed annually 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. After the review is done, the results and any changes to treatment or care are discussed with the person.

Source guidance

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

recommendation 1.5.2

Definitions of terms used in this quality statement

Assessments carried out during the past year

The comprehensive annual review carried out by a specialist cystic fibrosis multidisciplinary team includes a review of the following assessments:

- 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.

These assessments, which are components of the comprehensive annual review, 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.

Quality statement 2: Preventing cross-infection during hospital admissions

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 coughing and through social contact including play, sharing rooms, equipment, food or drink. The risk of cross-infection increases when people with cystic fibrosis 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 admission was to an individual room 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 and hospitals) ensure that all cystic fibrosis inpatient wards consist of individual rooms with en-suite facilities and that there are sufficient rooms to manage planned and emergency admissions. 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 child, young person or adult with cystic fibrosis and their family or carer. 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 ensure that service specifications require people with cystic fibrosis to be treated in single rooms with en-suite facilities when admitted as inpatients. They require providers to have a 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 in the ward.

Source guidance

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

Proportion of people with cystic fibrosis who have chronic Pseudomonas aeruginosa infection prescribed an inhaled antibiotic.

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

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.

Outcome

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.

What the quality statement means for different audiences

Service providers (such as cystic fibrosis centres and hospitals) 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 if eradication treatment has not worked, and discuss adherence and the benefits and harms of the treatment with the child, young person or adult with cystic

fibrosis and their family or carer.

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

People with cystic fibrosiswho 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. NICE guideline NG78 (2017), recommendation 1.6.35

Definitions of terms used in this quality statement

Chronic Pseudomonas aeruginosa

Pseudomonas aeruginosa is a bacterial infection that affects the lungs. It is chronic if there have been 3 or more isolates in the preceding 12 months.

[NICE's guideline on cystic fibrosis: diagnosis and management, glossary in full guideline and expert opinion]

Sustained treatment

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

[Expert opinion]

Quality statement 4: Choice of mucoactive agent

Quality statement

People with cystic fibrosis who have clinical evidence of lung disease are prescribed rhDNase 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.

At the time of publication (May 2018), rhDNase did not have a UK marketing authorisation for use in children under 5 years with cystic fibrosis for this indication. 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 unlicensed medicines for further information.

Quality measures

Structure

Evidence of the availability of devices for people with cystic fibrosis to take inhaled rhDNase.

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 and clinical evidence of lung disease who are 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 and 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

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.

What the quality statement means for different audiences

Service providers (such as cystic fibrosis centres) ensure that specialist cystic fibrosis multidisciplinary teams 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) offer rhDNase as the first-choice mucoactive agent to people with cystic fibrosis who have lung disease and identify an appropriate delivery device. They discuss the treatment and the importance of adherence with the child, young person or adult with

cystic fibrosis and their family or carer. They assess the effects of, and tolerance to, the treatment once it has started.

Commissioners 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 fibrosiswho have lung disease have a medicine called rhDNase, or dornase alfa, as their first treatment to help with breathing. The medicine is inhaled (breathed in). It makes mucus in the lungs less thick and sticky, so it is easier to cough out. This medicine also makes lung infections less likely.

Source guidance

Cystic fibrosis: diagnosis and management. NICE guideline NG78 (2017), 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. 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]

Update information

Minor changes since publication

December 2024: Source guidance references have been updated to align this quality standard with the updated NICE guideline on cystic fibrosis.

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 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.

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 for people with cystic fibrosis:

- 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 and Social Care outcome frameworks:

- Adult social care outcomes framework 2018 to 2019
- NHS outcomes framework 2016 to 2017
- Public health outcomes framework for England, 2016 to 2019.

Diversity, equality and language

Equality issues were considered during development and equality assessments for this quality standard 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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Endorsing organisation

This quality standard has been endorsed by NHS England, as required by the Health and

Social Care Act (2012)

Supporting organisations

Many organisations share NICE's commitment to quality improvement using evidence-based guidance. The following supporting organisations have recognised the benefit of the quality standard in improving care for patients, carers, service users and members of the public. They have agreed to work with NICE to ensure that those commissioning or providing services are made aware of and encouraged to use the quality standard.

- Royal College of Nursing (RCN)
- Association of Chartered Physiotherapists in Cystic Fibrosis
- British Thoracic Society (BTS)
- Royal College of General Practitioners (RCGP)
- British Society for Paediatric Endocrinology and Diabetes (BSPED)
- National Paediatric Respiratory and Allergy Nurses Group
- Association of Paediatric Chartered Physiotherapists