Cystic Fibrosis strength in numbers

UK CF Registry

1. Core Information

OVERVIEW

1.1.	What type of encounter are you recording?	Annual
1.2.	Date encounter booked for	
1.3.	Age of Patient at Encounter	(years) (months)
1.4.	Was the patient seen for this Annual Review?	○ Yes ○ No - Transferred to another centre or clinic ○ No - Did not attend ○ No - Patient died ○ No - Other ○ Not known
1.5.	Encounter setting	○ Out patient ○ Inpatient ○ Daycase ○ Virtual/Phone ○ Home visit
1.6.	Is this patient shared care?	○ Yes ○ No
1.7	Locations	
	a. Encounter Location	
	b. Where does this patient receive care?	
	c. Which is the patients' regional centre?	

Height / Weight

1.8.	Height	(cm)
	a. Height Percentile	(%)
1.9.	Weight	(kg)
	a. Weight Percentile	(%)
1.10.	BMI	(kg/m^2)
	a. BMI Percentile	(%)
1.11.	Height / Weight not supplied reason	○ Behavioural issues ○ Physical disability ○ Remote encounter

Oral antibiotics courses

1.13.	Number of courses of oral antibiotics taken since the last annual review	
	a. Is this an estimate or accurate?	○ Estimate ○ Accurate

Oxygen and ventilation

1.14. Oxygen therapy since last annual review? ○ Yes ○ No ○ Not known

Vaccinations

1.15. Has patient received an influenza vaccination since last annual review? ○ Yes ○ No ○ Not known

1.16. Has patient received a pneumococcal vaccination since last annual review? ○ Yes ○ No ○ Not known

Clinical trials

1.17. Has patient participated in any clinical drug trial since last annual review? ○ Yes ○ No ○ Not known

1.18. Has patient participated in any clinical study other than a drug study since last annual review? ○ Yes ○ No ○ Not known

2. Admissions & IVs

ADMISSIONS & IVs

Hospital IV Admissions

2.1. IV hospital admissions since last visit ▼

	Start	End	Total days	Admission reason
Hospital IV admission 1				▼
Hospital IV admission 2				▼
Hospital IV admission 3				▼
Hospital IV admission 4				▼
Hospital IV admission 5				▼
Total				(days)

Home IV Courses

2.2. Home IVs since last visit ▼

	Start	End	Total days	Reason for IVs
Home IV course 1				▼
Home IV course 2				▼
Home IV course 3				▼
Home IV course 4				▼
Home IV course 5				▼
Total				(days)

Non IV Hospital Admissions

2.3. Non IV hospital admissions since last visit ▼

	Start	End	Total days	Admission reason
Non IV hospital admission 1				▼
Non IV hospital admission 2				▼
Non IV hospital admission 3				▼
Non IV hospital admission 4				▼
Non IV hospital admission 5				▼
Total				(days)

3. Investigations

INVESTIGATIONS

Pulmonary function tests

3.1

FEV_1

a. FEV_1 raw value (l) ☐ Not measured

b. FEV_1 % predicted %

FVC

c. FVC raw value (l) ☐ Not measured

d. FVC % predicted %

FEF25-75

e. FEF25 - 75 raw value (l/s) ☐ Not measured

f. FEF25 % predicted %

Best FEV_1 since last annual review

3.2 Best FEV_1

a. Height at best FEV_1 value (cm)

b. Weight at best FEV_1 value (kg)

c. Date of best FEV_1 value

d. Best FEV_1 (l)

e. Best FEV_1 % predicted %

Faecal elastase

3.4. Faecal elastase ____ (mcg/ml) ☐ Not known or Not done

CF-related diabetes (CFRD)

3.5. Patient has been screened for CFRD?
- ○ Yes
- ○ No
- ○ No (Prior CFRD diagnosis)
- ○ Not known

a. Blood taken? ○ Yes ○ No

i. HBA1C value ____ (mmol/ml) ☐ Not done

ii. Random blood glucose ____ (mmol/l) ☐ Not done

iii. Fasting blood glucose taken ____ (mmol/l) ☐ Not done

iv. Oral glucose tolerance test fasting ____ (mmol/l) ☐ Not done

v. Oral glucose tolerance 1 hour post ____ (mmol/l) ☐ Not done

vi. Oral glucose tolerance 2 hour post ____ (mmol/l) ☐ Not done

vii. Continuous Glucose Monitoring result
- ○ Normal
- ○ Abnormal
- ○ CFRD
- ○ Not done

DEXA scan

3.6. DEXA scan performed
- ○ Normal
- ○ Abnormal
- ○ Not done
- ○ Not known

a. DEXA scan date ____

b. DEXA scan total body under 20 years of age ____ (z-score)

c. DEXA scan lumbar spine under 20 years of age ____ (z-score)

d. DEXA scan lumbar spine over 20 years of age ____ (z-score)

e. DEXA scan total hip over 20 years of age ____ (z-score)

f. DEXA scan femoral neck under 20 years of age ____ (z-score)

g. DEXA scan femoral neck over 20 years of age ____ (z-score)

h. DEXA scan lumbar spine over post menopausal women and men >50 years of age ____ (T-score)

i. DEXA scan total hip over post menopausal women and men >50 years of age ____ (T-score)

j. DEXA scan femoral neck over post menopausal women and men >50 years of age ____ (T-score)

X-ray / Scan

3.7. Chest x-ray result?
- ○ No change
- ○ New changes
- ○ Done but result Not known
- ○ Not done

Liver ultrasound

3.8. Liver ultra sound scan performed? ○ Yes ○ No ○ Not known

a. Liver ultra sound scan type ○ Normal ○ Abnormal

Serum creatinine

3.9. Serum creatinine (mmol/dl) Not done

Liver Tests

3.10. Laboratory liver enzymes done? Yes No Not Known

- A. ALT liver enzyme result
- B. AST liver enzyme result
- C. GGT liver enzyme result
- D. ALP liver enzyme result
- E. Total Bilirubin liver enzyme result

Immunoglobulin E

3.11. Total IgE at annual review (IU/ml) Not done

Serology tests - including COVID-19 Antibody blood tests

3.12 **Serology test details**

1. Serology type: *Please Select...* — Required
2. Serology date: DD/MM/YYYY Not known — Date required
3. Serology result: Positive / Negative / Inconclusive — Required

SARS-COV-2
Other

Chloride sweat tests

3.13 + Add new Sweat test

Add new

Chloride sweat test details

1. Sweat chloride value (mmol/litre) Not known — Required
2. Sweat chloride date DD/MM/YYYY Not known — Date required
3. Sweat test origin: Diagnosis / Investigations — Required

Drug name
(automatically set if created by chronic medication entry) *Please Select...*

4. Chronic Medications

CHRONIC MEDICATIONS

4.1. Has this patient had any chronic medications? ○ Yes ○ No

4.2.

	Drug name	Type	Frequency	Dosage	Start Date	End date (or N/A)	Stopping reason
1st							
2nd							
3rd							
4th							
5th							
6th							
7th							
8th							
9th							
10th							

4.3. Drug Intolerance (Please tick all that apply):

Inhaled	IVs	Oral	
☐ DNase	☐ Meropenem	☐ CFTR modifier	☑ Other
☐ Tobramycin	☐ Ceftadazime	☐ Voriconazole	☐ None known
☐ Colistin		☐ Macrolides	
☐ Hypertonic saline			

1. If 'Other' please specify

CFQ-R scores

4.4. Are CFQ-R results available for this patient since their last annual review? ◉ Yes ○ No

4.4. Please enter the scores (1-100) for each of the 12 CFQ-R domains. You can learn more, and calculate the scores, by navigating to here external website. 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.

A. Who completed this survey? ◉ Carer ○ Patient

B. Date CFQR completed by patient DD/MM/YYYY
Patient should be over 6 years of age

i. Is date an estimate? ○ Yes ○ No

1. Physical ☐ Not available

2. Vitality ☐ Not available

3. Emotion ☐ Not available

4.	Eat		Not available
5.	Treat		Not available
6.	Health		Not available
7.	Social		Not available
8.	Body		Not available
9.	Role		Not available
10.	Weight		Not available
11.	Respiratory		Not available
12.	Digestive		Not available

Vaccinations (COVID)

4.5 **Add new**

Vaccinations (COVID)

Currently this is just for COVID vaccines. Additional vaccines may be added in due course.

1. Which vaccine was received?
   - ○ Oxford-AstraZeneca (AZD1222)
   - ○ Valneva (VLA2001)
   - ○ Novavax (NVX-CoV2373)
   - ○ GlaxoSmithKline (SCB-2019)
   - ○ Pfizer-BioNTech (BNT162b2)
   - ○ Janssen and Johnson & Johnson (JNJ-78436735)
   - ○ Moderna (mRNA-1273)
   - ○ Other

   Required

2. Date received: DD/MM/YYYY

   Date required

   a. Is Received date an estimate?
   - ○ Yes - Estimate
   - ○ No - Accurate

   Required

5. Culture & Microbiology

CULTURE & MICROBIOLOGY

Respiratory microbiology

5.1.1 Number of samples

1. Number of sputum samples since last annual review
2. Number of cough/throat/nasal samples since last annual review
3. Number of Bronchoscopy samples since last annual review

5.1.2 Culture result

- ☐ Positive culture sample
- ☐ No growth
- ☐ Normal flora
- ☐ Awaited

5.1.3 Culture growth

1. Pseudomonas Aeruginosa

- ☐ Pseudomonas aeruginosa

a. Number of Pseudomonas aeruginosa samples since last annual review

b. *Pseudomonas* mucoid status

- ☐ Mucoid
- ☐ Non mucoid
- ☐ Not known

5.1.3 Bacterial growth

1. Pseudomonas Aeruginosa

- ☐ Pseudomonas aeruginosa
- ☐ Other Pseudomonas species

2. Burkholderia Cepacia complex

- ☐ Burkholderia cepacia complex

3. Staphylococcus aureus

- ☐ Staphylococcus aureus
- ☐ MRSA

4. Other Cultures

- ☐ Alcaligenes (Achromobacter) xylosoxidans
- ☐ Escherichia coli (e coli)
- ☐ Haemophilus influenzae
- ☐ Klebsiella species
- ☐ Pandoraea species
- ☐ Stenotrophomonas (Xanthomonas) maltophilia
- ☐ Bacterial Other

5.1.4. Fungal result

1. Fungal

- ☐ Aspergillus fumigatus
- ☐ Aspergillus species
- ☐ Scedosporium species
- ☐ Candida
- ☐ Fungal Other

5.1.5. Viral result

1. Viral

- ☐ SARS-COV-2
- ☐ Influenza
- ☐ RSV
- ☐ Viral Other

NTM - Non-tuberculosis Mycobacterium

5.2.1. Has the patient had NTM positive samples since last annual review? ○ Yes ◉ No

Negative Culture Result

- ○ Negative culture sample
- ○ Contaminated culture sample
- ○ No samples taken
- ○ Not known

5.2.2. NTM positive sample details:

NTM positive sample details

A. Date of Sample: DD/MM/YYYY
Date required

B. Sample Type

- ○ Sputum
- ○ Induced Sputum
- ○ Lung Biopsy
- ○ Broncho-alveolar lavage
- ○ Not known

Required

C. Species

- ☐ M. abscessus complex [MABSC] including M. abscessus, M. bolletii, M. massiliense
- ☐ M. avium complex (MAC) including M. avium, M. intracellulare
- ☐ M. chelonae
- ☐ M. fortuitum
- ☐ M. genavense
- ☐ M. gordonae
- ☐ M. haemophilum
- ☐ M. immunogenum
- ☐ M. kansasii
- ☐ M. malmoense
- ☐ M. marinum
- ☐ M. mucogenicum
- ☐ 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

a. Please select NTM species being treated

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

b. Has the patient stopped all NTM antibiotic treatment? (●) Yes ( ) No

i. Date of stopping treatment DD/MM/YYYY

ii. Reason for stopping

- ( ) Completed treatment
- ( ) Declined further treatment
- ( ) Intolerant of treatment
- ( ) Stopped treatment then later restarted it
- ( ) Other

5.2.4. Did the patient fulfil ATS criteria for NTM pulmonary disease before starting treatment? (●) Yes ( ) No

5.2.5. Was an intrevenous 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	☐ Azithromycin	☐ Capreomycin
☐ Cefoxitine	☐ Clarithromycin	☐ Ciprofloxacin
☐ Clofazimine	☐ Coamoxiclav	☐ Cotrimoxazole
☐ Cycloserine	☐ Doxycycline	☐ Ertepenem
☐ Ethambutol	☐ Ethionamide	☐ Imipenem
☐ Interferon gamma	☐ Isoniazid	☐ Levofloxacin
☐ Linezolid	☐ Meropenem	☐ Minocycline
☐ Moxifloxacin	☐ Ofloxacin	☐ Prothionamide
☐ Pyrazinamide	☐ Rifabutin	☐ Rifampicin
☐ Rifinah	☐ Rifater	☐ Streptomycin
☐ Tetracycline	☐ Tigecycline	☐ None

5.2.7. Has the patient been on oral corticosteroid since the last data set? ( ) Yes ( ) No ( ) Not known

COVID Test/Diagnosis

5.3

Acute COVID

1.	Primary COVID-19 test reason	(•) Symptoms ( ) Contact tracing ( ) Routine ( ) Monitoring previous positive result ( ) Other Required
2.	When was the COVID-19 test carried out?	DD/MM/YYYY Date required
3.	Type of test	( ) PCR ( ) Antigen (e.g. Lateral flow) Required
4.	Was the COVID-19 test positive?	(•) Yes ( ) No Required
5.	Was patient symptomatic?	( ) Yes ( ) No
6.	Was patient pregnant at time of diagnosis?	( ) Yes ( ) No
7.	Was patient admitted to hospital post diagnosis?	( ) Yes ( ) No
	a. If 'No', Was patient already an inpatient at time of COVID diagnosis?	( ) Yes ( ) No
	b. If 'Yes', Was COVID-19 diagnosis more than 9 days after original admission?	( ) Yes ( ) No

COVID-19 treatment

8.	Oral antibiotics	( ) Yes ( ) No
9.	IV antibiotics	( ) Yes ( ) No
10.	Treated with steroids (e.g. dexamethasone, prednisolone, hyrdrocortisone)	( ) Yes ( ) No
11.	Did patient receive new/additional oxygen?	( ) Yes ( ) No
12.	Did patient receive new/additional NIV?	( ) Yes ( ) No
13.	Was patient admitted to Intensive Care?	( ) Yes ( ) No
14.	Did patient receive mechanical ventilation?	( ) Yes ( ) No
15.	Was patient put on ECMO?	( ) Yes ( ) No

COMPLICATIONS

6. Any new or persisting complications since last encounter? ○ Yes ○ No

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

6.1. CFRD status:
- ○ CFRD
- ○ Steroid Induced Diabetes
- ○ Impaired glucose tolerance
- ○ Indeterminate / Undetermined
- ○ No CFRD

a. If 'CFRD', please specify
- ○ CFRD with fasting hyperglycaemia
- ○ CFRD without fasting hyperglycaemia
- ○ CFRD (fasting hyperglycaemia status unknown)

b. Complications
- ☐ None
- ☐ Diabetic Retinopathy
- ☐ Diabetic Microalbuminuria
- ☐ Other
- ☐ Not known

c. Treatment

i. Was patient prescribed treatment? ○ Yes ○ No

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

Cancer

6.2. Newly diagnosed cancer ○ Yes ○ No

a. If 'Yes', Cancer type

i. If 'Other' please specify

Septicaemia

6.3. Septicaemia positive blood cultures ○ Yes ○ No ○ Not known

a. Septicaemia related to indwelling port catheter ○ Yes ○ No ○ Not known

Number of episodes

	Date		Culture identified
1st episode		Not known	Select...
2nd episode		Not known	Select...
3rd episode		Not known	Select...
4th episode		Not known	Select...
5th episode		Not known	Select...

Haemoptysis

6.4. Haemoptysis massive, severe and/or moderate — Yes / No

a. Number of episodes — Not known

Massive/Severe/Moderate Haemoptysis episodes

	Type	Date	
1st episode			Not known
2nd episode			Not known
3rd episode			Not known
4th episode			Not known
5th episode			Not known

6.5. Haemoptysis scanty (<=5 mls in 24 hours) — Yes / No / Not known

a. Number of episodes

Chest tightness/wheezing

6.6. Acute chest tightness and/or wheezing related to medication — Yes / No / Not known

a. Acute chest tightness and/or wheezing related to medications number of episodes

	Date	Medication details
1st episode		
2nd episode		
3rd episode		
4th episode		
5th episode		

Cough Fracture

6.7. Cough fracture — Yes / No

a. Cough fracture number of episodes

1st date		Not known
2nd date		Not known
3rd date		Not known
4th date		Not known
5th date		Not known

Pulmonary

6.8. Pulmonary abscess ○ Yes ○ No

a. Number of episodes ▾ ☐ Not known

1st date ☐ Not known

2nd date ☐ Not known

3rd date ☐ Not known

4th date ☐ Not known

5th date ☐ Not known

Cardiac

6.9. Are there any cardiac complications ○ Yes ○ No

If 'Yes', please tick all that apply

☐ Arrhythmia

i. Arrhythmia type ○ Bradycarida ○ Tachyarhythmia

ii. Bradycardia options

a. HeartBlock ○ Yes ○ No

b. Pauses ○ Yes ○ No

c. Asymptomatic Bradycardia ○ Yes ○ No

iii. Tachyarrhythmia options

○ Atrial fibrillation
○ Vernticular fibrillation
○ Atrial flutter
○ Ventricular flutter
○ Paroxysm atrial tachycardia
○ Other
○ Ventricular tachycardia

a. If 'Other' please specify

☐ Cardiac arrest
☐ Cardiomyopathy
☐ Congenital heart disease
☐ Heart failure
☐ Ischaemic heart disease
☐ Valvular disease
☐ Other

Liver/ gall bladder (hepatobiliary)

6.10. Were there any liver / gall bladder complications (including gastrointestinal bleeds with varices as source) ○ Yes ○ No

a. Gall Bladder Disease ○ Yes ○ No

b. Raised Liver Enzymes ○ Yes ○ No

c. Liver disease ○ Yes ○ No

If 'Liver disease', is it:

i. Cystic fibrosis related liver disease ○ Yes ○ No

If 'CF related liver disease', are there any of the following additional findings:

1. Chronic liver Disease with no cirrhosis ○ Yes ○ No

2. Cirrhosis with portal hypertension ○ Yes ○ No

3. Cirrhosis with no portal hypertension ○ Yes ○ No

4. Gastrointestinal bleeding from varices ○ Yes ○ No

5. Hepatic Encephalopathy — Yes / No

6. Oesophageal injection or banding — Yes / No

ii. Other liver disease — Yes / No

If 'Other liver disease', 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 > 5 x ULN and duration of illness < 6 months) — Yes / No

A. Infectious — Yes / No

B. Drug induced liver disease — Yes / No

i. Suspected drug
- Levoflocaxin
- Not known
- Other

If 'Other', please specify

ii.Was a liver biopsy done? — Yes / No

If 'Yes', what were the results?
- Hepatitis
- Cholestatic
- Mixed
- Other

If 'Other', please specify

C. Other — Yes / No

If 'Other', please specify

D. Not known — Yes / No

Gut

6.11. Were there any Gut complications? — Yes / No

If 'Yes', check all complications that apply,

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

Kidney / Renal

6.12. Any Kidney/Renal complications — Yes / No

If 'Yes', check 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',

1. Tendon rupture? — Yes / No

2. Tendinitis? — Yes / No

3. Other tendinopathy? — Yes / No

ABPA - Allergic Bronchial Pulmonary Aspergillosis

6.14. ABPA — ◉ Yes ○ No

a. Highest IgE result since last annual review — _____ IU/ml ☐ Not known

b. Date of Highest IgE result — DD/MM/YYYY ☐ Not known

c. Was ABPA treated? — ◉ Yes ○ No

If 'Yes', which treatment was used

- ☐ Steroid
- ☐ Azole antifungals
- ☐ Nebulised amphoteracin
- ☐ Other

Other complications

6.15. Any other complications — ○ Yes ○ No

If 'Yes', check all complications that apply,

- ☐ Arthritis
- ☐ Arthropathy
- ☐ Asthma
- ☐ Bone fracture
- ☐ Depression
- ☐ Hearing loss
- ☐ Intensive care unit admission
- ☐ Nasal polyps
- ☐ Osteopenia
- ☐ Osteoporosis
- ☐ Pneumothorax requiring chest drain
- ☐ Port inserted or replaced since last annual review
- ☐ Sinus disease
- ☐ Absence of Vas deferens
- ☐ Other (please specify)

i. If 'Other', please specify _____

7. Growth & Nutrition

NUTRITIONAL ASSESSMENT

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
- ☐ Nasogastic
- ☐ 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? ○ Yes ○ No ○ Not known

PHYSIOTHERAPY

Airway clearance

8.1. Primary airway clearance

a. If 'Other', please specify

Secondary airway clearance

8.2 Secondary airway clearance (check all that apply)

- ☐ PEP
- ☐ Oscillating PEP
- ☐ Active Cycle of Breathing Techniques
- ☐ Assisted autogenic drainage
- ☐ Postural drainage
- ☐ VEST
- ☐ High Pressure PEP
- ☐ None
- ☐ Forced expiration
- ☐ Exercise
- ☐ Autogenic drainage
- ☐ Other

i. If 'Other', please specify

Exercise

8.3. Has an exercise test been performed? ○ Yes ○ No ○ Not known

a. If 'Yes', check all that apply

- ☐ Submaximal
- ☐ Shuttle test
- ☐ Walk test
- ☐ Step test
- ☐ Other

i. If 'Other', please specify

Continence & Posture

8.4. Urinary incontinence ○ Yes ○ No ○ Not known

8.5. Faecal incontinence ○ Yes ○ No ○ Not known

8.6. Postural abnormality ○ Yes ○ No ○ Not known

Lifestyle

Smoking

9.1. Does the patient smoke cigarettes or other forms of tobacco?

9.2. Is the patient regularly exposed to second hand smoke? ○ Yes ○ No ○ Not known

Education

9.3. Current education level of patient

Marital status

9.4. Patients marital status

Employment

9.5. What is the patients employment status?

- ○ Full time
- ○ Part time
- ○ Unemployed
- ○ Disabled

Student Not known

b. Does the patient have a 'Secondary' employment status?

Full time
Part time
Homemaker
Student
Unemployed
Disabled
Retired
Voluntary work
No - Not applicable

Pregnancy / Birth

9.6 Since the last annual review:

a. Has the patient or their partner been pregnant? Yes No Not known

b. Was conception via IVF? Yes No Not known

c. What was the outcome of the pregnancy?

d. Gestational age (weeks) Not known

e. Congenital abnormality Yes No

i. If 'Yes' please specify

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

a. Down Syndrome Yes No

i. Down Syndrome Karyotype
Karyotype confirmed
Karyotype pending

b. Suspected chromosomal disorder Yes No

i. Suspected chromosomal disorder Karyotype
Karyotype confirmed
Karyotype pending

Outcome

Death

10.1. Has the patient died? ○ Yes ◉ No

a. Date of death

i. Is date of death an estimate? ○ Yes ◉ No

b. Cause of Death

If 'Cancer', please specify
- ○ Bowel
- ○ Breast
- ○ Brain
- ○ Liver
- ○ Lung
- ○ Lymphoma
- ○ Pancreatic
- ○ Skin
- ○ Testicular
- ○ Other

Other

10.2. Diagnosis reversed? ○ Yes ◉ No

a. Diagnosis reversal date

b. Reason for reversal of diagnosis?

i. If 'Other', please specify

☐ Not known

Transplant journey

1. Does the CF MDT consider the patient sick enough to warrant transplant referral? ○ Yes ○ No

1. Has this patient been offered evaluation for transplant? ○ Yes ○ No

a. If 'No', please provide reason patient was not referred
- ○ Clinically not suitable
- ○ Patient declined evaluation

b. Transplant type offered evaluation for
- ☐ Lung
- ☐ Liver
- ☐ Other

2. Has this patient been evaluated for transplant?
- ○ Yes
- ○ No
- ○ Waiting for first evaluation

b. What was the outcome of the transplant evaluation?
- ○ Accepted
- ○ Declined
- ○ Deferred
- ○ Awaiting decision from transplant team

i. Date placed onto active transplant list DD/MM/YYYY

ii. Was patient removed from active transplant waiting list? ○ Yes ○ No

a. Date patient removed from active transplant waiting list DD/MM/YYYY

b. Reason for leaving waiting list
- ○ Patient Decision
- ○ Clinical status improved
- ○ Clinical status declined

2. Received transplant? ○ Yes ○ No

a. Transplant date DD/MM/YYYY

b. Transplant centre

c. Transplant type(s)

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

Transplant Complications

3. Within 12 months of surgery, select any complications suffered

- ☐ None
- ☐ Bronchiolitis obliterans
- ☐ Lymphoproliferative disorder
- ☐ Renal Failure
- ☐ Atypical Infection
- ☐ Other
- ☐ Unknown