# UK CF Registry

## 1. Core Information

### Overview

1.1. What type of encounter are you recording?  
   - Annual

1.2. Data encounter booked for
   - (years) (months)
   - Yes
   - No - Transferred to another centre or clinic
   - No - Did not attend
   - No - Patient died
   - No - Other
   - Not known

1.3. Age of Patient at Encounter
   - Out patient
   - Inpatient
   - Daycase
   - Virtual/Phone
   - Home visit

1.4. Was the patient seen for this Annual Review?

1.5. Encounter setting

1.6. Is this patient shared care?

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)
   - (%)

1.9. Weight
   - (kg)
   - (%)

1.10. BMI
   - (kg/m²)
   - (%)

1.11. Height / Weight not supplied reason

### Oxygen and ventilation

1.12. Oxygen therapy since last annual review?
   - Yes
   - No
   - Not known
   - Continuously
   - Nocturnal
   - During exacerbation
   - PRN

   a. When was oxygen therapy used?

   b. Used non invasive ventilation?
### Vaccinations

1.13. Has patient received an influenza vaccination since last annual review? (Yes / No / Not known)

1.14. Has patient received a pneumococcal vaccination since last annual review? (Yes / No / Not known)

### Clinical trials

1.15. Has patient participated in any clinical drug trial since last annual review? (Yes / No / Not known) 

Please specify

1.16. Has patient participated in any clinical study other than a drug study since last annual review? (Yes / No / Not known) 

Please specify

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### ADUSSIONS & IVs

#### Hospital IV Admissions

2.1. IV hospital admissions since last visit

<table>
<thead>
<tr>
<th>Start</th>
<th>End</th>
<th>Total days</th>
<th>Admission reason</th>
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</thead>
<tbody>
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</table>

#### Home IV Courses

2.2. Home IVs since last visit

<table>
<thead>
<tr>
<th>Start</th>
<th>End</th>
<th>Total days</th>
<th>Reason for IVs</th>
</tr>
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</table>

#### Non IV Hospital Admissions

2.3. Non IV hospital admissions since last visit

<table>
<thead>
<tr>
<th>Start</th>
<th>End</th>
<th>Total days</th>
<th>Admission reason</th>
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</table>
### INVESTIGATIONS

#### Pulmonary function tests

<table>
<thead>
<tr>
<th>3.1</th>
<th>FEV₁</th>
</tr>
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<tbody>
<tr>
<td>a.</td>
<td>FEV₁ raw value</td>
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<tr>
<td>b.</td>
<td>FEV₁ % predicted</td>
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</tbody>
</table>

<table>
<thead>
<tr>
<th>3.1</th>
<th>FVC</th>
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<td>c.</td>
<td>FVC raw value</td>
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<td>d.</td>
<td>FVC % predicted</td>
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<table>
<thead>
<tr>
<th>3.1</th>
<th>FEF25-75</th>
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<tbody>
<tr>
<td>e.</td>
<td>FEF25 - 75 raw value</td>
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<tr>
<td>f.</td>
<td>FEF25 % predicted</td>
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</table>

#### Best FEV₁ since last annual review

<table>
<thead>
<tr>
<th>3.2</th>
<th>Best FEV₁</th>
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<tbody>
<tr>
<td>a.</td>
<td>Height at best FEV₁ value</td>
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<tr>
<td>b.</td>
<td>Weight at best FEV₁ value</td>
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<tr>
<td>c.</td>
<td>Date of best FEV₁ value</td>
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<tr>
<td>d.</td>
<td>Best FEV₁</td>
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<tr>
<td>e.</td>
<td>Best FEV₁ % predicted</td>
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</table>

#### Faecal elastase

| 3.4 | Faecal elastase | (mcg/ml) | Not known or Not done |

#### CF-related diabetes (CFRD)

<table>
<thead>
<tr>
<th>3.5</th>
<th>Patient has been screened for CFRD?</th>
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<tbody>
<tr>
<td>a.</td>
<td>Blood taken?</td>
</tr>
<tr>
<td>i.</td>
<td>HBA1C value</td>
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<td>ii.</td>
<td>Random blood glucose</td>
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</tbody>
</table>
### DEXA scan

3.6. DEXA scan performed

- a. DEXA scan date
- b. DEXA scan total body under 20 years of age
- c. DEXA scan lumbar spine under 20 years of age
- d. DEXA scan lumbar spine over 20 years of age
- e. DEXA scan total hip over 20 years of age
- f. DEXA scan femoral neck under 20 years of age
- g. DEXA scan femoral neck over 20 years of age
- h. DEXA scan lumbar spine over post menopausal women and men >50 years of age
- i. DEXA scan total hip over post menopausal women and men >50 years of age
- j. DEXA scan femoral neck over post menopausal women and men >50 years of age

### 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 ultrasound scan performed?

- Yes
- No
- Not known

#### a. Liver ultrasound 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
4. Chronic Medications

**CHRONIC MEDICATIONS**

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

4.2.  

<table>
<thead>
<tr>
<th>Drug name</th>
<th>Type</th>
<th>Frequency</th>
<th>Dosage</th>
<th>Start Date</th>
<th>End date (or N/A)</th>
<th>Stopping reason</th>
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<tbody>
<tr>
<td>1st</td>
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4.3. Drug Intolerance (Please tick all that apply)

□ DNase  □ Tobramycin solution for inhalation  □ Colistin  □ Macrolide antibiotics  □ High-dose ibuprofen  □ Hypertonic saline  □ IV antibiotics

i. If ‘IV antibiotics’ please specify

□ None known

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

   a. Number of Pseudomonas aeruginosa samples since last annual review

   b. Pseudomonas mucoid status
d. Pseudomonas drug resistance

- Mucoid
- Non mucoid
- Not known

- Pseudomonas multi drug resistant
- Pseudomonas other resistance pattern
- Pseudomonas fully sensitive
- Pseudomonas ciprofloxacin resistant
- Chronic
- Intermittent

2. Burkholderia Cepacia complex

- Burkholderia cepacia
- Burkholderia cenocepacia
- Burkholderia multivorans
- Other Burkholderia cepacia species

3. Fungal

- Aspergillus fumigatus
- Scedosporium species
- Aspergillus species
- Candida

4. Staphylococcus aureus

- Staphylococcus aureus
- Chronic
- Intermittent

5. Other Cultures

- Alcaligenes (Achromobacter) xylosidans
- Pseudomonas Species
- Escherichia Coli
- Haemophilus influenza
- Klebsiella
- MRSA
- Pandorea
- Streptococcomonas (Xanthomas) malrophila
- Other

i. If ‘Other’, please specify

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NTM

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

- Yes
- No

<table>
<thead>
<tr>
<th>Species</th>
<th>Start date</th>
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<th>Start date</th>
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</thead>
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Has the patient stopped all NTM antibiotic treatment?

- Date of stopping treatment
- Reason for stopping

If 'Other' specify the reason

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

- Negative Culture Result

If yes, Date of Culture

Culture Type

Species

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

- Select reason why NTM treatment did not start

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

Which of the following antibiotics were prescribed as NTM treatment during the last period?

- Amikacin
- Cefoxitin
- Clofazimine
- Cycloserine
- Ethambutol
- Interferon gamma
- Linezolid
- Moxifloxacin
- Pyrazinamide
- Rifinah
- Tetracycline

- Azithromycin
- Clarithromycin
- Coamoxiclav
- Doxycycline
- Ethoniamide
- Isoniazid
- Meropenem
- Ofloxacin
- Rifabutin
- Rifaxin
- Tigecycline

- Capreomycin
- Ciprofloxacin
- Cotrimoxazole
- Ertaopenam
- Imipenem
- Levofloxacin
- Minocycline
- Prothionamide
- Rifampicin
- Streptomycin
- None

Has the patient been on oral corticosteroid since the last data set?

Has the patient been on immunosuppressive drugs since the last data set?
**COMPLICATIONS**

**HAS THE PATIENT HAD ANY OF THE FOLLOWING COMPLICATIONS SINCE THEIR LAST ANNUAL REVIEW?**

8. Any new or persisting complications since last encounter?

**CF-related diabetes (CFRD)**

**Diagnosis**

6.1 CFRD Status

- CFRD with fasting hyperglycaemia
- CFRD without fasting hyperglycaemia
- CFRD (fasting hyperglycaemia status unknown)
- Impaired glucose tolerance
- Indeterminate
- No CFRD

a. CFRD Complications

- None
- Diabetic Retinopathy
- Diabetic Microalbuminuria
- Other

- [ ] Not known

b. CFRD Treatment

i. Was patient prescribed treatment for CFRD?

- [ ] Yes
- [ ] No

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

**Cancer**

6.2 Newly diagnosed cancer

a. If 'Yes', Cancer type

i. If 'Other' please specify

**Septicaemia**

6.3 Septicaemia positive blood cultures

a. Septicaemia related to indwelling port catheter

Number of episodes

<table>
<thead>
<tr>
<th>Episode</th>
<th>Date</th>
<th>Culture identified</th>
</tr>
</thead>
<tbody>
<tr>
<td>1&lt;sup&gt;st&lt;/sup&gt; episode</td>
<td></td>
<td></td>
</tr>
<tr>
<td>2&lt;sup&gt;nd&lt;/sup&gt; episode</td>
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<tr>
<td>3&lt;sup&gt;rd&lt;/sup&gt; episode</td>
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<td>4&lt;sup&gt;th&lt;/sup&gt; episode</td>
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<tr>
<td>5&lt;sup&gt;th&lt;/sup&gt; episode</td>
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Haemoptysis

6.4. Haemoptysis massive, severe and/or moderate
   a. Number of episodes

   Massive/Severe/Moderate Haemoptysis episodes

   1st episode
   2nd episode
   3rd episode
   4th episode
   5th episode

6.5. Haemoptysis scanty (≤5 ml in 24 hours)
   a. Number of episodes

   Yes  No  Not known

Chest tightness/wheeze

6.6. Acute chest tightness and/or wheeze related to medication
   a. Acute chest tightness and/or wheeze related to medications number of episodes

   1st episode
   2nd episode
   3rd episode
   4th episode
   5th episode

   Date

   Medication details

Cough Fracture

6.7. Cough fracture
   a. Cough fracture number of episodes

   1st date
   2nd date
   3rd date
   4th date
   5th date

   Not known

Pulmonary

6.8. Pulmonary abscess
   a. Number of episodes

   1st date
   2nd date
   3rd date
   4th date
   5th date

   Not known
Cardiac

6.9. Are there any cardiac complications
   If 'Yes', please tick all that apply
   i. Arrhythmia type
   ii. Bradycardia options
      a. Heart Block
      b. Pauses
      c. Asymptomatic Bradycardia
   iii. Tachyarrhythmia options
      a. Atrial fibrillation
      b. Atrial flutter
      c. Paroxysmal atrial tachycardia
      d. Ventricular tachycardia
   a. If 'Other' please specify
      a. Cardiac arrest
      b. Cardiomyopathy
      c. Congenital heart disease
      d. Heart failure
      e. Ischaemic heart disease
      f. Valvular disease
      g. Other

Liver/ gall bladder (hepatobiliary)

8.10. Were there any liver / gall bladder complications (including gastrointestinal bleeds with varices as source)
   a. Gall Bladder Disease
   b. Raised Liver Enzymes
   c. Liver disease
   a. If 'Liver disease', is it:
      i. Cystic fibrosis related liver disease
   a. If 'CF related liver disease', are there any of the following additional findings:
      1. Chronic liver Disease with no cirrhosis
      2. Cirrhosis with portal hypertension
      3. Cirrhosis with no portal hypertension
      4. Gastrointestinal bleeding from varices
      5. Hepatic Encephalopathy
      6. Oesophageal injection or banding
   ii. Other liver disease
   a. If 'Other liver disease', was it:
      1. Acute liver failure
         (no underlying liver disease, ALT >3x ULN, INR > 2, not responsive to vitamin K)
      2. Acute hepatitis (ALT > 5 x ULN and duration of illness < 6 months)
         a. Infectious
         b. Drug induced liver disease
i. Suspected drug
   If 'Other', please specify
   ii. Was a liver biopsy done?
   If 'Yes', what were the results?
   If 'Other', please specify
   C. Other
   If 'Other', please specify
   D. Not known

Gut
6.11. Were there any Gut complications?
   If 'Yes', check all complications that apply.
   □ Diarrhoea
   □ Fatty liver disease
   □ Hepatitis
   □ Cholestatic
   □ Mixed
   □ Other
   □ Yes □ No

Kidney / Renal
   If 'Yes', check all complications that apply.
   □ Hypertension
   □ Kidney stones
   □ Acute kidney injury (plasma creatinine >150% of ULN for age; requiring dialysis / intensive monitoring)
   □ Chronic kidney disease (Chronic renal failure)
   □ Yes □ No

Tendon
6.13. Any tendon complications
   If 'Yes':
   1. Tendon rupture?
   2. Tendinitis?
   3. Other tendinopathy?
   □ Yes □ No

Other complications
6.14. Any other complications?
   □ Yes □ No
If 'Yes', check all complications that apply,

- Arthritis
- Arthropathy
- Allergic Bronchial Pulmonary Aspergillosis (ABPA)
- Asthma
- Bone fracture
- Depression
- Hearing loss
- Admission to intensive care unit
- Paediatric intensive care unit
- Nasal polyps
- Osteopenia
- Osteoporosis
- Pneumothorax requiring chest drain
- Port inserted or replaced in current 12 month period
- Sinus disease
- Absence of Yos deferens
- Other (please specify)

1. 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  
- Nasogastric  
- Jejunal tube  
- Yes but method unknown

7.5. Does the patient take pancreatic enzyme supplements?  
- Yes  
- No  
- Not known

7.6. Dose of Lipase  
- Not known  
- (u/l/kg per day)  
- Not applicable

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
   a. If 'Other', please specify

Exercise
8.3. Has an exercise test been performed?
   a. If 'Yes', check all that apply

Conti
8.4. Urinary incontinence
8.5. Faecal incontinence
8.6. Postural abnormality

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?

Education
9.3. Current education level of patient

Marital status
9.4. Patients marital status

Employment
9.5. What is the patient's employment status?
### 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?  
  - [ ] Not known

- **d.** Gestational age (weeks)  
  - [ ] Not known

- **e.** Congenital abnormality

  - i. If 'Yes' please specify

  - **a.** Down Syndrome  
    - Yes  
    - No

  - **ii.** Down Syndrome Karyotype
    - Karyotype confirmed  
    - Karyotype pending

- **b.** Suspected chromosomal disorder

  - i. Suspected chromosomal disorder Karyotype
    - Karyotype confirmed  
    - Karyotype pending
### Death

10.1. Has the patient died?
   - Date of death
   - Is date of death an estimate?
   - Cause of Death
     - If 'Cancer', please specify

Other

10.2. Diagnosis reversed?
   - Diagnosis reversal date
   - Reason for reversal of diagnosis?
     - If 'Other', please specify

### Transplants

10.3. Since the last annual review:
   - Has this patient been evaluated for transplant during the period since the last annual review?
     - What was the outcome of the transplant evaluation?
       - Acceptable
       - Declined
       - Deferred

   - Date placed onto transplant list
   - Received transplant since last annual review?
     - Transplant data
     - Transplant centre
     - Transplant type(s)
       - If 'Other', please specify

### Transplant Complications

10.4. Within 12 months of surgery, select any complications suffered
   - None
   - Bronchiolitis
   - Renal Failure
   - Other
   - Unknown

   - If 'Other', please specify