NATIONAL INSTITUTE FOR HEALTH AND CARE EXCELLENCE

HEALTH AND SOCIAL CARE DIRECTORATE

QUALITY STANDARD CONSULTATION

SUMMARY REPORT

1 Quality standard title

Cystic fibrosis

Date of quality standards advisory committee post-consultation meeting:
13 February 2018

2 Introduction

The draft quality standard for cystic fibrosis was made available on the NICE website for a 5-week public consultation period between 15 December 2017 and 19 January 2018. Registered stakeholders were notified by email and invited to submit consultation comments on the draft quality standard. General feedback on the quality standard and comments on individual quality statements were accepted.

Comments were received from 15 organisations, which included national organisations, professional bodies and others.

This report provides the quality standards advisory committee with a high-level summary of the consultation comments, prepared by the NICE quality standards team. It provides a basis for discussion by the committee as part of the final meeting where the committee will consider consultation comments. Where appropriate the quality standard will be refined with input from the committee.

Consultation comments that may result in changes to the quality standard have been highlighted within this report. Comments suggesting changes that are outside of the process have not been included in this summary. The types of comments typically

not included are those relating to source guidance recommendations and suggestions for non-accredited source guidance, requests to broaden statements out of scope, requests to include thresholds, targets, large volumes of supporting information, general comments on the role and purpose of quality standards and requests to change NICE templates. However, the committee should read this summary alongside the full set of consultation comments, which are provided in appendix 1.

3 Questions for consultation

Stakeholders were invited to respond to the following general questions:

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

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

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.

Stakeholders were also invited to respond to the following statement specific questions:

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?

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?

4 General comments

The following is a summary of general (non-statement-specific) comments on the quality standard.

- Quality standard is welcomed and reflects that people with cystic fibrosis (CF) are cared for in secondary care
- It reflects key areas for quality improvement but misses out transition
- Many statements appear to be adult orientated
- No quality measures of the early management after diagnosis through new-born screening or a network model of care in the paediatric age group
- A comment on adherence to treatment should also be included

Consultation comments on data collection (question 2)

- An essential member of the MDT is the CF coordinator (band 3 or 4) to gather the data, which could have some resource impact
- Systems are in place to collect data at a local level
- All CF specialist centres enter data into the UK CF registry which collects outcomes data for the proposed quality measures
- Using UK CF registry data will be efficient and effective. This will ensure the data is robust, and avoid duplication and unnecessary resource outlay
- The UK CF registry is undertaking a quality of life study and this data could form the quality of life outcomes
- It would be useful to include the SNOMED (a structured clinical vocabulary for use in an electronic health record) clinical terms so that NHS secondary care trusts can use a standard method for coding the data.

Consultation comments on resource impact (question 3)

- The quality standard is achievable
- Statements 1, 3 and 4 are achievable by local services given net resources

- Statements 1 and 2 cannot be achieved within net resources
- Statement 2 is part of the NHS England service specifications. Some providers do not achieve this and there may be a resource impact requirement for them

5 Summary of consultation feedback by draft statement

5.1 Draft statement 1

People with cystic fibrosis have a comprehensive annual review.

Consultation comments

Stakeholders made the following comments in relation to draft statement 1:

- This is achievable and is probably being achieved by the majority of CF centres
- This is measurable but not currently achieved
- The results of the annual review need to be fed back to patients
- Annual review should have input from the full CF MDT whether it is virtual, at a shared care clinic, or at the CF specialist centre
- Rationale and audience descriptors should incorporate shared decision-making
- Note that some people refuse to attend an annual review
- Lung function, health quality of life, and BMI should not be statement outcomes as they relate to general health, not the annual review
- Equality and diversity considerations should note that the annual review must be done either in a specialist CF centre or by the full specialist CF team seeing the person in a network centre
- The MDT should include a specialist social worker, specialist psychologist and professionals with specialist expertise in managing diabetes and liver disease
- Comprehensive information on annual review is available through the UK CF registry including data on people who had an annual review in person, virtually, did not attend or had no annual review
- Measuring medication review and adherence to therapies would be helpful
- The forced expiratory volume in 1 second [FEV_1] can be the one done on the day or the best that year; both are on the UK CF registry
- The best recorded FEV_1 from throughout the year should be used, not the measure from annual review

- Health related quality of life scores are not done routinely, are of limited value and, in children's services, are difficult to standardise
- Outcome on BMIs should be a snapshot or the maximum
- Further resources may be needed to achieve this statement
- Resources can vary. Funding is allocated / divided between specialist teams and local services which require collaborative working

Consultation question 4

Stakeholders made the following comments in relation to consultation question 4:

- All centres complete annual review and input into the UK CF registry
- MDT data gathering needs to be robust especially if all clinicians do not see the patient on the same day
- Most centres already have a system in place to collect and record data
- All people with CF have an annual review that includes all the components in the definition
- The annual review also includes: a review of medicines and compliance (usually by CF pharmacist) and feedback to the person which is essential
- People declining appointments needs to be recorded
- If someone did not come for annual review their data would be added to the registry so they are banded for the PBR tariff. This means not everyone entered had an annual review

5.2 Draft statement 2

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

Consultation comments

Stakeholders made the following comments in relation to draft statement 2:

- Important area for quality improvement as minimising risk is critical in clinical settings
- This does not consider the potential harms: distress being isolated from others, people being moved to more distant hospitals if facilities are not available locally, and delays in admission and urgent clinic appointments
- People should be offered isolation, not forced into it
- In CF units which do not have access to single rooms and en-suite facilities people may be designated a bathroom to achieve as good a level of infection control as possible
- CF centres need to adhere to local infection control policies
- It should be clearer that the need for single rooms is to prevent cross infection
- It can be more complex if people with CF and nontuberculous mycobacteria (NTM) or B Cepacia are admitted
- NHS England CF quality dashboard and service specifications state that people should not be admitted to wards where there is no CF expertise
- Data is easily collected as part of the NHS England CF quality dashboard
- Unclear how the incidence of cross infection in people with cystic fibrosis admitted as inpatients will be measured
- Data on incidence of cross infection is hard to collect. There are resource implications as it involves molecular typing of bacteria being carried out on anyone who has been an inpatient and has a new organism
- Statement will have a significant resource impact on some providers but it is part of the NHS England CF service specifications
- This should be aspired to but is unachievable for some centres in the current financial climate. Significant financial investment is needed to achieve this

- Ideally negative pressure en-suite rooms with space to exercise would be available

5.3 Draft statement 3

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

Consultation comments

Stakeholders made the following comments in relation to draft statement 3:

- Include oral macrolide therapy as well as inhaled antibiotics for chronic pseudomonas suppression
- An agreed definition of chronic pseudomonas aeruginosa needs to be included. Suggestion to use the CF UK registry definition
- This may be hard to measure due to the funding disparity in service provision of nebuliser equipment
- Lung function is not affected solely by the use of inhaled antibiotics so should not be used as an outcome measure
- Need to clarify if exacerbation means clinical syndrome or the need for antibiotics
- People experiencing pulmonary exacerbation is difficult to define. The number of days of IV antibiotic use could be used instead for significant exacerbations. This is recorded on the UK CF registry
- The data is available on the UK CF registry and NHS England CF quality dashboard, apart from adherence to treatment
- The statement should be achievable but may require changes in commissioning

Consultation question 5

Stakeholders made the following comments in relation to consultation question 5:

- Not possible to say whether the medication is taken. Prescription data is entered into the database, not whether the person is taking the medication
- Reasons for the variation in prescribing rates are not known
- Some high cost drugs such as DNase and nebuliser antibiotics are stopped due to non-adherence and patient choice
- DNase should be offered to people over 6 years and the initiation of nebulised antibiotics should be timely

- Offering more dry powder inhalers instead of nebulised liquid may improve adherence rates
- Nebulised antibiotics are only tolerated in 30-40% so this may account for the variability. Some people take oral macrolides rather than nebulised antibiotics for chronic pseudomonas suppression

5.4 Draft statement 4

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

Consultation comments

Stakeholders made the following comments in relation to draft statement 4:

- This is an important and achievable statement
- The BNF for children states rhDNase is licensed for age 5 years and above
- The statement should be that people are prescribed any mucoactive agent. This is collected in the NHS England CF quality dashboard
- DNase is not tolerated by everyone. It should be offered first but may be refused, not tolerated or substituted
- Shared decision-making should be taken into account
- All CF centres have a radiology department and can perform spirometry
- A chest x-ray is a crude measure particularly in well children. Children under 6 years are unable to do reliable lung function
- This may be hard to measure due to the funding disparity in service provision of nebuliser equipment
- Statement suggests that DNase is used for people with clinical evidence of lung disease. All children with CF have lung disease from birth but DNase is licensed for five years and above
- This would be a very difficult quality statement to adhere to and to monitor

[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.

6 Suggestions for additional statements

The following is a summary of stakeholder suggestions for additional statements.

Transition

- A stakeholder commented that the quality standard misses out the important area of transition. This was discussed at the first QSAC for cystic fibrosis. The committee agreed not to progress a statement on transition due to challenges with measurability and overlaps with existing statements on transition

Genetic testing and patient journeys

- A stakeholder felt that a statement is needed on new pathogens, pseudomonas, B. cepacia and NTM (nontuberculous mycobacteria) being sent for genetic analysis. All patient journeys should be traced though out-patients and chest units as well as in-patient stays. There are no recommendations in the development source which cover this

Appendix 1: Quality standard consultation comments table – registered stakeholders

ID	Stakeholder	Statement No	Comments[2]
1	British Thoracic Society (Royal College of Physicians endorse this response)	General	BTS is pleased to support the quality statements. We have the following general observations: If the standards are used for quality dashboards then they need to be more carefully defined. The current quality dashboards are not well defined Many statements appear to be adult orientated. For example, clinical evidence of lung disease is difficult in children under 6 years of age where lung function may not be reliable and rhDNAse is not licensed in children under 5. Some may already have radiological and clinical evidence of lung disease. There are no quality measures of the early management after diagnosis through newborn screening. There are no quality measures of a network model of care as is commissioned in the paediatric age group.
2	Cystic Fibrosis Trust	General	We welcome the cystic fibrosis quality standard. We hope it will act to support and sustain evidence-based improvements in the quality of care for people with cystic fibrosis.
3	Cystic Fibrosis Trust	General	We support the comments made by the NHS England Clinical Reference Group (CRG)
4	NHS England (CF Trust support the comments made by the NHS England Clinical Reference Group)	General	In the second paragraph it suggests that desired levels of achievement should be defined locally. Our CRG advice is that this could allow a centre not to follow standard protocols, NICE guidelines, service specifications or standards or care, by simply giving a low level of desired achievements. For example if a centre does not believe that annual review is worthwhile then they can simply say that their local desired level of achievement is 0% for annual reviews which they will inevitably achieve.
5	Royal College of General Practitioners	General	The care of patients with cystic fibrosis is within secondary care and the quality standards reflect this
6	Royal College of Paediatrics and Child Health	General	Generally OK, there are many other possible quality measures but this list is manageable.
7	UK Psychosocial Professionals in CF Group (UKPPCF)	General	For future quality standards measurement of quality of life (including psychological health/wellbeing and achievement of of age appropriate social functioning eg participation in education, employment, relationships etc) would be an important area to monitor.

[2]PLEASE NOTE: Comments received in the course of consultations carried out by NICE are published in the interests of openness and transparency, and to promote understanding of how quality standards are developed. The comments are published as a record of the submissions that NICE has received, and are not endorsed by NICE, its staff or its advisory committees.

ID	Stakeholder	Statement No	Comments[2]
8	Association of Paediatric Chartered Physiotherapists (APCP)	Consultation question 1	AR- ideally a comment on there adherence to treatment should also be included
9	British Thoracic Society (Royal College of Physicians endorse this response)	Consultation question 1	The draft reflects the key areas of quality improvement. Comments on each statement and how to expand on each is given above.
10	Cystic Fibrosis Trust	Consultation question 1	The quality standard reflects key evidence-based areas of quality cystic fibrosis care. However, we are disappointed that data completeness is not included. 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 have been demonstrated to be a proxy indicator of a well-functioning unit.
11	NHS England (CF Trust support the comments made by the NHS England Clinical Reference Group)	Consultation question 1	The standard does reflect key areas for quality improvement but the advice from our CRG is that it misses out the extremely important area of transition.
12	Association of Paediatric Chartered Physiotherapists (APCP)	Consultation question 2	An Essential member of the MDT is the CF coordinator to gather all the data required. This doesn't have to be a nurse they can do other nurse related jobs. It could therefore have resource implications but not as big as if there were another CF nurse, use of Band 3 or 4 physiotherapy technician for example / or research staff at each centre.
13	British Thoracic Society (Royal College of Physicians endorse this response)	Consultation question 2	The data can be collected through the CF registry, and are achievable by local services
14	Cystic Fibrosis Trust	Consultation question 2	All cystic fibrosis specialist centres enter data into the UK CF Registry. The national UK CF Registry collects outcomes data for the proposed quality measures. Measuring quality statements using data from the UK CF

ID	Stakeholder	Statement No	Comments[2]
			Registry data will be efficient and effective. Using registry data would ensure the data is robust, and avoid duplication and unnecessary resource outlay.
15	Cystic Fibrosis Trust	Consultation question 2	The UK CF Registry is undertaking a quality of life study using national ONS wellness measures, the EQ5D-5L, and the CFQoL. These data could support quality of life outcomes data for Quality Statement 1 and Quality Statement 2. Using UK CF Registry data ensures data are robust, and avoid duplication and unnecessary resource outlay.
16	NHS England (CF Trust support the comments made by the NHS England Clinical Reference Group)	Consultation question 2	Systems are in place to collect data at local level and most importantly the UK CF registry will have some of this data which can be broken down by centres. There are some issues however and these are highlighted at the relevant part.
17	Royal College of General Practitioners	Consultation question 2	It would be useful to include the SNOMED clinical terms for this standard so that NHS secondary care trusts can use a standard method or coding the data to measure their performance to these standards and not to rely on paper based audit and quality improvement. SNOMED CT must be adopted by all GPs and in systems used by general practice service providers, before 1 April 2018 and by secondary care by 1 April 2020.
18	British Thoracic Society (Royal College of Physicians endorse this response)	Consultation question 3	Yes, this is achievable, data available through the CF Registry.
19	Cystic Fibrosis Trust	Consultation question 3	Achieving Quality Statement 1 will require further resources depending on the existing infrastructure at that service. Quality Statement 2 cannot be achieved within net resources
20	NHS England (CF Trust support the comments made by the NHS England Clinical Reference Group)	Consultation question 3	In terms of annual review, use of inhaled antibiotics, and use of mucoactive agents, these standard certainly are achievable by local services given net resources. In terms of statement two in which people should have individual rooms with ensuite facilities when admitted, this is contained within the NHS England service specifications. Some providers do not achieve this and there may be a resource impact requirement for them which would not be covered by the payment methodology.
21	British Thoracic Society (Royal College of Physicians endorse this response)	Consultation question 6	No further comments

ID	Stakeholder	Statement No	Comments[2]
22	Association of Paediatric Chartered Physiotherapists (APCP)	Statement 1	Admission delay is a bigger problem with patients waiting weeks/months for admission and can be cancelled last minuite, also relevant to time delay to urgent clinic appointments These will be exacerbated by the need for single en suite rooms and segregation clinics
23	British Society for Paediatric Endocrinology and Diabetes (BSPED)	Statement 1	Evidence also that this team includes professionals with specialist expertise in managing diabetes and liver disease, where relevant.
24	British Thoracic Society (Royal College of Physicians endorse this response)	Statement 1	This statement is valid. Comprehensive information on annual review is available through the CF registry. However it would also be helpful to measure a review of medicines and indication of patient adherence to therapies – this is not currently measured on the CF registry. In addition testing for example CF-related Diabetes testing would be helpful. t is also important to "feedback the results of annual review to the patient" – this is not currently part of the standard, but would be easy to record. Other points: Outcome 1a – FEV p4 FEV1 should be stated here as either that done on the day OR the best that year – the CF Trust database asks for both. Outcome 1c BMIS should be snapshot OR maximum as above
25	Cystic Fibrosis Trust	Statement 1	Achieving Quality Statement 1 will require further resources depending on the existing infrastructure at that service. Quality Statement 2 cannot be achieved within net resources
26	Cystic Fibrosis Trust	Statement 1	Annual review is an important process to assess health, reflect, and make changes to care strategies.
27	Cystic Fibrosis Trust	Statement 1	The annual review should take place with input from the full cystic fibrosis multidisciplinary team whether it is virtual, at a shared care clinic, or at the cystic fibrosis specialist centre.
28	Cystic Fibrosis Trust	Statement 1	The Quality Statement 1 rationale should explicitly recognise the important role of people with cystic fibrosis and/or their carers in decision-making about their own treatment and care following an annual review. We propose replacing the sentence - "[The Annual Review] 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." We believe the following phrase is more inclusive and reflective of shared decision-making best practice - "[The Annual Review] enables people with cystic fibrosis and their specialist team to understand an individual's health

ID	Stakeholder	Statement No	Comments[2]
			status and progression of disease and make changes to prevent or limit the symptoms and complications of cystic fibrosis that reflect the individual's healthcare priorities."
29	Cystic Fibrosis Trust	Statement 1	Reflecting the NICE Clinical Guideline on cystic fibrosis, the multidisciplinary team should include, or have access to, specialist social workers. This sentence should reflect this.
30	Cystic Fibrosis Trust	Statement 1	The number of people who have completed a core set of clinical assessments each year is measured in the UK CF Registry. The UK CF Registry records centre-level information, where clinical teams self-report on patients who had an annual review in person, people who had a virtual annual review, people who did not attend, and people who had no annual review. These data are subject to validation and provide the best available measure of annual review.
31	Cystic Fibrosis Trust	Statement 1	FEV1 is recorded on the UK CF Registry. The outcome measure should be the best recorded FEV1 from throughout the year instead of the measure from annual review.
32	Cystic Fibrosis Trust	Statement 1	The UK CF Registry is undertaking a quality of life study using national ONS wellness measures, the EQ5D-5L, and the CFQoL. These data could form the quality of life outcomes data for Quality Statement 1 and Quality Statement 2. Using registry data would ensure the data is robust, and avoid duplication and unnecessary resource outlay.
33	Cystic Fibrosis Trust	Statement 1	The information about what the quality statement means for people with cystic fibrosis does not recognise the role of people with cystic fibrosis and or/their carers in decision making. We propose the following text: "People with cystic fibrosis have the results of all their medical tests and checks from the last year to review with the cystic fibrosis multidisciplinary team. This review is done every year and means people with cystic fibrosis and their specialists can see how well treatment is working and whether any changes are needed."
34	Cystic Fibrosis Trust	Statement 1	The UK CF Registry is undertaking a quality of life study using national ONS wellness measures, the EQ5D-5L, and the CFQoL. These data could form the quality of life outcomes data for Quality Statement 1 and Quality Statement 2. Using registry data would ensure the data is robust, and avoid duplication and unnecessary resource outlay.
35	NHS England (CF Trust support the comments made by the NHS England Clinical Reference Group)	Statement 1	We suspect compliance with this will be fairly high. It has to be remembered that some patients simply refuse to come for an annual review and there is nothing that can be done about that. We understand the principle of checking when an annual review was done but do not understand why you then suggest outcomes on page five of lung function, health quality of life, and BMI. They do not relate to whether patients received an annual review but simply relate to the person's general health. Page 5 suggests the health related quality of life scores should be used as a data source. This is not done routinely and in clinical practice is of limited value. It is mainly used in controlled trials as an outcome measure although a few centres do carry this out. On page 7 under equality and diversity considerations, it needs to be stressed here that the annual review must be done either in a specialist CF centre or by the full specialist CF team seeing the patient in a network centre.

ID	Stakeholder	Statement No	Comments[2]
36	National Paediatric Respiratory & Allergy Nurses Group (NPRANG)	Statement 1	Whilst this key standard is what each CF team should aspire to and meet, resources vary in different hospitals. Funding is allocated / divided between specialist teams and local services which require collaborative working. The components of the annual review will all be addressed but maybe to a different level depending on staffing/expertise. QoL scores are not standard in annual review reports. Difficult to standardise in children's services and there is a psychological assessment for parents to partake in for the younger children & babies.
37	UK Psychosocial Professionals in CF Group (UKPPCF)	Statement 1	Currently states "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." Social worker should also be included in this list of specialist CF professionals that make up the MDT.
38	UK Psychosocial Professionals in CF Group (UKPPCF)	Statement 1	Currently states that "a psychological assessment" should be carried out at annual review. It should also be clear that this assessment should be by a specialist clinical psychologist (as is stated for other CF MDT members in this section)
39	UK Psychosocial Professionals in CF Group (UKPPCF)	Statement 1	This statement is measurable but not currently achieved eg some CF MDTs do still not contain specialist clinical psychology and social work staff, so this part of the annual review cannot be achieved in these cases.
40	Association of Chartered Physiotherapists in Cystic Fibrosis (ACPCF)	Consultation question 4	Annual reviews appear to be routine in all centres and network clinics already. Despite the fact that all MDT investigations are not completed on the same day, they are planned annually and conducted appropriately. The majority of centres will already have a system in place to collect and record data. The components listed in the definition of statement 1 are generally collected by centres as part of their annual review process. This standard is achievable and is likely already being achieved by the majority of CF centres.
41	Association of Paediatric Chartered Physiotherapists (APCP)	Consultation question 4	All centres complete AR and put there information into the CF registry as far as I am aware (our centre does) Data gathering of MDT involvement needs to be robust especially if not everyone sees the pt on the same day. Also recording offering appointments and pts declining needs to be recorded.
42	British Thoracic Society (Royal College of Physicians endorse this response)	Consultation question 4	All patients with CF have an Annual Review. The annual review includes all the components mention in the definition of statement 1. The annual review also has a review of medicines and compliance, usually by CF pharmacist. Annual review also includes Feedback to the patient - which is an essential part of the process.
43	NHS England (CF Trust support the comments made by the NHS England	Consultation question 4	It is not certain what proportion of patients do have annual reviews whilst your briefing paper (page 17) states the registry report shows 95% of patients have completed data based on their annual review. If the patient did not come for annual review their clinic data would be entered in order for that patient to be banded for the PBR tariff. It therefore does not necessarily mean that every patient entered actually had an annual review.

ID	Stakeholder	Statement No	Comments[2]
	Clinical Reference Group)		
44	Association of Chartered Physiotherapists in Cystic Fibrosis (ACPCF)	Statement 2	This standard is a definite area suitable for quality improvement. Not all CF units have access to single rooms and en suite facilities. In these cases, if more than one patient is resident on a ward without en suites, they may be specifically designated a bathroom/toilet, aiming to achieve as good a level of infection control as possible in the circumstances. This situation can be further complicated if CF patients with NTM or B Cepacia are admitted. It will be difficult for some centres to achieve this standard without significant financial investment in infrastructure. In fact, the ideal would be to have negative pressure en suite rooms with space to exercise.
45	Association of Paediatric Chartered Physiotherapists (APCP)	Statement 2	Ensuite rooms. Not all hospitals can offer this. They may instead offer single cf pt bathrooms. The ideal would be a negative pressure en suite room with room to exercise in. But without significant investment and space this is unlikely to happen for all centres.
46	British Thoracic Society (Royal College of Physicians endorse this response)	Statement 2	This statement should acknowledge that CF centres need to adhere to local infection control policies. The need for single rooms is to prevent cross infection - and this does not come across strongly enough in this statement. This statement is not currently measured through the CF registry - but data would be easy to collect.
47	Cystic Fibrosis Trust	Statement 2	Achieving Quality Statement 1 will require further resources depending on the existing infrastructure at that service. Quality Statement 2 cannot be achieved within net resources
48	Cystic Fibrosis Trust	Statement 2	Minimising risk is critical in clinical settings. We support the intent of this quality statement.
49	Cystic Fibrosis Trust	Statement 2	Transparency relating to cross infection prevention measures and outcomes will further develop best practice models and help drive quality improvement initiatives.
50	Cystic Fibrosis Trust	Statement 2	We are unaware of a way in which “incidence of cross infection in people with cystic fibrosis admitted as inpatients” will be measured. Tracking the incidence of bacterial strains within the UK cystic fibrosis population will require additional resources.
51	Cystic Fibrosis Trust	Statement 2	The UK CF Registry is undertaking a quality of life study using national ONS wellness measures, the EQ5D-5L, and the CFQoL. These data could form the quality of life outcomes data for Quality Statement 1 and Quality Statement 2. Using registry data would ensure the data is robust, and avoid duplication and unnecessary resource outlay.
52	NHS England (CF Trust support the comments made by the NHS England	Statement 2	This data is always being collected as part of the Quality dashboard. On page nine, one of the outcomes is incidence of cross infection in people with CF admitted as inpatients. This is extremely hard to collect and to know with certainty. This may have resource implications for providers as it involves molecular typing of bacteria being carried out on any patient who has been an inpatient and has a new organism. This is a costly undertaking, and

ID	Stakeholder	Statement No	Comments[2]
	Clinical Reference Group)		indeed whilst this testing was previously free, CF centres are now being charged for this. This would not be covered under the current payment methodology. Therefore the incidence data of cross infection will be limited. Again health related quality of life score appears as an outcome and should be omitted. On page nine under service providers, it is suggested that systems should be in place to make sure people with CF treated outside specialist CF wards are allocated individual rooms with en-suite facilities. It is part of the NHS England CF quality dashboard and service specifications, that patients should not be admitted to wards where there is not a cystic fibrosis expertise, although we are aware that this may occur.
53	NHS England (CF Trust support the comments made by the NHS England Clinical Reference Group)	Statement 2	Our CRG have raised the concern that the resource impact of insisting that every patient is in an en-suite cubicle, even though that may be desirable, is likely to have a significant resource impact on some providers. It is however stated in our NHS England CF service specifications.
54	National Paediatric Respiratory & Allergy Nurses Group (NPRANG)	Statement 2	NPRANG agrees that this is a statement that should be aspired to but due to the ongoing / current financial climate many services are unable to provide en suite facilities making this statement unachievable. Until purpose built environments are available for people with CF, teams have to work within the limits of services and ensure that local protective isolation policies are adhered to.
55	National Paediatric Respiratory & Allergy Nurses Group (NPRANG)	Statement 2	This statement may be hard to measure as who collects or checks the evidence. The UK CF registry does not retrieve this information and whilst the CF Trust previously performed peer reviews, this system is currently not taking place.
56	Royal College of General Practitioners	Statement 2	The evidence is found in the full document (pp 696-711, paras 11.4.3 to 11.6.4) and supports the guidance. However, in the quality standard statement no attention is given to the potential harms of adhering to this policy. The most likely is the potential distress in being isolated from other patients. This was considered in the full document (para 11.6.3.1.3, on p709) where variable responses from patients are reported. My own interpretation of this evidence is that the emotional response is variable (as would be expected). Another potential harm would result if such facilities were unavailable locally, so that patients were moved to more distant hospitals, though I would admit that I don't know how likely this is. Given some potential harms basic principles of good medical practice would dictate that patients should be offered isolation, not forced into it. The offer would be based on the best estimates of reducing cross infection i.e. not based on any false and implied prediction that isolation could avoid the problem altogether.
57	Association of Chartered	Statement 3	Inhalation of antibiotics for the management of chronic pseudomonas aeruginosa is a cornerstone in the management of CF. Reasons for lack of uptake are probably numerous. Poor adherence to treatment is very

ID	Stakeholder	Statement No	Comments[2]
	Physiotherapists in Cystic Fibrosis (ACPCF)		common across the board with airway clearance and nebulisers being most problematic. Once smart technologies are in place we may find even lower rates of adherence. A comprehensive nebuliser service should provide devices that deliver drugs in a fast and efficient manner. This statement is the ideal but not always achievable. There is a cost implication for e flows and I nebs and the ongoing consumables and replacement parts. These may be prohibitive to many CF units. Dry powder devices are available but only to patients unable to tolerate nebulisers. Potentially this needs to be reviewed and DPI's made available to all chronically infected CF patients. This could improve uptake Some of the newer inhaled therapies such as Levofloxacin and Cayston are not readily available in all parts of the UK. They may offer something if other inhaled therapies are not tolerated by a patient. This standard should be achievable but may require changes in commissioning
58	British Thoracic Society (Royal College of Physicians endorse this response)	Statement 3	This statement should also include oral macrolide therapy as well as inhaled antibiotics for chronic pseudomonas suppression. Macrolide therapy (Azithromycin) is recorded on the CF registry. The proportion prescribed is confounded by those who refuse, or are intolerant of one or more nebulised antibiotic. The detail should be recorded – has a person been offered a nebulised antibiotic as well as are they prescribed it. If someone NOT on an inhaled antibiotic it should be documented that they have been offered and what the reason for non prescription is. What isn't available is a registry of adherence to treatment and testing ie CF related Diabete testing-this would be helpful, when measure standards across different centres to take into account different adherence rates to different treatments and tests Outcome b: What is meant by an exacerbation? A clinical syndrome or the need for oral or IV antibiotics? The standard definition of chronic Pseudomonas infection should be used nationally (presumably the same as that used in the CF Registry) in order to compare regional differences. This information is collected.
59	Cystic Fibrosis Trust	Statement 3	People with cystic fibrosis who have chronic pseudomonas aeruginosa infection have sustained treatment with an inhaled antibiotic
60	Cystic Fibrosis Trust	Statement 3	The number of people with chronic pseudomonas aeruginosa infection is stored on the UK CF Registry. The number of people with chronic pseudomonas who are prescribed an inhaled antibiotic is also available on the UK CF Registry alongside the reasons for a person with cystic fibrosis starting or stopping an inhaled antibiotic.
61	Cystic Fibrosis Trust	Statement 3	The UK CF Registry currently measures 'intent to treat' regarding inhaled antibiotics, and reasons a person started to stopped medications. The UK CF Registry cannot measure adherence.
62	Cystic Fibrosis Trust	Statement 3	An agreed definition of "chronic Pseudomonas aeruginosa" included within or referenced in this section.
63	NHS England (CF Trust support the	Statement 3	It is likely that adherence to this metric will be high. It has already been collected as part of the NHS England CF quality dashboard and there is also UK CF registry data available.

ID	Stakeholder	Statement No	Comments[2]
	comments made by the NHS England Clinical Reference Group)		On page 12, concerning the denominator, it would be most important to define chronic pseudomonas infection as there is not a consistent approach by centres. Again we are unclear why lung function is taken as outcome measure directly from this issue, as it will be affected by so many factors, and not just use of inhaled antibiotics. Another outcome suggested is - patients experiencing pulmonary exacerbation. Again this is difficult to define and although there are definitions used in research studies, they are not used in clinical practice. The number of days intravenous antibiotic use could however be taken as a surrogate marker for significant exacerbations, and is already recorded on CF Registry.
64	National Paediatric Respiratory & Allergy Nurses Group (NPRANG)	Statement 3	This statement may be hard to measure due to the funding disparity in service provision of nebuliser equipment. Commonly in children's services, nebuliser equipment is obtained from charity sources. Outlying local services may have to use equipment that is available until effective & time efficient nebuliser equipment can be obtained.
65	National Paediatric Respiratory & Allergy Nurses Group (NPRANG)	Statement 3	Regarding proportion of people with CF who have chronic Pseudomonas aeruginosa infection – this may be hard to measure accurately due to the variation in the definition of chronic isolation. How many isolates equates to chronic isolation. One document states >3 isolates, whilst another gives a broader definition of Pseudomonas aeruginosa that has not been eradicated.
66	Royal College of Paediatrics and Child Health	Statement 3	'Pseudomonas infection' needs to be defined. Eg Single isolation or Symptoms. Prevalence depends on how often samples taken, hence variation between clinics.
67	Association of Paediatric Chartered Physiotherapists (APCP)	Consultation question 5	Some high cost drugs like Dnase / neb antibiotics are stopped due to non-adherence and patient choice, I think the importance is that Dnase is offered in the over 6 population and the initiaition of nebulised antibiotics is timley – support for the chronic growers like the CF hub trial /ineb downloads is also very useful why don't pts always use nebuliser A/B or DNAse. Once smart technologies are in place I believe we will find even lower adherence rates than medicines processions would indicate. But also remember patients do have a right to not use prescribed medicines. My concerns are how do we protect children's lungs where parents do not give nebulised drugs , poor understanding by safeguarding teams and social services to enforce a minimum adherence level in these at risk children. Also offering more DRy powder inhalers instead of nebulised liquid A/bs May improved adherence rates. Reviewing the guidelines for allowing people to convert to DPIs without having to show intolerance of nebulised A/Bs
68	British Thoracic Society (Royal College of	Consultation question 5	No comment

ID	Stakeholder	Statement No	Comments[2]
	Physicians endorse this response)		
69	British Thoracic Society (Royal College of Physicians endorse this response)	Consultation question 5	Nebulised antibiotics are only tolerated in 30-40% and therefore this may account for the variability. Also some patients are taking oral Macrolides (Azithromycin) rather than nebulised antibiotics for chronic pseudomonas suppression.
70	NHS England (CF Trust support the comments made by the NHS England Clinical Reference Group)	Consultation question 5	We cannot say whether the medication is not taken, we simply know if it is prescribed and it is the prescription data that is entered into the database rather than whether the person is actually taking the medication. We currently do not understand reasons for the variation in prescribing rates.
71	Association of Chartered Physiotherapists in Cystic Fibrosis (ACPCF)	Statement 4	This is an important and achievable standard.
72	British Thoracic Society (Royal College of Physicians endorse this response)	Statement 4	DNase is not tolerated by all patients. DNASe should be offered first but may be refused or not tolerated or substituted by Mannitol or 7N saline these are not failures but clinical expediency. Therefore it is important to distinguish between offered treatment and tolerated treatment in this case. Hypertonic saline is also an effective mucolytic and numbers of patients on this therapy could also be collected through the CF registry. The rate of inhaled antibiotics and mucolytics (DNase) should take in to account **'shared patient decision making'** (e.g. patient decides against the treatment for reasons such as burden of treatment) and **intolerance** i.e patient not able to tolerate inhaled antibiotic.
73	Cystic Fibrosis Trust	Statement 4	Statement 4 should refer to 'an accepted mucolytic agent for use in cystic fibrosis' rather than referring specifically to nrDNase.
74	NHS England (CF Trust support the comments made by the NHS England		There is a statement that DNase does not have marketing authorisation for use in children with cystic fibrosis. This is untrue and it appears in the BNF for children and within the SPC that it is licensed for children of 5 years and above.

ID	Stakeholder	Statement No	Comments[2]
	Clinical Reference Group)		
75	NHS England (CF Trust support the comments made by the NHS England Clinical Reference Group)	Statement 4	We do not understand this metric that patients with lung disease are prescribed DNase as first choice mucoactive agent. Surely the metric should be that they are prescribed any mucoactive agent which is exactly what is collected in the quality dashboard. If a patient had been on DNase and it has not helped, and then is switched to Hypertonic Saline and doing well, then clearly that is the appropriate treatment. This metric seems to be a one off outcome of whether they were offered DNase in the first place. The next big problem here is suggesting that DNase is used for patients who have clinical evidence of lung disease. One could say that every child with cystic fibrosis has lung disease right from birth given data we have on lung inflammation, CT scan changes and infant lung function, that shows that many children are affected very soon. DNase is licensed for five years and above although it is used, on occasions, in younger children. We think this would be a very difficult quality statement to adhere to and to monitor. Our CRG advice is that it would be much more useful to have transition as quality statement 4 rather than this. The document is also asking for evidence of local arrangements to provide radiological imaging and lung function testing, to identify people with cystic fibrosis who have lung disease. There will not be a single CF centre that does not have a radiology department and ability to perform spirometry. It should also be pointed out that a chest x-ray is a crude measure particularly in well children, and children under 5 to 6 years of age are unable to do reliable lung function.
76	National Paediatric Respiratory & Allergy Nurses Group (NPRANG)	Statement 4	DNase available for age > 5 years but not mentioned in the standard. Although this age group would be performing lung function, results may not be reproducible therefore clinical evidence of lung disease would not be available however a clinical decision may be made to commence treatment.
77	National Paediatric Respiratory & Allergy Nurses Group (NPRANG)	Statement 4	This statement may be hard to measure due to the funding disparity in service provision of nebuliser equipment. Commonly in children's services, nebuliser equipment is obtained from charity sources. Outlying local services may have to use equipment that is available until effective & time efficient nebuliser equipment can be obtained.
78	Cystic Fibrosis Dietitians Group	Comments on briefing paper	On the briefing paper table 4 'Summary of Suggested Quality Improvement Areas • Exploration of nutritional management strategies according to class of mutation • Nutritional strategies to promote increased lean body mass.

ID	Stakeholder	Statement No	Comments[2]
79	NHS England (CF Trust support the comments made by the NHS England Clinical Reference Group)	Comments on briefing paper	Our CRG advises that there would be resource implications for providers as regards ensuite facilities for patients and some of the testing relating to cross-infection in patients.
80	UK Psychosocial Professionals in CF Group (UKPPCF)	Comments on briefing paper	CF social worker is not included in the list of CF MDT members that should complete annual reviews.
81	UK Psychosocial Professionals in CF Group (UKPPCF)	Comments on briefing paper	Areas for assessment at annual review should also include “the family system around a child”
82	UK Psychosocial Professionals in CF Group (UKPPCF)	Comments on briefing paper	“Psychosocial indicators” (in areas for assessment by CF specialist psychologist at annual review) is rather unclear and would benefit from more definition. Also annual review should include comprehensive assessment of social functioning by CF specialist social worker.
83	UK Psychosocial Professionals in CF Group (UKPPCF)	Comments on briefing paper	References to social worker should be specialist social worker as for other CF professionals
84	Association of Paediatric Chartered Physiotherapists (APCP)	Additional areas	We should, as an absolute rule , have a standard that all new pathogens pseudomonas, b cepacia, NTM etc should be sent for genetic analysis and tracing all pt journeys though out patients and chest units as well as in pt stays, so that hospitals have the information to change practice and demand changes by their managers to the environment.
85	The British Society for Antimicrobial Chemotherapy (BSAC)	No comments	Members of The British Society for Antimicrobial Chemotherapy (BSAC) have no comments for this Quality Standard Consultation on Cystic Fibrosis.
86	Department of Health & Social Care	No comments	I wish to confirm that the Department of Health and Social Care has no substantive comments to make, regarding this consultation.
87	Royal College of Nursing	No comments	Nurses caring for people with cystic fibrosis were invited to review the draft quality standard. There are no comments to make on this document on behalf of the Royal College of Nursing.

Registered stakeholders who submitted comments at consultation

- Association of Chartered Physiotherapists in Cystic Fibrosis (ACPCF)
- Association of Paediatric Chartered Physiotherapists (APCP)
- The British Society for Antimicrobial Chemotherapy (BSAC)
- British Society for Paediatric Endocrinology and Diabetes (BSPED)
- British Thoracic Society (Royal College of Physicians endorse this response)
- Cystic Fibrosis Dietitians Group
- Cystic Fibrosis Trust
- Department of Health and Social Care
- NHS England (Cystic Fibrosis Trust support the comments made by the NHS England Clinical Reference Group)
- National Paediatric Respiratory and Allergy Nurses Group (NPRANG)
- Royal College of General Practitioners
- Royal College of Nursing
- Royal College of Paediatrics and Child Health
- Royal College of Physicians
- UK Psychosocial Professionals in Cystic Fibrosis Group (UKPPCF)