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

Oxygen and ventilaltion

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

a. When was oxygen therapy used?
- Continuously
- Nocturnal and/or with exertion
- During exacerbation
- PRN

b. Used non invasive ventillation? Yes No Not known

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

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

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)

- ☐ 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

- ☐ Pseudomonas aeruginosa

a. Number of Pseudomonas aeruginosa samples since last annual review

b. *Pseudomonas* mucoid status

- [ ] Mucoid
- [ ] Non mucoid
- [ ] Not known

c. *Pseudomonas* drug resistance

- [ ] Pseudomonas multi drug resistant
- [ ] Pseudomonas other resistance pattern
- [ ] Pseudomonas fully sensitive
- [ ] Pseudomonas ciprofloxacin resistant

d. Is Pseudomonas Chronic or Intermittent? ○ Chronic ○ Intermittent

2. Bukholderia Cepacia complex

- [ ] Bukholderia cepacia
- [ ] Bukholderia cenocepacia
- [ ] Bukholderia multivorans
- [ ] Other Bukholderia cepacia species

3. Fungal

- [ ] Aspergillus fumigatus
- [ ] Scedosporium species
- [ ] Aspergillus species
- [ ] Candida

4. Staphylococcus aureus

- [ ] Staphylococcus aureus

i. Is Staphylococcus aureus Chronic or Intermittent? ○ Chronic ○ Intermittent

5. Other Cultures

- [ ] Alcaligenes (Achromobacter) xylosoxidans
- [ ] Pseudomonas Species
- [ ] Escherichia Coli
- [ ] Haemophilus influenza
- [ ] Klebsiella
- [ ] MRSA
- [ ] Pandoraea
- [ ] Strenotrophomonas (Xanthomas) maltophilia
- [ ] Other

i. If 'Other', please specify

NTM

5.2.1. Has the patient been on treatment for NTM pulmonary disease at any time since last annual review? ○ Yes ○ No

Species

i. Start date

Species

i. Start date

b. Has the patient stopped all NTM antibiotic treatment? ○ Yes ○ No

i. Date of stopping treatment

ii. Reason for stopping
- ○ Completed treatment
- ○ Declined further treatment
- ○ Intolerant of treatment
- ○ Stopped treatment then later restarted it
- ○ Other

a. If 'Other' specify the reason

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

a. Negative Culture Result
- ○ Negative culture sample
- ○ Contaminated culture sample
- ○ No samples taken
- ○ Not known

5.2.3.

A. If yes, Date of Culture

B. Culture Type
- ○ Sputum
- ○ Induced Sputum
- ○ Lung Biopsy
- ○ Broncho-alveolar lavage
- ○ Not known

C. Species

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

a. Select reason why NTM treatment did not start
- ○ The patient is about to start NTM treatment
- ○ The patient declined NTM treatment
- ○ The clinical team did not think that NTM treatment was appropriate
- ○ Other

5.2.5. Was an intrevenous induction 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

5.2.9. Has the patient been on oral corticosteroid since the last data set? ○ Yes ○ No ○ Not known

5.2.10. Has the patient been on immunosuppressive drugs since the last data set? ○ Yes ○ No ○ Not known

COMPLICATIONS

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

6. 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

i. If 'Other', please specify

b. CFRD Treatment

i. Was patient prescribed treatment for CFRD? ○ Yes ○ No

If 'Yes',

- ☐ 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

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 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

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

Transplants

10.3 Since the last annual review:

a. Has this patient been evaluated for transplant during the period since the last annual review? ○ Yes ○ No

i. What was the outcome of the transplant evaluation?

- ○ Accepted
- ○ Declined
- ○ Deferred

a. Date placed onto transplant list

b. Received transplant since last annual review? ○ Yes ○ No

i. Transplant date

ii. Transplant centre

iii. Transplant type(s)

a. If 'Other', please specify

Transplant Complications

10.4 Within 12 months of surgery, select any complications suffered

- ☐ None
- ☐ LymphoProlifDisorder
- ☐ Atypical Infection
- ☐ Bronchiolitis
- ☐ Renal Failure
- ☐ Other
- ☐ Unknown

i. If 'Other', please specify,