

Cystic Fibrosis Trust

UK CF Registry

Demographics proforma

June 2024
Version 1.1

This document displays all of the questions that are available on the Registry. Not all questions will be applicable to all patients.

Please direct any queries to registry@cysticfibrosis.org.uk.

Changelog		
Version	Date	Changes made
1.1	June 2024	Updates made to question 1.6 (sex and gender)
1.0	December 2023	Document published

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Patient details			
Name			
Date of birth	___ / ___ / ____	Registry ID	

1. Patient information

Consent

- 1.1 Has a **valid** UK CF Registry consent form been signed and dated for this patient?
- Yes
 - Consent withdrawn

Basic information

- 1.3 Surname _____
- 1.3a Surname at birth (if different) _____
- 1.4 Forename _____
- 1.5 Middle name _____ Not known
- 1.6a Sex registered at birth
- Male
 - Female
- 1.6b *This question is optional and should only be completed if a patient has asked for their gender to be changed within their medical record.*
- Gender
- Male
 - Female
 - Non-binary
 - Other:
- 1.7 Date of birth _____ / _____ / _____

1.8 Ethnicity

- | | |
|--|--|
| <input type="checkbox"/> White (British) | <input type="checkbox"/> Asian (Other) |
| <input type="checkbox"/> White (Irish) | <input type="checkbox"/> Mixed (White and Black Caribbean) |
| <input type="checkbox"/> White (Other) | <input type="checkbox"/> Mixed (White and Black African) |
| <input type="checkbox"/> Black (Caribbean) | <input type="checkbox"/> Mixed (White and Asian) |
| <input type="checkbox"/> Black (African) | <input type="checkbox"/> Mixed (Other) |
| <input type="checkbox"/> Black (Other) | <input type="checkbox"/> Other (Chinese) |
| <input type="checkbox"/> Asian (Indian) | <input type="checkbox"/> Other (Any other ethnic group) |
| <input type="checkbox"/> Asian (Pakistani) | <input type="checkbox"/> Prefer not to say |
| <input type="checkbox"/> Asian (Bangladeshi) | <input type="checkbox"/> Not known |

If 'other', please specify: _____

1.9 NHS number _____ Not known / not applicable

1.10 CHI number _____ Not known / not applicable

1.11 Postcode _____

1.12 Was the patient born in the UK?

- Yes
- No

If yes, county of birth: _____

If no, country of birth: _____

1.13 GP postcode _____ Not known

2. Genotyping

2. Has the patient been genotyped?

- Yes
- No

2.1 Date genotyping sample taken __ / __ / ____

Not known

2.2 How many mutations have been identified?

- 0
- 1
- 2
- 3

2.3 Genetic mutation 1 _____

Mutation 1 Poly-T tract

- 5T
- 7T
- 9T
- Not known / not applicable

2.4 Genetic mutation 2 _____

Mutation 2 Poly-T tract

- 5T
- 7T
- 9T
- Not known / not applicable

2.5 Genetic mutation 3 _____

Mutation 3 Poly-T tract

- 5T
- 7T
- 9T
- Not known / not applicable

3. Diagnosis

3.0 Birthweight _____ kg Not known

3.1 Date diagnosis of CF was confirmed with the patient/parent ___ / ___ / ____

Is this date an estimate or exact?

- Estimate
- Exact

3.2 How did the patient present with CF? Check all applicable.

- Family history
- Newborn screening
- Not known
- Genotyping
- Prenatal / antenatal
- Other

3.2a Other CF associated complications / signs

- Persistent or acute respiratory infection
- Oedema
- Electrolyte imbalance
- Failure to thrive / malnutrition
- Bronchiectasis
- Pancreatitis
- Fertility investigations
- Liver disease
- Meconium ileus
- Nasal polyps
- Rectal prolapse
- Steatorrhea / abnormal stools / malabsorption
- Other:

If the patient had meconium ileus, how was this managed?

- Surgically
- Medically
- Not known

Chloride sweat tests

	Date	Value	Origin
1	___ / ___ / ____	_____ mmol / litre <input type="checkbox"/> Not known	<input type="checkbox"/> Diagnosis <input type="checkbox"/> Investigations
2	___ / ___ / ____	_____ mmol / litre <input type="checkbox"/> Not known	<input type="checkbox"/> Diagnosis <input type="checkbox"/> Investigations
3	___ / ___ / ____	_____ mmol / litre <input type="checkbox"/> Not known	<input type="checkbox"/> Diagnosis <input type="checkbox"/> Investigations
4	___ / ___ / ____	_____ mmol / litre <input type="checkbox"/> Not known	<input type="checkbox"/> Diagnosis <input type="checkbox"/> Investigations
5	___ / ___ / ____	_____ mmol / litre <input type="checkbox"/> Not known	<input type="checkbox"/> Diagnosis <input type="checkbox"/> Investigations

Newborn screening

3.4 NBS undertaken?

- Yes
- No

3.4a NBS result

<input type="checkbox"/> Carrier	<input type="checkbox"/> IRT-1 raised, one CF-causing mutation and normal sweat test (Welsh protocol) <input type="checkbox"/> Raised IRT-1, one mut, IRT-2 below cutoff
<input type="checkbox"/> Equivocal	<input type="checkbox"/> IRT-1 raised, one mutation and indeterminate sweat test result (Welsh protocol) <input type="checkbox"/> Raised IRT-1, no mut, raised IRT-2, indeterminate sweat test <input type="checkbox"/> Raised IRT-1, one mut, raised IRT-2, indeterminate sweat test <input type="checkbox"/> Raised IRT-1, two muts, normal or indeterminate sweat test
<input type="checkbox"/> Suspected	<input type="checkbox"/> IRT-1 raised, one or two CF-causing mutations and positive sweat test <input type="checkbox"/> Raised first IRT <input type="checkbox"/> Raised IRT-1 (>99.9 th centile) <input type="checkbox"/> Raised second IRT
<input type="checkbox"/> Not suspected	<input type="checkbox"/> IRT-1 below cutoff (Welsh protocol) <input type="checkbox"/> IRT-1 normal (<99.5 th centile) <input type="checkbox"/> IRT-1 raised (>99.9 th centile), no mutations, IRT-2 below cutoff <input type="checkbox"/> IRT1 raised (<99.9 th centile), no mutations
<input type="checkbox"/> Not known	

3.4b NBS IRT results

Date 1st IRT sample taken ___ / ___ / ___ Not known

IRT result _____ ng/ml or µg/l – mean value

Date 2nd IRT sample taken ___ / ___ / ___ Not known Not taken

IRT result _____ ng/ml or µg/l – mean value

3.4c Laboratory details

Which laboratory was the sample sent to?

- Belfast, Northern Ireland
- Bristol
- Cambridge
- Cardiff, Wales (please note different Welsh protocol)
- Glasgow, Scotland
- Leeds
- Liverpool
- Manchester
- Not known
- NE Thames (GOSH)
- Newcastle
- Oxford
- Portsmouth
- SE Thames
- Sheffield
- SW Thames
- West Midlands
- Other:

Date sample sent to lab ___ / ___ / ___ Not known

3.5 Referral details

Date patient referred to regional CF team __ / __ / ____ Not known

Date patient seen by regional CF centre __ / __ / ____ Not known

Date patient seen by local CF centre __ / __ / ____ Not known
 N/A (not shared care)

3.6 Was a diagnostic faecal elastase sample taken?

Yes

No

Date of sample __ / __ / ____ Not known

Result ____ mcg/ml Not known

Outcome

3.7 Has the patient died?

Yes

No

If yes:

Date of death __ / __ / ____

Is date of death an estimate?

Yes

No

Cause of death

Cancer

Liver disease or failure

Respiratory or cardiorespiratory

Suicide

Transplantation related

Trauma

Other:

Not known

If cancer, please specify type:

- | | |
|---------------------------------|-------------------------------------|
| <input type="checkbox"/> Bowel | <input type="checkbox"/> Lymphoma |
| <input type="checkbox"/> Brain | <input type="checkbox"/> Pancreatic |
| <input type="checkbox"/> Breast | <input type="checkbox"/> Skin |
| <input type="checkbox"/> Liver | <input type="checkbox"/> Testicular |
| <input type="checkbox"/> Lung | <input type="checkbox"/> Other: |

3.8 Diagnosis reversed?

- Yes
- No

If yes:

Diagnosis reversal date __ / __ / ____

Reason for reversal of diagnosis:

- DNA analysis
- Repeat normal sweat testing
- Transepithelial potential differences
- Other:
- Not known

4. Medications and others

4.1 Medications

Drug name	Start date	End date	Reason for stopping
	__ / __ / ____	__ / __ / ____	
	__ / __ / ____	__ / __ / ____	
	__ / __ / ____	__ / __ / ____	
	__ / __ / ____	__ / __ / ____	
	__ / __ / ____	__ / __ / ____	

4.2 CFQ-R scores

Please see Appendix A to view the CFQ-R domain scores and questionnaires.

4.3 Covid vaccinations

	Vaccine name	Date received
1	<input type="checkbox"/> Oxford-AstraZeneca (AZD1222) <input type="checkbox"/> Valneva (VLA2001) <input type="checkbox"/> Novavax (NVX-CoV2373) <input type="checkbox"/> GlaxoSmithKline (SCB-2019) <input type="checkbox"/> Pfizer-BioNTech (BNT162b2) <input type="checkbox"/> Janssen and Johnson & <input type="checkbox"/> Moderna (mRNA-1273) Johnson (JNJ-78436735) <input type="checkbox"/> Other:	__ / __ / ____ <input type="checkbox"/> Accurate <input type="checkbox"/> Estimate
2	<input type="checkbox"/> Oxford-AstraZeneca (AZD1222) <input type="checkbox"/> Valneva (VLA2001) <input type="checkbox"/> Novavax (NVX-CoV2373) <input type="checkbox"/> GlaxoSmithKline (SCB-2019) <input type="checkbox"/> Pfizer-BioNTech (BNT162b2) <input type="checkbox"/> Janssen and Johnson & <input type="checkbox"/> Moderna (mRNA-1273) Johnson (JNJ-78436735) <input type="checkbox"/> Other:	__ / __ / ____ <input type="checkbox"/> Accurate <input type="checkbox"/> Estimate
3	<input type="checkbox"/> Oxford-AstraZeneca (AZD1222) <input type="checkbox"/> Valneva (VLA2001) <input type="checkbox"/> Novavax (NVX-CoV2373) <input type="checkbox"/> GlaxoSmithKline (SCB-2019) <input type="checkbox"/> Pfizer-BioNTech (BNT162b2) <input type="checkbox"/> Janssen and Johnson & <input type="checkbox"/> Moderna (mRNA-1273) Johnson (JNJ-78436735) <input type="checkbox"/> Other:	__ / __ / ____ <input type="checkbox"/> Accurate <input type="checkbox"/> Estimate

Covid tests

4.4i Primary covid test reason

- Symptoms
- Contact tracing
- Routine
- Monitoring previous positive result
- Other:

4.4ii When was the covid test carried out? __ / __ / ____

- 4.4iii Type of test
- PCR
 - Antigen (e.g. lateral flow)

- 4.4iv Was the covid test positive?
- Yes
 - No

If the test was positive:

- 4.4v Was the patient symptomatic?
- Yes
 - No

If yes, how did they present? Please tick all that apply.

- Fever
- Fatigue (tiredness)
- Altered cough
- Myalgia (muscle pain)
- Loss of smell
- Dyspnoea (shortness of breath)
- Loss of taste
- Other:

- 4.4vi Was the patient pregnant at time of diagnosis?
- Yes
 - No

- 4.4vii Was the patient admitted to hospital post diagnosis?
- Yes
 - No

If no, was the patient already an inpatient at the time of diagnosis?

- Yes
- No

If yes, was the covid diagnosis more than 9 days after the initial admission?

- Yes
- No

Please list any notable covid complications (e.g. stroke, secondary infection):

- Not applicable

- 4.5b AST liver enzyme result Not done
- Normal
 - >1 to ≤3 x ULN
 - >3 to ≤5 x ULN
 - >5 to ≤8 x ULN
 - >8 x ULN
- 4.5c GGT liver enzyme result Not done
- Normal
 - >1 to ≤3 x ULN
 - >3 to ≤5 x ULN
 - >5 to ≤8 x ULN
 - >8 x ULN
- 4.5d ALP liver enzyme result Not done
- Normal
 - >1 to ≤2 x ULN
 - >2 x ULN
- 4.5e Total bilirubin liver enzyme result Not done
- Normal
 - >1 to ≤2 x ULN
 - >2 x ULN

Appendix A – CFQ-R

A1 – CFQ-R domain scores

Please enter the scores (0-100) for each of the CFQ-R domains. You can learn more, and calculate the scores, by navigating to an external website [here](#). Alternatively, you can fill in the relevant questionnaire and the domain scores will be calculated upon completion.

You can enter multiple CFQ-R surveys per year, which can also be viewed, edited or created via the patient's 'Demographics' section from the Patient Management screen.

- | | | | |
|----|-------------|-------|--|
| 1 | Physical | _____ | <input type="checkbox"/> Not available |
| 2 | Vitality | _____ | <input type="checkbox"/> Not available |
| 3 | Emotion | _____ | <input type="checkbox"/> Not available |
| 4 | Eat | _____ | <input type="checkbox"/> Not available |
| 5 | Treat | _____ | <input type="checkbox"/> Not available |
| 6 | Health | _____ | <input type="checkbox"/> Not available |
| 7 | Social | _____ | <input type="checkbox"/> Not available |
| 8 | Body | _____ | <input type="checkbox"/> Not available |
| 9 | Role | _____ | <input type="checkbox"/> Not available |
| 10 | Weight | _____ | <input type="checkbox"/> Not available |
| 11 | Respiratory | _____ | <input type="checkbox"/> Not available |
| 12 | Digestive | _____ | <input type="checkbox"/> Not available |
| 13 | School | _____ | <input type="checkbox"/> Not available |

A2 – CFQ-R questionnaire for adolescents / adults (age 14 and over)

Understanding the impact of your illness and treatments on your everyday life can help your healthcare team keep track of your health and adjust your treatments. For this reason, this questionnaire was specifically developed for people who have cystic fibrosis. Thank you for your willingness to complete this form.

The following questions are about the current state of your health, as you perceive it. This information will allow us to better understand how you feel in your everyday life. Please answer all the questions. There are no right or wrong answers! If you are not sure how to answer, choose the response that seems closest to your situation.

Section I. Demographics

- A What is your date of birth? ___ / ___ / ____
- B What is your gender?
 Male
 Female
- C During the past two weeks, have you been on holiday or out of school or work for reasons NOT related to your health?
 Yes
 No
- D What is your current marital status?
 Single/never married Separated
 Married Remarried
 Widowed With a partner
 Divorced
- E Which of the following best describes your racial background?
 White (British) Black (Caribbean)
 White (Irish) Black (African)
 White (Other) Black (Other)
 Mixed (White and Black Caribbean) Asian (Indian)
 Mixed (White and Black African) Asian (Pakistani)
 Mixed (White and Asian) Asian (Bangladeshi)
 Mixed (Other) Asian (Other)
 Other (Chinese) Other (Any other ethnic group)
 Prefer not to say Not known

- F What is the highest level of education you have completed?
- Some secondary school or less
 - GCSEs/ O-levels
 - A/AS-levels
 - Other higher education
 - University degree
 - Professional qualification or post-graduate study
- G Which of the following best describes your current work or school status?
- Attending school outside the home
 - Taking educational courses at home
 - Seeking work
 - Working full or part time (either outside the home or at a home-based business)
 - Full time homemaker
 - Not attending school or working due to my health
 - Not working for other reasons

Section II. Quality of Life

During the past two weeks, to what extent have you had difficulty:

- 1 Performing vigorous activities such as running or playing sports
- A lot of difficulty
 - Some difficulty
 - A little difficulty
 - No difficulty
- 2 Walking as fast as others
- A lot of difficulty
 - Some difficulty
 - A little difficulty
 - No difficulty
- 3 Carrying or lifting heavy things such as books, shopping, or school bags
- A lot of difficulty
 - Some difficulty
 - A little difficulty
 - No difficulty
- 4 Climbing one flight of stairs
- A lot of difficulty
 - Some difficulty
 - A little difficulty
 - No difficulty

5 Climbing stairs as fast as others

- A lot of difficulty
- Some difficulty
- A little difficulty
- No difficulty

During the past two weeks, indicate how often:

6 You felt well

- Always
- Often
- Sometimes
- Never

7 You felt worried

- Always
- Often
- Sometimes
- Never

8 You felt useless

- Always
- Often
- Sometimes
- Never

9 You felt tired

- Always
- Often
- Sometimes
- Never

10 You felt full of energy

- Always
- Often
- Sometimes
- Never

11 You felt exhausted

- Always
- Often
- Sometimes
- Never

- 12 You felt sad
- Always
 - Often
 - Sometimes
 - Never

Thinking about the state of your health over the last two weeks:

- 13 To what extent do you have difficulty walking?
- You can walk a long time without getting tired
 - You can walk a long time but you get tired
 - You cannot walk a long time because you get tired quickly
 - You avoid walking whenever possible because it's too tiring for you
- 14 How do you feel about eating?
- Just thinking about food makes you feel sick
 - You never enjoy eating
 - You are sometimes able to enjoy eating
 - You are always able to enjoy eating
- 15 To what extent do your treatments make your daily life more difficult?
- Not at all
 - A little
 - Moderately
 - A lot
- 16 How much time do you currently spend each day on your treatments?
- A lot
 - Some
 - A little
 - Not very much
- 17 How difficult is it for you to do your treatments (including medications) each day?
- Not at all
 - A little
 - Moderately
 - Very
- 18 How do you think your health is now?
- Excellent
 - Good
 - Fair
 - Poor

Thinking about your health during the last two weeks, indicate the extent to which each sentence is true or false for you:

- 19 I have trouble recovering after physical effort
- Very true
 - Somewhat true
 - Somewhat false
 - Very false
- 20 I have to limit vigorous activities such as running or playing sports
- Very true
 - Somewhat true
 - Somewhat false
 - Very false
- 21 I have to force myself to eat
- Very true
 - Somewhat true
 - Somewhat false
 - Very false
- 22 I have to stay at home more than I want to
- Very true
 - Somewhat true
 - Somewhat false
 - Very false
- 23 I feel comfortable discussing my illness with others
- Very true
 - Somewhat true
 - Somewhat false
 - Very false
- 24 I think I am too thin
- Very true
 - Somewhat true
 - Somewhat false
 - Very false
- 25 I think I look different from others my age
- Very true
 - Somewhat true
 - Somewhat false
 - Very false

- 26 I feel bad about my physical appearance
- Very true
 - Somewhat true
 - Somewhat false
 - Very false
- 27 People are afraid that I may be contagious
- Very true
 - Somewhat true
 - Somewhat false
 - Very false
- 28 I get together with my friends a lot
- Very true
 - Somewhat true
 - Somewhat false
 - Very false
- 29 I think my coughing bothers others
- Very true
 - Somewhat true
 - Somewhat false
 - Very false
- 30 I feel comfortable going out at night
- Very true
 - Somewhat true
 - Somewhat false
 - Very false
- 31 I often feel lonely
- Very true
 - Somewhat true
 - Somewhat false
 - Very false
- 32 I feel healthy
- Very true
 - Somewhat true
 - Somewhat false
 - Very false

- 33 It is difficult to make plans for the future (for example, going to college, getting married, getting promoted at work, etc.)
- Very true
 - Somewhat true
 - Somewhat false
 - Very false
- 34 I lead a normal life
- Very true
 - Somewhat true
 - Somewhat false
 - Very false

Section III. School, Work, or Daily Activities

- 35 To what extent did you have trouble keeping up with your schoolwork, professional work, or other daily activities during the past two weeks?
- You have had no trouble keeping up
 - You have managed to keep up but it's been difficult
 - You have been behind
 - You have not been able to do these activities at all
- 36 How often were you absent from school, work, or unable to complete daily activities during the last two weeks because of your illness or treatments?
- Always
 - Often
 - Sometimes
 - Never
- 37 How often does CF get in the way of meeting your school, work, or personal goals?
- Always
 - Often
 - Sometimes
 - Never
- 38 How often does CF interfere with getting out of the house to run errands such as shopping or going to the bank?
- Always
 - Often
 - Sometimes
 - Never

Section IV. Symptom Difficulties

Indicate how you have been feeling during the past two weeks:

- 39 Have you had trouble gaining weight?
- A great deal
 - Somewhat
 - A little
 - Not at all
- 40 Have you been congested?
- A great deal
 - Somewhat
 - A little
 - Not at all
- 41 Have you been coughing during the day?
- A great deal
 - Somewhat
 - A little
 - Not at all
- 42 Have you had to cough up mucus?
- A great deal
 - Somewhat
 - A little
 - Not at all
- 43 Has your mucus been mostly:
- Clear
 - Clear to yellow
 - Yellowish-green
 - Green with traces of blood
 - Don't know

How often in the past two weeks:

- 44 Have you been wheezing?
- Always
 - Often
 - Sometimes
 - Never
- 45 Have you had trouble breathing?
- Always
 - Often
 - Sometimes
 - Never

- 46 Have you woken up during the night because you were coughing?
- Always
 - Often
 - Sometimes
 - Never
- 47 Have you had problems with wind?
- Always
 - Often
 - Sometimes
 - Never
- 48 Have you had diarrhoea?
- Always
 - Often
 - Sometimes
 - Never
- 49 Have you had abdominal pain?
- Always
 - Often
 - Sometimes
 - Never
- 50 Have you had eating problems?
- Always
 - Often
 - Sometimes
 - Never

A3 – CFQ-R questionnaire for 12-13 year olds (self-report format)

These questions are for children like you who have cystic fibrosis. Your answers will help us understand what this disease is like and how your treatments help you. So, answering these questions will help you and others like you in the future.

Please answer all the questions. There are no right or wrong answers! If you are not sure how to answer, choose the response that seems closest to your situation.

- A What is your date of birth? __ / __ / ____
- B Are you...
- Male
 - Female
- C During the past two weeks, have you been on holiday or out of school for reasons NOT related to your health?
- Yes
 - No
- D Which of the following best describes your racial background?
- | | |
|--|---|
| <input type="checkbox"/> White (British) | <input type="checkbox"/> Black (Caribbean) |
| <input type="checkbox"/> White (Irish) | <input type="checkbox"/> Black (African) |
| <input type="checkbox"/> White (Other) | <input type="checkbox"/> Black (Other) |
| <input type="checkbox"/> Mixed (White and Black Caribbean) | <input type="checkbox"/> Asian (Indian) |
| <input type="checkbox"/> Mixed (White and Black African) | <input type="checkbox"/> Asian (Pakistani) |
| <input type="checkbox"/> Mixed (White and Asian) | <input type="checkbox"/> Asian (Bangladeshi) |
| <input type="checkbox"/> Mixed (Other) | <input type="checkbox"/> Asian (Other) |
| <input type="checkbox"/> Other (Chinese) | <input type="checkbox"/> Other (Any other ethnic group) |
| <input type="checkbox"/> Prefer not to say | <input type="checkbox"/> Not known |
- E What year are you in now at school? If it's summer, the year you just finished.
- Reception
 - Year 1
 - Year 2
 - Year 3
 - Year 4
 - Year 5
 - Year 6
 - Year 7
 - Not in school

In the past two weeks:

- 1 You were able to walk as fast as others
 - Very true
 - Mostly true
 - Somewhat true
 - Not at all true

- 2 You were able to climb stairs as fast as others
 - Very true
 - Mostly true
 - Somewhat true
 - Not at all true

- 3 You were able to run, jump and climb as you wanted
 - Very true
 - Mostly true
 - Somewhat true
 - Not at all true

- 4 You were able to run as quickly and for as long as others
 - Very true
 - Mostly true
 - Somewhat true
 - Not at all true

- 5 You were able to participate in sports that you enjoy (e.g., swimming, football, dancing or others)
 - Very true
 - Mostly true
 - Somewhat true
 - Not at all true

- 6 You had difficulty carrying or lifting heavy things such as books, your school bag, or a rucksack
 - Very true
 - Mostly true
 - Somewhat true
 - Not at all true

And during these past two weeks, indicate how often:

- 7 You felt tired
- Always
 - Often
 - Sometimes
 - Never
- 8 You felt mad
- Always
 - Often
 - Sometimes
 - Never
- 9 You felt grouchy
- Always
 - Often
 - Sometimes
 - Never
- 10 You felt worried
- Always
 - Often
 - Sometimes
 - Never
- 11 You felt sad
- Always
 - Often
 - Sometimes
 - Never
- 12 You had trouble falling asleep
- Always
 - Often
 - Sometimes
 - Never
- 13 You had bad dreams or nightmares
- Always
 - Often
 - Sometimes
 - Never

14 You felt good about yourself

- Always
- Often
- Sometimes
- Never

15 You had trouble eating

- Always
- Often
- Sometimes
- Never

And during these past two weeks, indicate how often:

16 You had to stop fun activities to do your treatments

- Always
- Often
- Sometimes
- Never

17 You were forced to eat

- Always
- Often
- Sometimes
- Never

During the past two weeks:

18 You were able to do all of your treatments

- Very true
- Mostly true
- Somewhat true
- Not at all true

19 You enjoyed eating

- Very true
- Mostly true
- Somewhat true
- Not at all true

20 You got together with friends a lot

- Very true
- Mostly true
- Somewhat true
- Not at all true

- 21 You stayed at home more than you wanted to
- Very true
 - Mostly true
 - Somewhat true
 - Not at all true
- 22 You felt comfortable sleeping away from home (at a friend or family member's house or elsewhere)
- Very true
 - Mostly true
 - Somewhat true
 - Not at all true
- 23 You felt left out
- Very true
 - Mostly true
 - Somewhat true
 - Not at all true
- 24 You often invited friends to your house
- Very true
 - Mostly true
 - Somewhat true
 - Not at all true
- 25 You were teased by other children
- Very true
 - Mostly true
 - Somewhat true
 - Not at all true
- 26 You felt comfortable discussing your illness with others (friends, teachers)
- Very true
 - Mostly true
 - Somewhat true
 - Not at all true
- 27 You thought you were too short
- Very true
 - Mostly true
 - Somewhat true
 - Not at all true

- 28 You thought you were too thin
- Very true
 - Mostly true
 - Somewhat true
 - Not at all true
- 29 You thought you were physically different from others your age
- Very true
 - Mostly true
 - Somewhat true
 - Not at all true
- 30 Doing your treatments bothered you
- Very true
 - Mostly true
 - Somewhat true
 - Not at all true

Let us know how often in the past two weeks:

- 31 You coughed during the day
- Always
 - Often
 - Sometimes
 - Never
- 32 You woke up during the night because you were coughing
- Always
 - Often
 - Sometimes
 - Never
- 33 You had to cough up mucus
- Always
 - Often
 - Sometimes
 - Never
- 34 You had trouble breathing
- Always
 - Often
 - Sometimes
 - Never

35 Your stomach hurt

- Always
- Often
- Sometimes
- Never

A4 – CFQ-R questionnaire for 6-13 year olds (completed by parent/carer)

Understanding the impact of your child's illness and treatments on his or her everyday life can help your healthcare team keep track of your child's health and adjust his or her treatments. For this reason, we have developed a quality of life questionnaire specifically for parents of children with cystic fibrosis. We thank you for your willingness to complete this questionnaire.

Instructions: The following questions are about the current state of your child's health, as he or she perceives it. This information will allow us to better understand how he or she feels in everyday life.

Please answer all the questions. There are no right or wrong answers! If you are not sure how to answer, choose the response that seems closest to your child's situation.

Section I. Demographics

A What is your child's date of birth? __ / __ / ____

B What is your relationship to the child?

- | | |
|---|---|
| <input type="checkbox"/> Mother | <input type="checkbox"/> Other relative |
| <input type="checkbox"/> Father | <input type="checkbox"/> Foster mother |
| <input type="checkbox"/> Grandmother | <input type="checkbox"/> Foster father |
| <input type="checkbox"/> Grandfather | |
| <input type="checkbox"/> Other (please describe): | |

C Which of the following best describes your child's racial background?

- | | |
|--|---|
| <input type="checkbox"/> White (British) | <input type="checkbox"/> Black (Caribbean) |
| <input type="checkbox"/> White (Irish) | <input type="checkbox"/> Black (African) |
| <input type="checkbox"/> White (Other) | <input type="checkbox"/> Black (Other) |
| <input type="checkbox"/> Mixed (White and Black Caribbean) | <input type="checkbox"/> Asian (Indian) |
| <input type="checkbox"/> Mixed (White and Black African) | <input type="checkbox"/> Asian (Pakistani) |
| <input type="checkbox"/> Mixed (White and Asian) | <input type="checkbox"/> Asian (Bangladeshi) |
| <input type="checkbox"/> Mixed (Other) | <input type="checkbox"/> Asian (Other) |
| <input type="checkbox"/> Other (Chinese) | <input type="checkbox"/> Other (Any other ethnic group) |
| <input type="checkbox"/> Prefer not to say | <input type="checkbox"/> Not known |

D During the past two weeks, has your child been on holiday or out of school for reasons NOT related to their health?

- Yes
 No

Section II. Quality of Life

To what extent has your child had difficulty:

- 1 Performing vigorous activities such as running or playing sports
 - A lot of difficulty
 - Some difficulty
 - A little difficulty
 - No difficulty

- 2 Walking as fast as others
 - A lot of difficulty
 - Some difficulty
 - A little difficulty
 - No difficulty

- 3 Climbing stairs as fast as others
 - A lot of difficulty
 - Some difficulty
 - A little difficulty
 - No difficulty

- 4 Carrying or lifting heavy objects such as books, a school bag, or rucksack
 - A lot of difficulty
 - Some difficulty
 - A little difficulty
 - No difficulty

- 5 Climbing several flights of stairs
 - A lot of difficulty
 - Some difficulty
 - A little difficulty
 - No difficulty

During the past two weeks, indicate how often your child:

- 6 Seemed happy
 - Always
 - Often
 - Sometimes
 - Never

- 7 Seemed worried
- Always
 - Often
 - Sometimes
 - Never
- 8 Seemed tired
- Always
 - Often
 - Sometimes
 - Never
- 9 Seemed short-tempered
- Always
 - Often
 - Sometimes
 - Never
- 10 Seemed well
- Always
 - Often
 - Sometimes
 - Never
- 11 Seemed grouchy
- Always
 - Often
 - Sometimes
 - Never
- 12 Seemed full of energy
- Always
 - Often
 - Sometimes
 - Never
- 13 Was absent or late for school or other activities because of his/her illness or treatments
- Always
 - Often
 - Sometimes
 - Never

Thinking about the state of your child's health over the past two weeks, indicate:

- 14 The extent to which your child participated in sports and other physical activities, such as P.E. (physical education)
- Has not participated in physical activities
 - Has participated less than usual in sports
 - Has participated as much as usual but with some difficulty
 - Has been able to participate in physical activities without any difficulty
- 15 The extent to which your child has difficulty walking
- He or she can walk a long time without getting tired
 - He or she can walk a long time but gets tired
 - He or she cannot walk a long time, because he or she gets tired quickly
 - He or she avoids walking whenever possible, because it's too tiring for him or her

Thinking about your child's state of health during the past two weeks, indicate the extent to which each sentence is true or false for your child:

- 16 My child has trouble recovering after physical effort
- Very true
 - Mostly true
 - Somewhat true
 - Not at all true
- 17 Mealtimes are a struggle
- Very true
 - Mostly true
 - Somewhat true
 - Not at all true
- 18 My child's treatments get in the way of his/her activities
- Very true
 - Mostly true
 - Somewhat true
 - Not at all true
- 19 My child feels small compared to other kids the same age
- Very true
 - Mostly true
 - Somewhat true
 - Not at all true

- 20 My child feels physically different from other kids the same age
- Very true
 - Mostly true
 - Somewhat true
 - Not at all true
- 21 My child thinks that he/she is too thin
- Very true
 - Mostly true
 - Somewhat true
 - Not at all true
- 22 My child feels healthy
- Very true
 - Mostly true
 - Somewhat true
 - Not at all true
- 23 My child tends to be withdrawn
- Very true
 - Mostly true
 - Somewhat true
 - Not at all true
- 24 My child leads a normal life
- Very true
 - Mostly true
 - Somewhat true
 - Not at all true
- 25 My child has less fun than usual
- Very true
 - Mostly true
 - Somewhat true
 - Not at all true
- 26 My child has trouble getting along with others
- Very true
 - Mostly true
 - Somewhat true
 - Not at all true

- 27 My child has trouble concentrating
- Very true
 - Mostly true
 - Somewhat true
 - Not at all true
- 28 My child is able to keep up with his/her school work or holiday activities
- Very true
 - Mostly true
 - Somewhat true
 - Not at all true
- 29 My child is not doing as well as usual in school or holiday activities
- Very true
 - Mostly true
 - Somewhat true
 - Not at all true
- 30 My child spends a lot of time on his/her treatments everyday
- Very true
 - Mostly true
 - Somewhat true
 - Not at all true
- 31 How difficult is it for your child to do his/her treatments (including medications) each day?
- Not at all
 - A little
 - Moderately
 - Very
- 32 How do you think your child's health is now?
- Excellent
 - Good
 - Fair
 - Poor

Section III. Symptom Difficulties

The next set of questions is designed to determine the frequency with which your child has certain respiratory difficulties, such as coughing or shortness of breath.

Please indicate how your child has been feeling during the past two weeks:

33 My child had trouble gaining weight

- A great deal
- Somewhat
- A little
- Not at all

34 My child was congested

- A great deal
- Somewhat
- A little
- Not at all

35 My child coughed during the day

- A great deal
- Somewhat
- A little
- Not at all

36 My child had to cough up mucus

- A great deal
- Somewhat
- A little
- Not at all

37 My child's mucus has been mostly:

- Clear
- Clear to yellow
- Yellowish-green
- Green with traces of blood
- Don't know

During the past two weeks:

38 My child wheezed

- Always
- Often
- Sometimes
- Never

39 My child had trouble breathing

- Always
- Often
- Sometimes
- Never

- 40 My child woke up during the night because he/she was coughing
- Always
 - Often
 - Sometimes
 - Never
- 41 My child had wind
- Always
 - Often
 - Sometimes
 - Never
- 42 My child had diarrhoea
- Always
 - Often
 - Sometimes
 - Never
- 43 My child had abdominal pain
- Always
 - Often
 - Sometimes
 - Never
- 44 My child has had eating problems
- Always
 - Often
 - Sometimes
 - Never

A5 – CFQ-R questionnaire for 6-11 year olds (interviewer format)

This questionnaire is formatted for use by an interviewer. Please use this format for younger children. For older children who seem able to read and answer the questions on their own, such as 12 and 13 year olds, use this questionnaire in its self-report format.

There are directions for the interviewer for each section of the questionnaire. Directions that you should read to the child are indicated by quotation marks. Directions that you are to follow are underlined and set in italics.

Interviewer: *Please ask the following questions*

- A What is your date of birth? __ / __ / ____
- B Are you...
 Male
 Female
- C During the past two weeks, have you been on holiday or out of school for reasons NOT related to your health?
 Yes
 No
- D Which of the following best describes your racial background?
 White (British) Black (Caribbean)
 White (Irish) Black (African)
 White (Other) Black (Other)
 Mixed (White and Black Caribbean) Asian (Indian)
 Mixed (White and Black African) Asian (Pakistani)
 Mixed (White and Asian) Asian (Bangladeshi)
 Mixed (Other) Asian (Other)
 Other (Chinese) Other (Any other ethnic group)
 Prefer not to say Not known
- E What year are you in now at school? If it's summer, the year you just finished.
 Reception
 Year 1
 Year 2
 Year 3
 Year 4
 Year 5
 Year 6
 Year 7
 Not in school

Interviewer: Please read the following to the child:

"These questions are for children like you who have cystic fibrosis. Your answers will help us understand what this disease is like and how your treatments help you. So, answering these questions will help you and others like you in the future."

"For each question that I ask, choose one of the answers on the cards I'm about to show you."

Present the orange card to the child.

"Look at this card and read with me what it says: **very true, mostly true, somewhat true, not at all true.**"

"Here's an example: If I asked you if it is **very true, mostly true, somewhat true, not at all true** that elephants can fly, which one of the four answers on the card would you choose?"

Present the blue card to the child.

"Now, look at this card and read with me what it says: **always / often / sometimes / never.**"

"Here's another example: If I asked you if you go to the moon **always, often, sometimes, or never**, which answer on the card would you choose?"

Present the orange card to the child.

"Now, I will ask you some questions about your everyday life."

"**Tell me if you find the statements I read to you to be** very true, mostly true, somewhat true, or not at all true."

Please tick the box indicating the child's response.

During the past **two weeks**:

- 1 You were able to walk as fast as others
- Very true
 - Mostly true
 - Somewhat true
 - Not at all true

- 2 You were able to climb stairs as fast as others
- Very true
 - Mostly true
 - Somewhat true
 - Not at all true
- 3 You were able to run, jump and climb as you wanted
- Very true
 - Mostly true
 - Somewhat true
 - Not at all true
- 4 You were able to run as quickly and for as long as others
- Very true
 - Mostly true
 - Somewhat true
 - Not at all true
- 5 You were able to participate in sports that you enjoy (e.g., swimming, football, dancing or others)
- Very true
 - Mostly true
 - Somewhat true
 - Not at all true
- 6 You had difficulty carrying or lifting heavy things such as books, your school bag, or a rucksack
- Very true
 - Mostly true
 - Somewhat true
 - Not at all true

Interviewer: Present the blue card to the child.

Please tick the box indicating the child's response.

And during these past **two weeks**, tell me how often:

- 7 You felt tired
- Always
 - Often
 - Sometimes
 - Never

- 8 You felt mad
- Always
 - Often
 - Sometimes
 - Never
- 9 You felt grouchy
- Always
 - Often
 - Sometimes
 - Never
- 10 You felt worried
- Always
 - Often
 - Sometimes
 - Never
- 11 You felt sad
- Always
 - Often
 - Sometimes
 - Never
- 12 You had trouble falling asleep
- Always
 - Often
 - Sometimes
 - Never
- 13 You had bad dreams or nightmares
- Always
 - Often
 - Sometimes
 - Never
- 14 You felt good about yourself
- Always
 - Often
 - Sometimes
 - Never

- 15 You had trouble eating
- Always
 - Often
 - Sometimes
 - Never
- 16 You had to stop fun activities to do your treatments
- Always
 - Often
 - Sometimes
 - Never
- 17 You were forced to eat
- Always
 - Often
 - Sometimes
 - Never

Interviewer: Present the orange card to the child.

"Now tell me if you find the statements I read to you to be very true, mostly true, somewhat true, or not at all true."

During the past **two weeks**:

- 18 You were able to do all of your treatments
- Very true
 - Mostly true
 - Somewhat true
 - Not at all true
- 19 You enjoyed eating
- Very true
 - Mostly true
 - Somewhat true
 - Not at all true
- 20 You got together with friends a lot
- Very true
 - Mostly true
 - Somewhat true
 - Not at all true

- 21 You stayed at home more than you wanted to
- Very true
 - Mostly true
 - Somewhat true
 - Not at all true
- 22 You felt comfortable sleeping away from home (at a friend or family member's house or elsewhere)
- Very true
 - Mostly true
 - Somewhat true
 - Not at all true

During the past **two weeks**:

- 23 You felt left out
- Very true
 - Mostly true
 - Somewhat true
 - Not at all true
- 24 You often invited friends to your house
- Very true
 - Mostly true
 - Somewhat true
 - Not at all true
- 25 You were teased by other children
- Very true
 - Mostly true
 - Somewhat true
 - Not at all true
- 26 You felt comfortable discussing your illness with others (friends, teachers)
- Very true
 - Mostly true
 - Somewhat true
 - Not at all true
- 27 You thought you were too short
- Very true
 - Mostly true
 - Somewhat true
 - Not at all true

- 28 You thought you were too thin
- Very true
 - Mostly true
 - Somewhat true
 - Not at all true
- 29 You thought you were physically different from others your age
- Very true
 - Mostly true
 - Somewhat true
 - Not at all true
- 30 Doing your treatments bothered you
- Very true
 - Mostly true
 - Somewhat true
 - Not at all true

Interviewer: Present the blue card to the child again

Tell me how often in the past **two weeks**:

- 31 You coughed during the day
- Always
 - Often
 - Sometimes
 - Never
- 32 You woke up during the night because you were coughing
- Always
 - Often
 - Sometimes
 - Never
- 33 You had to cough up mucus
- Always
 - Often
 - Sometimes
 - Never
- 34 You had trouble breathing
- Always
 - Often
 - Sometimes
 - Never

35 Your stomach hurt

- Always
- Often
- Sometimes
- Never