

# **Cystic Fibrosis Trust**

## **UK CF Registry**

### **Annual review proforma**

October 2025

Version 1.4

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](mailto:registry@cysticfibrosis.org.uk).

<b>Changelog</b>		
<b>Version</b>	<b>Date</b>	<b>Changes made</b>
1.4	October 2025	Added colonoscopy question to investigations section (3.17) Added induced sputum samples (5.1) Updated NTM antibiotics list (5.2.6) Added cataracts option to 'other complications' (6.15)
1.3	February 2025	Changes made to section 9.7 (parenthood)
1.2	June 2024	Added 'patient declined' option to question 1.11 (weight not supplied) Updates made to sections 9.1-9.3 (smoking and vaping)
1.1	February 2024	Updates made to section 9.6 (parenthood)
1.0	June 2023	Document published

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

## 1. Core information

1.1 Type of encounter

- Annual review
- Encounter

1.2 Date of encounter \_\_\_ / \_\_\_ / \_\_\_\_\_

1.3 Patient age \_\_\_ years \_\_\_ months

1.4 Did the patient have an annual review?

- Yes
- No – transferred to another centre or clinic
- No – did not attend appointment
- No – patient died
- No – other
- Not known

1.5 Encounter setting

- Outpatient
- Inpatient
- Day case
- Virtual/phone
- Home visit

1.6 Is this patient shared care?

- Yes
- No

1.7 If yes:

Encounter location	
Where does this patient receive care?	
What is the patient's regional centre?	

## Height and weight

1.8 Height \_\_\_\_\_ cm  Not known

1.9 Weight \_\_\_\_\_ kg  Not known

1.10 BMI \_\_\_\_\_ kg/m<sup>2</sup>

1.11 If height/weight not supplied, please give a reason:

- Behavioural issues
- Physical disability
- Remote encounter
- Patient declined

## Oral antibiotics

1.13 Number of courses of oral antibiotics taken since the last annual review: \_\_\_\_\_

1.13a Is this an estimate?

- Estimate
- Accurate

## Oxygen and ventilation

1.14 Oxygen therapy since last annual review

- Yes
- No
- Not known

1.14a If yes, when was oxygen therapy used?

- Continuously
- Nocturnal and/or with exertion
- During exacerbation
- PRN

## Vaccinations

1.15 Has the patient received an influenza vaccine since last annual review?

- Yes
- No
- Not known

1.16 Has the patient received a pneumococcal vaccine since last annual review?

- Yes
- No
- Not known

### **Clinical trials**

1.17 Has the patient participated in any clinical drug trial since last annual review?

- Yes
- No
- Not known

1.18 Has the patient participated in any clinical study other than a drug study since last annual review?

- Yes
- No
- Not known

## 2. Admissions and IVs

2.1 IV hospital admissions since last visit: \_\_\_\_\_ Total days: \_\_\_\_\_

	Start date	End date	Total days	Admission reason
1	__/__/__	__/__/__		<input type="checkbox"/> Pulmonary exacerbation <input type="checkbox"/> Eradication of pseudomonas <input type="checkbox"/> Induction NTM <input type="checkbox"/> Haemoptysis <input type="checkbox"/> Planned IVs <input type="checkbox"/> Bronchoscopy <input type="checkbox"/> Not known <input type="checkbox"/> Other
2	__/__/__	__/__/__		<input type="checkbox"/> Pulmonary exacerbation <input type="checkbox"/> Eradication of pseudomonas <input type="checkbox"/> Induction NTM <input type="checkbox"/> Haemoptysis <input type="checkbox"/> Planned IVs <input type="checkbox"/> Bronchoscopy <input type="checkbox"/> Not known <input type="checkbox"/> Other
3	__/__/__	__/__/__		<input type="checkbox"/> Pulmonary exacerbation <input type="checkbox"/> Eradication of pseudomonas <input type="checkbox"/> Induction NTM <input type="checkbox"/> Haemoptysis <input type="checkbox"/> Planned IVs <input type="checkbox"/> Bronchoscopy <input type="checkbox"/> Not known <input type="checkbox"/> Other
4	__/__/__	__/__/__		<input type="checkbox"/> Pulmonary exacerbation <input type="checkbox"/> Eradication of pseudomonas <input type="checkbox"/> Induction NTM <input type="checkbox"/> Haemoptysis <input type="checkbox"/> Planned IVs <input type="checkbox"/> Bronchoscopy <input type="checkbox"/> Not known <input type="checkbox"/> Other
5	__/__/__	__/__/__		<input type="checkbox"/> Pulmonary exacerbation <input type="checkbox"/> Eradication of pseudomonas <input type="checkbox"/> Induction NTM <input type="checkbox"/> Haemoptysis <input type="checkbox"/> Planned IVs <input type="checkbox"/> Bronchoscopy <input type="checkbox"/> Not known <input type="checkbox"/> Other

2.2 Home IVs since last visit: \_\_\_\_\_ Total days: \_\_\_\_\_

	Start date	End date	Total days	Reason for IVs
1	__/__/__	__/__/__		<input type="checkbox"/> Pulmonary exacerbation <input type="checkbox"/> Sinus infection <input type="checkbox"/> Not known <input type="checkbox"/> Planned IVs <input type="checkbox"/> Haemoptysis <input type="checkbox"/> Other
2	__/__/__	__/__/__		<input type="checkbox"/> Pulmonary exacerbation <input type="checkbox"/> Sinus infection <input type="checkbox"/> Not known <input type="checkbox"/> Planned IVs <input type="checkbox"/> Haemoptysis <input type="checkbox"/> Other
3	__/__/__	__/__/__		<input type="checkbox"/> Pulmonary exacerbation <input type="checkbox"/> Sinus infection <input type="checkbox"/> Not known <input type="checkbox"/> Planned IVs <input type="checkbox"/> Haemoptysis <input type="checkbox"/> Other
4	__/__/__	__/__/__		<input type="checkbox"/> Pulmonary exacerbation <input type="checkbox"/> Sinus infection <input type="checkbox"/> Not known <input type="checkbox"/> Planned IVs <input type="checkbox"/> Haemoptysis <input type="checkbox"/> Other
5	__/__/__	__/__/__		<input type="checkbox"/> Pulmonary exacerbation <input type="checkbox"/> Sinus infection <input type="checkbox"/> Not known <input type="checkbox"/> Planned IVs <input type="checkbox"/> Haemoptysis <input type="checkbox"/> Other

2.3 Non-IV hospital admissions since last visit: \_\_\_\_\_

Total days: \_\_\_\_\_

	Start date	End date	Total days	Admission reason
1	__/__/__	__/__/__		<input type="checkbox"/> Non-exacerbation pulmonary complication <input type="checkbox"/> Transplant related <input type="checkbox"/> Bowel <input type="checkbox"/> Haemoptysis <input type="checkbox"/> Not known <input type="checkbox"/> GI complication <input type="checkbox"/> Non-transplant surgery <input type="checkbox"/> Liver <input type="checkbox"/> Diabetes <input type="checkbox"/> Other
2	__/__/__	__/__/__		<input type="checkbox"/> Non-exacerbation pulmonary complication <input type="checkbox"/> Transplant related <input type="checkbox"/> Bowel <input type="checkbox"/> Haemoptysis <input type="checkbox"/> Not known <input type="checkbox"/> GI complication <input type="checkbox"/> Non-transplant surgery <input type="checkbox"/> Liver <input type="checkbox"/> Diabetes <input type="checkbox"/> Other
3	__/__/__	__/__/__		<input type="checkbox"/> Non-exacerbation pulmonary complication <input type="checkbox"/> Transplant related <input type="checkbox"/> Bowel <input type="checkbox"/> Haemoptysis <input type="checkbox"/> Not known <input type="checkbox"/> GI complication <input type="checkbox"/> Non-transplant surgery <input type="checkbox"/> Liver <input type="checkbox"/> Diabetes <input type="checkbox"/> Other
4	__/__/__	__/__/__		<input type="checkbox"/> Non-exacerbation pulmonary complication <input type="checkbox"/> Transplant related <input type="checkbox"/> Bowel <input type="checkbox"/> Haemoptysis <input type="checkbox"/> Not known <input type="checkbox"/> GI complication <input type="checkbox"/> Non-transplant surgery <input type="checkbox"/> Liver <input type="checkbox"/> Diabetes <input type="checkbox"/> Other
5	__/__/__	__/__/__		<input type="checkbox"/> Non-exacerbation pulmonary complication <input type="checkbox"/> Transplant related <input type="checkbox"/> Bowel <input type="checkbox"/> Haemoptysis <input type="checkbox"/> Not known <input type="checkbox"/> GI complication <input type="checkbox"/> Non-transplant surgery <input type="checkbox"/> Liver <input type="checkbox"/> Diabetes <input type="checkbox"/> Other

### 3. Investigations

#### Pulmonary function tests

- 3.1a FEV<sub>1</sub> raw value \_\_\_\_\_ l  Not measured
- 3.1b FEV<sub>1</sub> % predicted \_\_\_\_\_ %
- 3.1c FVC raw value \_\_\_\_\_ l  Not measured
- 3.1d FVC % predicted \_\_\_\_\_ %
- 3.1e FEF 25-75 raw value \_\_\_\_\_ l/s  Not measured
- 3.1f FEF 25-75 % predicted \_\_\_\_\_ %
- 3.1g Were these spirometer readings taken at home or in hospital?
- Patient's own/home spirometer
  - Hospital spirometer
  - Not known

#### Best FEV<sub>1</sub> since last annual review

- Best FEV<sub>1</sub> not measured
- 3.2a Height at best FEV<sub>1</sub> value \_\_\_\_\_ cm
- 3.2b Weight at best FEV<sub>1</sub> value \_\_\_\_\_ kg
- 3.2c Date of best FEV<sub>1</sub> value \_\_\_ / \_\_\_ / \_\_\_\_
- 3.2d Best FEV<sub>1</sub> \_\_\_\_\_ l
- 3.2e Best FEV<sub>1</sub> % predicted \_\_\_\_\_ %
- 3.2f Were these spirometer readings taken at home or in hospital?
- Patient's own/home spirometer
  - Hospital spirometer
  - Not known

#### Faecal elastase

- 3.4 Faecal elastase \_\_\_\_\_ mcg/ml  Not known/not done

### CF-related diabetes (CFRD)

3.5 Patient screened for CFRD?

- Yes
- No
- No – prior diagnosis of CFRD
- Not known

3.5a Bloods taken?

- Yes
- No

HbA1c value \_\_\_\_\_ mmol/ml  Not measured

Random blood glucose \_\_\_\_\_ mmol/l  Not measured

Fasting blood glucose \_\_\_\_\_ mmol/l  Not measured

Oral glucose tolerance test fasting \_\_\_\_\_ mmol/l  Not measured

Oral glucose tolerance 1 hour post \_\_\_\_\_ mmol/l  Not measured

Oral glucose tolerance 2 hours post \_\_\_\_\_ mmol/l  Not measured

Continuous glucose monitoring result

- Normal  CFRD
- Abnormal  Not done

### DEXA scan

3.6 DEXA scan performed?

- Normal  Not done
- Abnormal  Not known

If DEXA scan performed:

3.6a DEXA scan date \_\_\_\_\_ / \_\_\_\_\_ / \_\_\_\_\_

3.6d DEXA scan lumbar spine over 20 years of age \_\_\_\_\_ (z-score)

3.6e DEXA scan total hip over 20 years of age \_\_\_\_\_ (z-score)

3.6g DEXA scan femoral neck over 20 years of age \_\_\_\_\_ (z-score)

## **X-ray**

- 3.7 Chest x-ray result
- No change
  - New changes
  - Done but result not known
  - Not done

## **Liver ultrasound**

- 3.8 Liver ultrasound performed?
- Yes
  - No
  - Not known

- 3.8a If liver ultrasound scan done, result:
- Normal
  - Abnormal

## **Serum creatinine**

- 3.9 Serum creatinine \_\_\_\_\_  $\mu\text{mol/l}$   Not measured

## **Liver tests**

- 3.10 Have laboratory liver enzymes been done since last encounter?
- Yes
  - No
  - Not known

If yes:

Date of liver test \_\_\_\_\_ / \_\_\_\_\_ / \_\_\_\_\_

- 3.10a ALT liver enzyme result  Not done
- Normal
  - >1 to  $\leq 3$  x ULN
  - >3 to  $\leq 5$  x ULN
  - >5 to  $\leq 8$  x ULN
  - >8 x ULN

- 3.10b 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

- 3.10c 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

- 3.10d ALP liver enzyme result  Not done
- Normal
  - >1 to ≤2 x ULN
  - >2 x ULN

- 3.10e Total bilirubin liver enzyme result  Not done
- Normal
  - >1 to ≤2 x ULN
  - >2 x ULN

### **Immunoglobulin E**

- 3.11 Total IgE at annual review \_\_\_\_\_ iu/ml  Not measured
- 3.12 Aspergillus specific IgE at annual review \_\_\_\_\_ iu/ml  Not measured

### **Immunoglobulin G**

- 3.13 Aspergillus specific IgG at annual review \_\_\_\_\_ iu/ml  Not measured

### **Eosinophils**

- 3.14 Eosinophil count at annual review \_\_\_\_\_ x10<sup>9</sup>/l  Not measured

### **Serology tests including covid-19 antibody blood tests**

- 3.15 Serology type  Serology not done
- SAR-COV-2
  - Other

Serology date      \_\_ / \_\_ / \_\_\_\_

Serology result

- Positive
- Negative
- Inconclusive

### Chloride sweat tests

3.16    Sweat chloride value                      \_\_\_\_ mmol/l                       Not measured

Sweat chloride date                      \_\_ / \_\_ / \_\_\_\_

Sweat test origin

- Diagnosis
- Investigations

### Colonoscopy

3.17    Has a colonoscopy been offered?

- Performed – normal
- Performed – abnormal finding
- Not done – not offered
- Not done – inadequate bowel prep
- Not known
- Patient declined

3.17a    Colonoscopy date (month/year)      \_\_ / \_\_\_\_                       Not known

3.17b    Was the colonoscopy performed as the result of (please tick all that apply):

- Bowel cancer screening
- FIT test
- Patient symptoms
- Not known

3.17c    Colonoscopy outcome? (Please tick all that apply)

- Adenomatous polyps
- Other polyps
- Cancer
- Diverticulitis
- Colitis
- Other:

## 4. Chronic medications

4.1 Does this patient take any chronic medications?

- Yes
- No

4.2 Chronic medication details

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

4.3 Drug intolerance (tick all that apply)

### Inhaled

- DNase
- Tobramycin
- Colistin
- Hypertonic saline

### IVs

- Meropenem
- Ceftazidime

### Oral

- CFTR modulator
- Voriconazole
- Macrolides

Other:

None known

## CFQ-R

4.4 Are CFQ-R scores available for this patient since their last annual review?

- Yes
- No

4.4i Who completed the CFQ-R questionnaire?

- Patient
- Parent or carer

4.4ii Date of CFQ-R questionnaire: \_\_ / \_\_ / \_\_\_\_

4.4iii How should the CFQ-R score be entered? (Please attach the scores/questionnaire if using this form to collect clinical data)

- Domain scores only
- Full questionnaire

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

## Covid vaccinations

### 4.5 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

## 5. Culture and microbiology

5.1.1 Number of sputum samples since last annual review \_\_\_\_\_

Number of cough/throat/nasal samples since last annual review \_\_\_\_\_

Number of bronchoscopy samples since last annual review \_\_\_\_\_

Number of induced sputum samples since last annual review \_\_\_\_\_

5.1.2 Results

- Positive sample
- No growth

5.1.3 Bacterial growth (if applicable)

- |   |  |
|---|--|
| <input type="checkbox"/> Pseudomonas aeruginosa                     | <input type="checkbox"/> Alcaligenes (Achromobacter) xylooxidans |
| <input type="checkbox"/> Other Pseudomonas species                  | <input type="checkbox"/> Escherichia coli (E coli)               |
| <input type="checkbox"/> Burkholderia cepacia complex               | <input type="checkbox"/> Haemophilus influenzae                  |
| <input type="checkbox"/> Staphylococcus aureus                      | <input type="checkbox"/> Klebsiella species                      |
| <input type="checkbox"/> MRSA                                       | <input type="checkbox"/> Pandoraea species                       |
| <input type="checkbox"/> Stenotrophomonas (Xanthomonas) maltophilia | <input type="checkbox"/> Other:                                  |

5.1.4 Fungal result (if applicable)

- |  |                                  |
|--|----------------------------------|
| <input type="checkbox"/> Aspergillus fumigatus | <input type="checkbox"/> Candida |
| <input type="checkbox"/> Aspergillus species   | <input type="checkbox"/> Other:  |
| <input type="checkbox"/> Scedosporium species  |                                  |

5.1.5 Viral result (if applicable)

- |                                     |                                 |
|-------------------------------------|---------------------------------|
| <input type="checkbox"/> SARS-COV-2 | <input type="checkbox"/> RSV    |
| <input type="checkbox"/> Influenza  | <input type="checkbox"/> Other: |

### NTM: non-tuberculosis mycobacterium

5.2.1 Has the patient had NTM positive samples since their last annual review?

- Yes
- No – negative culture sample
- No – contaminated culture sample
- No – no samples taken
- No – unknown reason

5.2.2 NTM positive sample details, if applicable:

Date of sample      \_\_ / \_\_ / \_\_\_\_

Sample type

- Sputum
- Induced sputum
- Lung biopsy
- Broncho-alveolar lavage
- Not known

Species

- M. abscessus complex (MABSC) including M. abscessus, M. bolletii, M. massiliense
- M. chelonae
- M. fortuitum
- M. genavense
- M. gordonae
- M. haemophilum
- M. immunogenum
- M. kansasii
- M. malmoense
- M. marinum
- M. mucogenicum
- M. avium complex (MAC) including M. avium, M. intracellulare
- M. nonchromogenicum
- M. scrofulaceum
- M. simiae
- M. smegmatis
- M. szulgai
- M. terrae complex
- M. ulcerans
- M. xenopi
- Mycobacterium species (unidentified)

5.2.3 Has the patient been on treatment for NTM pulmonary disease at any time since last annual review?

- Yes
- No

5.2.3a Please select NTM species being treated.

- M. abscessus complex (MABSC) including M. abscessus, M. bolletii, M. massiliense
- M. chelonae
- M. fortuitum
- M. genavense
- M. gordonae
- M. haemophilum
- M. immunogenum
- M. kansasii
- M. malmoense
- M. marinum
- M. mucogenicum
- M. avium complex (MAC) including M. avium, M. intracellulare
- M. nonchromogenicum
- M. scrofulaceum
- M. simiae
- M. smegmatis
- M. szulgai
- M. terrae complex
- M. ulcerans
- M. xenopi
- Mycobacterium species (unidentified)

5.2.3b Has the patient stopped all NTM treatment?

Yes

No

If patient has stopped treatment:

Date of stopping treatment      \_\_ / \_\_ / \_\_\_\_

Reason for stopping

Completed treatment

Declined further treatment

Intolerant of treatment

Stopped treatment then later restarted it

Other (please specify):

5.2.4 Did the patient fulfil ATS criteria for NTM pulmonary disease before starting treatment?

Yes

No

5.2.5 Was an intravenous intensive regimen used at the beginning of the NTM treatment?

Yes

No

5.2.6 Which of the following antibiotics were prescribed as NTM treatment during the last period? Please tick all that apply.

Amikacin

Avibactam/Ceftazidime

Azithromycin

Bedaquiline

Capreomycin

Cefoxitine

Clarithromycin

Ciprofloxacin

Clofazimine

Co-amoxiclav

Cotrimoxazole

Cycloserine

Doxycycline

Ertepenem

Ethambutol

Ethionamide

Imipenem

Imipenem/Relebactam

Interferon gamma

Isoniazid

Levofloxacin

Linezolid

Meropenem

Minocycline

Moxifloxacin

Ofloxacin

Prothionamide

Pyrazinamide

Rifabutin

Rifampicin

Rifinah

Rifater

Streptomycin

Tedizolid

Tetracycline

Tigecycline

None

5.2.7 Has the patient been on oral corticosteroids since the last data set?

- Yes
- No
- Not known

## Covid tests

If covid test done:

5.3i Primary covid test reason

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

5.3ii When was the covid test carried out?      \_\_ / \_\_ / \_\_\_\_

5.3iii Type of test

- PCR
- Antigen (e.g. lateral flow)

5.3iv Was the covid test positive?

- Yes
- No

If the test was positive:

5.3v Was the patient symptomatic?

- Yes
- No

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

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

5.3vi Was the patient pregnant at time of diagnosis?

- Yes
- No

5.3vii 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

#### Covid treatment

5.3viii Oral antibiotics

- Yes
- No

5.3ix IV antibiotics

- Yes
- No

5.3x Treated with steroids (e.g. dexamethasone, prednisolone, hydrocortisone)

- Yes
- No

5.3xi Did patient receive new/additional oxygen?

- Yes
- No

5.3xii Did patient receive new/additional NIV?

- Yes
- No

5.3xiii Was patient admitted to intensive care?

- Yes
- No

5.3xiv Did patient receive mechanical ventilation?

- Yes
- No

5.3xv Was patient put on ECMO?

- Yes
- No

## 6. Complications

### CF-related diabetes (CFRD) or impaired glucose tolerance

6.1 CFRD status:

- CFRD
- Steroid induced diabetes
- Impaired glucose tolerance
- Indeterminate
- No CFRD

6.1a If CFRD, please specify:

- CFRD with fasting hyperglycaemia
- CFRD without fasting hyperglycaemia
- CFRD (fasting hyperglycaemia status unknown)

If CFRD or steroid induced diabetes:

6.1b Complications:

- None
- Diabetic retinopathy
- Diabetic microalbuminuria
- Other:
- Not known

6.1c Was patient prescribed treatment?

- Yes
- No

If yes:

- Dietary change
- Oral hypoglycaemic agents
- Intermittent insulin
- Chronic insulin

### Cancer

6.2 Newly diagnosed cancer

- Yes
- No

If yes, cancer 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:     |

## Septicaemia

### 6.3 Septicaemia with positive blood cultures

- Yes
- No
- Not known

If yes, related to indwelling port catheter?

- Yes
- No
- Not known

Number of episodes \_\_\_\_\_

	Date	Culture identified	
1	___ / ___ / ____ <input type="checkbox"/> Not known	<input type="checkbox"/> Burkholderia cepacia <input type="checkbox"/> Candida <input type="checkbox"/> MRSA (methicillin resistant staphylococcus aureus) <input type="checkbox"/> Pseudomonas aeruginosa <input type="checkbox"/> Staphylococcus aureus	<input type="checkbox"/> Stenotrophomonas (Xanthomonas) maltophilia <input type="checkbox"/> Staphylococcus aureus <input type="checkbox"/> Not known <input type="checkbox"/> Other:
2	___ / ___ / ____ <input type="checkbox"/> Not known	<input type="checkbox"/> Burkholderia cepacia <input type="checkbox"/> Candida <input type="checkbox"/> MRSA (methicillin resistant staphylococcus aureus) <input type="checkbox"/> Pseudomonas aeruginosa <input type="checkbox"/> Staphylococcus aureus	<input type="checkbox"/> Stenotrophomonas (Xanthomonas) maltophilia <input type="checkbox"/> Staphylococcus aureus <input type="checkbox"/> Not known <input type="checkbox"/> Other:
3	___ / ___ / ____ <input type="checkbox"/> Not known	<input type="checkbox"/> Burkholderia cepacia <input type="checkbox"/> Candida <input type="checkbox"/> MRSA (methicillin resistant staphylococcus aureus) <input type="checkbox"/> Pseudomonas aeruginosa <input type="checkbox"/> Staphylococcus aureus	<input type="checkbox"/> Stenotrophomonas (Xanthomonas) maltophilia <input type="checkbox"/> Staphylococcus aureus <input type="checkbox"/> Not known <input type="checkbox"/> Other:
4	___ / ___ / ____ <input type="checkbox"/> Not known	<input type="checkbox"/> Burkholderia cepacia <input type="checkbox"/> Candida <input type="checkbox"/> MRSA (methicillin resistant staphylococcus aureus) <input type="checkbox"/> Pseudomonas aeruginosa <input type="checkbox"/> Staphylococcus aureus	<input type="checkbox"/> Stenotrophomonas (Xanthomonas) maltophilia <input type="checkbox"/> Staphylococcus aureus <input type="checkbox"/> Not known <input type="checkbox"/> Other:

5	___ / ___ / ___ <input type="checkbox"/> Not known	<input type="checkbox"/> Burkholderia cepacia <input type="checkbox"/> Candida <input type="checkbox"/> MRSA (methicillin resistant staphylococcus aureus) <input type="checkbox"/> Pseudomonas aeruginosa <input type="checkbox"/> Staphylococcus aureus	<input type="checkbox"/> Stenotrophomonas (Xanthomonas) maltophilia <input type="checkbox"/> Staphylococcus aureus <input type="checkbox"/> Not known <input type="checkbox"/> Other:
---	---	---	--

## Haemoptysis

### 6.4 Haemoptysis massive, severe and/or moderate

- Yes  
 No

If yes, number of episodes \_\_\_\_\_

Not known

	Type	Date
1	<input type="checkbox"/> Massive (>240ml in 24 hours) <input type="checkbox"/> Severe (>60ml and <240ml in 24 hours) <input type="checkbox"/> Moderate (>5ml and <60ml in 24 hours)	___ / ___ / ___ <input type="checkbox"/> Not known
2	<input type="checkbox"/> Massive (>240ml in 24 hours) <input type="checkbox"/> Severe (>60ml and <240ml in 24 hours) <input type="checkbox"/> Moderate (>5ml and <60ml in 24 hours)	___ / ___ / ___ <input type="checkbox"/> Not known
3	<input type="checkbox"/> Massive (>240ml in 24 hours) <input type="checkbox"/> Severe (>60ml and <240ml in 24 hours) <input type="checkbox"/> Moderate (>5ml and <60ml in 24 hours)	___ / ___ / ___ <input type="checkbox"/> Not known
4	<input type="checkbox"/> Massive (>240ml in 24 hours) <input type="checkbox"/> Severe (>60ml and <240ml in 24 hours) <input type="checkbox"/> Moderate (>5ml and <60ml in 24 hours)	___ / ___ / ___ <input type="checkbox"/> Not known
5	<input type="checkbox"/> Massive (>240ml in 24 hours) <input type="checkbox"/> Severe (>60ml and <240ml in 24 hours) <input type="checkbox"/> Moderate (>5ml and <60ml in 24 hours)	___ / ___ / ___ <input type="checkbox"/> Not known

### 6.5 Haemoptysis scanty ( $\leq$ 5mls in 24 hours)

- Yes  
 No

If yes, number of episodes \_\_\_\_\_

## Chest tightness / wheezing

### 6.6 Acute chest tightness and/or wheezing related to medication

- Yes  
 No  
 Not known

If yes, number of episodes \_\_\_\_\_

	Date	Medication details
1	__ / __ / ____	
2	__ / __ / ____	
3	__ / __ / ____	
4	__ / __ / ____	
5	__ / __ / ____	

### Cough fracture

6.7 Cough fracture

- Yes
- No

If yes, number of episodes \_\_\_\_

	Date	
1	__ / __ / ____	<input type="checkbox"/> Not known
2	__ / __ / ____	<input type="checkbox"/> Not known
3	__ / __ / ____	<input type="checkbox"/> Not known
4	__ / __ / ____	<input type="checkbox"/> Not known
5	__ / __ / ____	<input type="checkbox"/> Not known

### Pulmonary

6.8 Pulmonary abscess

- Yes
- No

If yes, number of episodes \_\_\_\_  Not known

	Date	
1	__ / __ / ____	<input type="checkbox"/> Not known
2	__ / __ / ____	<input type="checkbox"/> Not known
3	__ / __ / ____	<input type="checkbox"/> Not known
4	__ / __ / ____	<input type="checkbox"/> Not known
5	__ / __ / ____	<input type="checkbox"/> Not known

## Cardiac

6.9 Any cardiac complications

- Yes
- No

If yes, please tick all that apply:

- |   |   |
|---|---|
| <input type="checkbox"/> Arrhythmia (bradycardia)     | <input type="checkbox"/> Congenital heart disease |
| <input type="checkbox"/> Arrhythmia (tachyarrhythmia) | <input type="checkbox"/> Heart failure            |
| <input type="checkbox"/> Cardiac arrest               | <input type="checkbox"/> Ischaemic heart disease  |
| <input type="checkbox"/> Cardiomyopathy               | <input type="checkbox"/> Valvular disease         |
| <input type="checkbox"/> Congenital heart disease     | <input type="checkbox"/> Other:                   |

## Liver / gallbladder (hepatobiliary)

6.10 Any liver/gallbladder complications (including gastrointestinal bleeds with varices as source)

- Yes
- No

If yes:

Gallbladder disease (including gallbladder stones)

- Yes
- No

Raised liver enzymes

- Yes
- No

Liver disease

- Yes
- No

6.10i If liver disease, is it cystic fibrosis related liver disease?

- Yes
- No

If yes, are there any of the following additional findings?

- |  |                              |                             |
|--|------------------------------|-----------------------------|
| Hepatic steatosis (fatty liver disease)                  | <input type="checkbox"/> Yes | <input type="checkbox"/> No |
| Chronic liver disease with no cirrhosis (early fibrosis) | <input type="checkbox"/> Yes | <input type="checkbox"/> No |
| Cirrhosis with portal hypertension                       | <input type="checkbox"/> Yes | <input type="checkbox"/> No |
| Cirrhosis with no portal hypertension                    | <input type="checkbox"/> Yes | <input type="checkbox"/> No |

Please specify complications relating to cirrhosis:

- |   |                              |                             |
|---|------------------------------|-----------------------------|
| Gastrointestinal bleeding from varices              | <input type="checkbox"/> Yes | <input type="checkbox"/> No |
| Oesophageal injection or banding                    | <input type="checkbox"/> Yes | <input type="checkbox"/> No |
| Hypersplenism (i.e. WBC <3.0 or platelets <100,000) | <input type="checkbox"/> Yes | <input type="checkbox"/> No |
| Ascites   | <input type="checkbox"/> Yes | <input type="checkbox"/> No |
| Hepatic encephalopathy                              | <input type="checkbox"/> Yes | <input type="checkbox"/> No |

6.10ii Acute liver complications

- Yes
- No

If yes, was it:

1. Acute liver failure (no underlying liver disease, ALT >3x ULN, INR >2, not responsive to vitamin K)
  - Yes
  - No
2. Acute hepatitis (ALT >5x ULN and duration of illness <6 months)
  - Yes
  - No

If yes to hepatitis:

- 2a. Infectious hepatitis (Hepatitis A, B, C, EBV, CMV or other known infectious disease)
  - Yes
  - No
- 2b. Drug induced liver disease
  - Yes
  - No
- 2c. Other non-infectious (autoimmune, alcohol, or other known cause)
  - Yes – please specify:
  - No
- 2d. Not known
  - Yes
  - No

If yes to drug induced liver disease:

2bi. Suspected drug

- Levofloxacin
- Other
- Not known

2bii. Was a liver biopsy done?

- Yes
- No

2biii. If yes, what were the results?

- Hepatitis
- Cholestatic
- Mixed
- Other:

## **Gut**

6.11 Any gut complications

- Yes
- No

If yes, select all complications that apply:

- DIOS (distal intestinal obstruction syndrome)
- Fibrosing colonopathy / colonic stricture
- Intestinal obstruction
- Gastro-oesophageal reflux disease
- Gastrointestinal bleeding (non varices as source)
- Pancreatitis
- Peptic ulcer
- Rectal prolapse

## **Kidney / renal**

6.12 Any kidney or renal complications (including hypertension)

- Yes
- No

If yes, select all complications that apply:

- Hypertension
- Kidney stones
- Acute kidney injury (plasma creatinine >50% of ULN for age; requiring dialysis/intensive monitoring)
- Chronic kidney disease (chronic renal failure)

## Tendon

### 6.13 Any tendon complications

- Yes
- No

If yes:

- |                    |                              |                             |
|--------------------|------------------------------|-----------------------------|
| Tendon rupture     | <input type="checkbox"/> Yes | <input type="checkbox"/> No |
| Tendinitis         | <input type="checkbox"/> Yes | <input type="checkbox"/> No |
| Other tendinopathy | <input type="checkbox"/> Yes | <input type="checkbox"/> No |

If yes to tendon rupture:

#### A. Specify location of tendon rupture

- Shoulder/forearm
- Wrist/hand
- Hip/knee
- Ankle/foot – if yes:     Achilles     Other:
- Other:

#### B. Was the tendon rupture unilateral or bilateral?

- Unilateral – left
- Unilateral – right
- Bilateral

#### C. Treatment for tendon rupture

- NSAIDs
- Narcotics
- Other medication (please give details):
- Physiotherapy
- Injection (please specify location):
- Surgery (please specify type):

#### D. Date of tendon rupture                      \_\_ / \_\_ / \_\_\_\_

#### E. Tendon rupture outcome

- Resolved – please specify stop date: \_\_ / \_\_ / \_\_\_\_
- Permanent disability
- Ongoing
- Other:

## **ABPA (Allergic Broncho-Pulmonary Aspergillosis) / other Aspergillus disease**

### 6.14 ABPA

- |   |                              |                             |
|---|------------------------------|-----------------------------|
| ABPA (Allergic Broncho-Pulmonary Aspergillosis) | <input type="checkbox"/> Yes | <input type="checkbox"/> No |
| Aspergillosis bronchitis                        | <input type="checkbox"/> Yes | <input type="checkbox"/> No |
| Aspergilloma                                    | <input type="checkbox"/> Yes | <input type="checkbox"/> No |
| Other   | <input type="checkbox"/> Yes | <input type="checkbox"/> No |

If yes to any of the above:

Highest IgE result since last annual review: \_\_\_\_\_ IU/ml  Not known

Date of highest IgE result \_\_\_\_\_ / \_\_\_\_\_ / \_\_\_\_\_  Not known

Has patient received any active treatment for this since last annual review?

- Yes
- No

If yes, which treatment was used?

- |  |   |
|--|---|
| <input type="checkbox"/> Steroid           | <input type="checkbox"/> Nebulised amphotericin |
| <input type="checkbox"/> Azole antifungals | <input type="checkbox"/> Anti-IgE               |
| <input type="checkbox"/> Other:            |   |

## **Other complications**

### 6.15 Any other complications

- Yes
- No

If yes, select all that apply:

- |  |  |
|--|--|
| <input type="checkbox"/> Arthritis                     | <input type="checkbox"/> Osteopenia                                      |
| <input type="checkbox"/> Arthropathy                   | <input type="checkbox"/> Osteoporosis                                    |
| <input type="checkbox"/> Asthma                        | <input type="checkbox"/> Pneumothorax requiring chest drain              |
| <input type="checkbox"/> Bone fracture                 | <input type="checkbox"/> Port inserted/replaced since last annual review |
| <input type="checkbox"/> Cataract (lens opacity)       | <input type="checkbox"/> Sinus disease                                   |
| <input type="checkbox"/> Depression                    | <input type="checkbox"/> Absence of vas deferens                         |
| <input type="checkbox"/> Hearing loss                  | <input type="checkbox"/> Other:  |
| <input type="checkbox"/> Intensive care unit admission |  |
| <input type="checkbox"/> Nasal polyps                  |  |

## 7. Growth and nutrition

Please ensure you have recorded all relevant prescribed medications in the chronic medication tab e.g. Ursodeoxycholic acid.

- 7.1 Nutritional assessment carried out this encounter?  
 Yes  
 No
- 7.2 Seen by specialist CF dietitian  
 Yes  
 No
- 7.3 Assessed for oral intake  
 Yes  
 No
- 7.4 Supplemental feeding  
 None  
 Nasogastric  
 Jejunal tube  
 Yes, but method unknown  
 Oral  
 Gastrostomy  
 Parenteral  
 Not known
- 7.5 Does the patient take pancreatic enzyme supplements?  
 Yes  
 No  
 Not known
- 7.6 Dose of lipase \_\_\_\_\_ iu/kg per day  
 Not applicable  
 Not known
- 7.7 Has the patient been on oestrogen/testosterone?  
 Yes  
 No  
 Not known

## 8. Physiotherapy

### Airway clearance

#### 8.1 Primary airway clearance

- |   |   |
|---|---|
| <input type="checkbox"/> Active cycle of breathing techniques       | <input type="checkbox"/> Manual techniques (percussion, over pressures, vibrations) |
| <input type="checkbox"/> Assisted autogenic drainage                | <input type="checkbox"/> NIV (non-invasive ventilation)                             |
| <input type="checkbox"/> Autogenic drainage                         | <input type="checkbox"/> Oscillating PEP  |
| <input type="checkbox"/> Exercise                                   | <input type="checkbox"/> PEP  |
| <input type="checkbox"/> Forced expiration                          | <input type="checkbox"/> Postural drainage  |
| <input type="checkbox"/> High pressure PEP                          | <input type="checkbox"/> Vest   |
| <input type="checkbox"/> Incentive spirometer                       | <input type="checkbox"/> None   |
| <input type="checkbox"/> Manual in/ex-sufflation (aka cough assist) | <input type="checkbox"/> Other:   |

#### 8.2 Secondary airway clearance (please select all that apply)

- |   |   |
|---|---|
| <input type="checkbox"/> Active cycle of breathing techniques       | <input type="checkbox"/> Manual techniques (percussion, over pressures, vibrations) |
| <input type="checkbox"/> Assisted autogenic drainage                | <input type="checkbox"/> NIV (non-invasive ventilation)                             |
| <input type="checkbox"/> Autogenic drainage                         | <input type="checkbox"/> Oscillating PEP  |
| <input type="checkbox"/> Exercise                                   | <input type="checkbox"/> PEP  |
| <input type="checkbox"/> Forced expiration                          | <input type="checkbox"/> Postural drainage  |
| <input type="checkbox"/> High pressure PEP                          | <input type="checkbox"/> Vest   |
| <input type="checkbox"/> Incentive spirometer                       | <input type="checkbox"/> None   |
| <input type="checkbox"/> Manual in/ex-sufflation (aka cough assist) | <input type="checkbox"/> Other:   |

### NIV

#### 8.3 Has non-invasive ventilation (NIV) been used since last annual review? (Not for airway clearance)

- Yes
- No
- Not known

If yes:

For respiratory failure/to relieve work of breathing/breathlessness?

- Yes, long term
- Yes, during an exacerbation only
- No
- Not known

For any other reason? If yes, please specify the reason:

- Yes, long term
- Yes, during an exacerbation only
- No
- Not known

## **Exercise**

8.4 Has an exercise test been performed? (Please attach the results if using this form to collect clinical data)

- Yes
- No
- Not known

Please see Appendix B to view the exercise test forms.

## **Continence and posture**

8.5 Urinary incontinence

- Yes
- No
- Not known

8.6 Faecal incontinence

- Yes
- No
- Not known

8.7 Postural anomaly

- Yes
- No
- Not known

## 9. Lifestyle

### Smoking

- 9.1 Does the patient smoke cigarettes or other forms of tobacco?
- Yes, regularly, 1 pack a day or more
  - Yes, regularly, <1 pack a day
  - Yes, occasionally
  - Yes, amount unknown
  - No
  - Not asked
  - Declined to answer
- 9.2 Is the patient regularly exposed to second hand cigarette smoke and/or vape aerosol?
- Yes
  - No
  - Not known
- 9.3 Does the patient use e-cigarettes?
- Yes, regularly, every day
  - Yes, regularly, weekly
  - Yes, occasionally
  - Yes, amount unknown
  - No
  - Not asked
  - Declined to answer
- 9.3a Does the patient use nicotine in their e-cigarettes?
- Nicotine free only
  - Nicotine only
  - Both nicotine-free and nicotine
  - Not known

### Education

- 9.4 Current education level of patient
- |  |                                     |
|--|-------------------------------------|
| <input type="checkbox"/> Less than GCSE        | <input type="checkbox"/> College    |
| <input type="checkbox"/> GCSE or equivalent    | <input type="checkbox"/> University |
| <input type="checkbox"/> A level or equivalent | <input type="checkbox"/> Not known  |

## Marital status

9.5 Patient's marital status

- |  |                                    |
|--|------------------------------------|
| <input type="checkbox"/> Single, never married     | <input type="checkbox"/> Separated |
| <input type="checkbox"/> Long term partner         | <input type="checkbox"/> Divorced  |
| <input type="checkbox"/> Married/civil partnership | <input type="checkbox"/> Widowed   |
| <input type="checkbox"/> Not known                 |                                    |

## Employment

9.6a What is the patient's primary employment status?

- |                                     |   |
|-------------------------------------|---|
| <input type="checkbox"/> Full time  | <input type="checkbox"/> Disabled       |
| <input type="checkbox"/> Part time  | <input type="checkbox"/> Retired        |
| <input type="checkbox"/> Homemaker  | <input type="checkbox"/> Voluntary work |
| <input type="checkbox"/> Student    | <input type="checkbox"/> Not known      |
| <input type="checkbox"/> Unemployed |   |

9.6b Does the patient have a secondary employment status?

- |                                     |  |
|-------------------------------------|--|
| <input type="checkbox"/> Full time  | <input type="checkbox"/> Disabled            |
| <input type="checkbox"/> Part time  | <input type="checkbox"/> Retired             |
| <input type="checkbox"/> Homemaker  | <input type="checkbox"/> Voluntary work      |
| <input type="checkbox"/> Student    | <input type="checkbox"/> No – not applicable |
| <input type="checkbox"/> Unemployed |  |

## Parenthood

9.7 Since the last annual review, has the patient or their partner been pregnant?

- Yes
- No
- Not known

If yes:

9.7a Was the conception via IVF?

- Yes
- No
- Not known

9.7b Due date (month/year)    \_\_\_ / \_\_\_\_     Not known

9.7c What was the outcome of the pregnancy?

- Live birth
- Spontaneous abortion
- Stillbirth
- Therapeutic abortion
- Undelivered
- Not known

If live birth or stillbirth:

Gestational age \_\_\_\_\_ weeks  Not known

Delivery date (month/year) \_\_ / \_\_\_\_  Not known

9.7d Congenital abnormality?

- Yes
- No

If yes, please specify:

- Anencephaly
- Meningomyelocele/spina bifida
- Cyanotic congenital heart disease
- Congenital diaphragmatic hernia
- Gastroschisis
- Limb reduction defect (excluding congenital amputation & dwarfing syndromes)
- None of the above
- Omphalocele
- Gastroschisis
- Cleft lip (with or without cleft palate)
- Cleft palate alone
- Hypospadias

Down syndrome?

- Yes
- No

If yes, down syndrome karyotype:

- Karyotype confirmed
- Karyotype pending

Suspected chromosomal disorder?

- Yes
- No

If yes, chromosomal disorder karyotype:

- Karyotype confirmed
- Karyotype pending

## 10. Outcome

### Death

10.1 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:     |

### Diagnosis reversal

10.2 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

## Transplant journey

10.3i Does the CF MDT consider the patient sick enough to warrant transplant referral?

- Yes
- No

If yes, has this patient been offered evaluation for transplant?

- Yes
- No – clinically not suitable
- No – patient declined evaluation

If yes, transplant type offered evaluation for:

- Lung
- Liver
- Other:

Has the patient been evaluated for transplant?

- Yes
- No
- Waiting for first evaluation

If yes, what was the outcome of the transplant evaluation?

- Accepted
- Deferred
- Declined
- Awaiting decision from transplant team

If accepted:

Date placed onto active transplant waiting list     \_\_\_ / \_\_\_ / \_\_\_\_

Was the patient removed from the active transplant waiting list?

- Yes
- No

If yes:

Date removed from active transplant waiting list     \_\_\_ / \_\_\_ / \_\_\_\_

Reason for leaving waiting list

- Patient decision
- Clinical status improved
- Clinical status declined

10.3ii Received transplant?

- Yes
- No

If yes:

Transplant date                    \_\_ / \_\_ / \_\_\_\_

Transplant centre

- Addenbrooke's Hospital, Cambridge
- Edinburgh Royal Infirmary, Edinburgh
- Freeman Hospital, Newcastle
- Great Ormond Street Hospital, London
- Harefield Hospital, London
- Nottingham City Hospital, Nottingham
- Papworth Hospital, Cambridge
- Queen Elizabeth Hospital, Birmingham
- Royal Free Hospital, London
- St James' University Hospital, Leeds
- Wythenshawe Hospital, Manchester
- Other:

Transplant type(s):

- Bilateral lung
- Lobe from cadaver
- Liver
- Heart and lung
- Lobe from living donor
- Other:

10.3iii Within 12 months of surgery, select any complications suffered:

- None
- Lymphoproliferative disorder
- Atypical infection
- Unknown
- Bronchiolitis obliterans
- Renal failure
- Other:

## 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?
- |  |   |
|--|---|
| <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

**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

## Appendix B – exercise tests

### B1 – shuttle test

Test date                                    \_\_ / \_\_ / \_\_\_\_

Level achieved                            \_\_\_\_                                     Not known

Additional distance achieved            \_\_\_\_ metres                             Not known

Heart rate

a. Maximal heart rate                    \_\_\_\_ beats per minute                 Not known

b. Resting heart rate                    \_\_\_\_ beats per minute                 Not known

O2 saturation

a. O2 saturations (resting/baseline)    \_\_\_\_%                                     Not known

b. O2 saturations (lowest)              \_\_\_\_%                                     Not known

Recovery time                            \_\_\_\_ minutes                             Not known

Supplemental O2 required?             Yes                     No                                     Not known

BORG scale                     0-10 scale                     6-20 scale                     Not done

0-10 scale:

a. Modified BORG (0-10) – baseline

- |  |   |
|--|---|
| <input type="checkbox"/> 0 – nothing at all                        | <input type="checkbox"/> 5 – severe                             |
| <input type="checkbox"/> 0.5 – very, very slight (just noticeable) | <input type="checkbox"/> 6                                      |
| <input type="checkbox"/> 1 – very slight                           | <input type="checkbox"/> 7 – very severe                        |
| <input type="checkbox"/> 2 – slight                                | <input type="checkbox"/> 8                                      |
| <input type="checkbox"/> 3 – moderate                              | <input type="checkbox"/> 9 – very, very severe (almost maximal) |
| <input type="checkbox"/> 4 – somewhat severe                       | <input type="checkbox"/> 10 – maximal                           |

b. Modified BORG (0-10) – maximal

- |  |   |
|--|---|
| <input type="checkbox"/> 0 – nothing at all                        | <input type="checkbox"/> 5 – severe                             |
| <input type="checkbox"/> 0.5 – very, very slight (just noticeable) | <input type="checkbox"/> 6                                      |
| <input type="checkbox"/> 1 – very slight                           | <input type="checkbox"/> 7 – very severe                        |
| <input type="checkbox"/> 2 – slight                                | <input type="checkbox"/> 8                                      |
| <input type="checkbox"/> 3 – moderate                              | <input type="checkbox"/> 9 – very, very severe (almost maximal) |
| <input type="checkbox"/> 4 – somewhat severe                       | <input type="checkbox"/> 10 – maximal                           |

6-20 scale:

a. BORG (6-20) – baseline

- |   |  |
|---|--|
| <input type="checkbox"/> 6 – no exertion at all | <input type="checkbox"/> 14                    |
| <input type="checkbox"/> 7 – extremely light    | <input type="checkbox"/> 15 – hard             |
| <input type="checkbox"/> 8                      | <input type="checkbox"/> 16                    |
| <input type="checkbox"/> 9 – very light         | <input type="checkbox"/> 17 – very hard        |
| <input type="checkbox"/> 10                     | <input type="checkbox"/> 18                    |
| <input type="checkbox"/> 11 – light             | <input type="checkbox"/> 19 – extremely hard   |
| <input type="checkbox"/> 12                     | <input type="checkbox"/> 20 – maximal exertion |
| <input type="checkbox"/> 13 – somewhat hard     |  |

b. BORG (6-20) – maximal

- |   |  |
|---|--|
| <input type="checkbox"/> 6 – no exertion at all | <input type="checkbox"/> 14                    |
| <input type="checkbox"/> 7 – extremely light    | <input type="checkbox"/> 15 – hard             |
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