Encounters Variables - Valid from 2023 Year of follow-up

Section	Sub-Section, Group	Variable	Mandatory in Encounter	Mandatory in Annual Summary
Date	N/A			
		– Name of Clinic	Yes (filled automatically)	No
		– Date of Encounter	Yes	No
		– Doctor (Doctor or user name):	Yes (filled automatically)	No
Status/Pregnancy/ Transplant	Status			
		– Has the patient died since the last encounter?	Yes (default *Alive).*	Yes
		– Date of death	Yes, if patient died	Yes
		– Cause of death	Yes, if patient died	Yes
	Pregnancy			
		– Is the patient pregnant / has the patient been	Only for female patients (if male,	Yes
		– Start date	Yes, if pregnant	No
		– Stop date	Yes, if pregnancy stopped	No
		– Reason for stopping	Yes, if pregnancy stopped	Yes
	Transplant			
		– Has the patient been assessed for, or had an active	No	No
		– Transplant type	No	Yes
		– Referred : awaiting assessment	No	No
		– Assessed : awaiting decision:	No	No
		– Decision made : unsuitable for listing:	No	No
		– Decision made : on active list:	No	No
		– Transplanted	No	Yes
		– Date of transplant	Yes, if transplanted	Yes (year latest Tx)
		– Re-transplant	No	Yes (if retransplant)

Section	Sub-Section, Group	Variable	Mandatory in Encounter	Mandatory in Annual Summary
Respiration and Anthropometry	**N/A**			
		– Weight	Yes (-1 needed if not measured)	Yes (-1 needed if not measured)
		– Height	Yes (-1 needed if not measured)	Yes (-1 needed if not measured)
		– FEV1	Yes (-1 needed if not measured)	Yes (-1 needed if not measured)
		– FVC	Yes (-1 needed if not measured)	Yes (-1 needed if not measured)
		– Date of FEV1 and/or height, weight measurement(s):	Yes, if any of the above measurements	Yes, if any of the above measurements
		– FEF 25 - 75:	No	No
		– Result of LCI 2.5%:	No, but recommended if measured	Yes, lowest value (-1 needed if not measured)
		– Date of LCI 2.5% measurement:	Yes, if value entered for LCI 2.5%	Yes, date of lowest value
		– Device used for LCI measurement:	Yes, if value entered for LCI 2.5%	Yes, if value recorded
		– Seen by physiotherapist:	No	No
		– Seen by psychologist	No	No
		– Seen by dietician	No	No
Regular medications For all regular medications	**N/A**			
		– Frequency	No	There are no dates in the Patient Annual Summary (PAS) but Encounter dates are used to calculate therapy duration. ≥ 3months continuous in Encounters = *Yes* in the PAS (but see definitions for Oxygen therapy and Inhaled antibiotics in the PAS). No therapy or < 3 months in Encounters = *No* in the PAS
		– Start date	Yes, if therapy selected	
		– End date	Yes, if stopped	
		– Continuing	Yes, if continuing	
	Antibiotics & Anti-fungals			
	Nebulised	– Nebulised Tobramycin	No, but if prescribed, should be added.	Yes - Inhaled antibiotics (nebulised and inhaled antibiotics from Encounters)
		– Nebulised Colomycin		
		– Nebulised Aztreonam		
		– Nebulised Levofloxacin		
	Inhaled	– Tobramycin dry powder inhaler		
		– Colomycin dry powder inhaler		
		– Nebulised other		
	Macrolides	– Azithromycin	No, but if prescribed, should be added.	Yes - Azithromycin or other macrolides
		– Clarithromycin		
		– Erythromycin		

Section	Sub-Section, Group	Variable	Mandatory in Encounter	Mandatory in Annual Summary
Regular medications - *cont.*	*(Antibiotics and anti-fungals, cont)* . Oral antibiotics	– Quinolone (Ciprofloxacin, Moxifloxacin etc.) – Sulphonamide (Trimethoprim, sulfamethoxazole) – Cephalosporin (Cephalexin, Cefuroxime etc.) – Penicillin (Amoxicillin, inc. with clavulanic acid) – Flucloxacillin etc. – Tetracycline (doxycycline, minocycline etc.) – Metronidazole – Clindamycin – Rifampicin – Sodium fusiadate – Chloramphenicol – Chronic oral antibiotics - other	No, but if prescribed, should be added.	No, oral antibiotics are not in the PAS
	Oral anti-fungals	– Itraconazole – Voriconazole – Posaconazole – Chronic oral anti-funglas- other	No, but if prescribed, should be added.	No, oran anti-fungals are not in the PAS
	Respiratory			
		– rhDNase	No, but if prescribed, should be added.	Yes - rhDNase
		– Hypertonic saline 3% – Hypertonic saline 6% – Hypertonic saline 7% – Hypertonic saline 11% – Hypertonic saline other	No, but if prescribed, should be added.	Yes, Hypertonic saline 3% and over
		– Mannitol	No, but if prescribed, should be added.	Yes
	Inhaled short-acting Bronchodilators	– Salbuterol – Terbutaline – Inhaled SA bronchodilators - other	No, but if prescribed, should be added.	Yes - inhaled bronchodilators, long-acting or short-acting
	Inhaled long-acting Bronchodilators	– Salmetorol – Inhaled LA bronchodilators - other	No, but if prescribed, should be added.	Yes - inhaled bronchodilators, long-acting or short-acting
	Inhaled Steroid alone	– Fluticasone – Budesonide – Beclomethasone – Inhaled steroid alone - other	No, but if prescribed, should be added.	Yes - inhaled steroids
	Inhaled Steroid with LABA	– Fluticasone - Salmeterol – Budesonide - Formoterol – Inhaled steroid with LABA - other	No, but if prescribed, should be added.	Yes - inhaled steroids / inhaled bronchodilators

Section	Sub-Section, Group	Variable	Mandatory in Encounter	Mandatory in Annual Summary
Regular medications - *cont.*	*Respiratory cont.*	– Tiotropium	No, but if prescribed, should be added.	No, LAMA are not in the PAS
	Inhaled LAMA	– Inhaled LAMA - other		
	Other	– Oral steroid	No, but if prescribed, should be added.	Yes
		– High dose Ibuprofen	No, but if prescribed, should be added.	No, Ibuprofen is not in the PAS
		– Leukotrien receptor antagonist	No, but if prescribed, should be added.	No, LRAs are not in the PAS
		– Oxygen therapy	No, but if prescribed, should be added.	Yes
	Non-invasive positive pressure ventilation (NIPPV)	– CPAP	No, but if prescribed, should be added.	Yes - NIPPV CPAP, BPAP
		– BPAP		
	Nutrition / GI			
		– Pancreatic enzyme replacement	No, but if prescribed, should be added.	Yes
		– Ursodeoxycholic acid	No, but if prescribed, should be added.	Yes
		– Proton pump inhibitors	No, but if prescribed, should be added.	Yes
		– H2 Blocker	No, but if prescribed, should be added.	No, H2 blockers are not in the PAS
	Enteral feeding	– PEG	No, but if prescribed, should be added.	No, Enteral feeding variables are not in the PA
		– NG		
	Vitamin Supplements	– Vitamin A	No, but if prescribed, should be added.	No, Vitamin supplements are not in the PAS
		– Vitamin D		
		– Vitamin E		
		– Vitamin K		
	Endocrine			
	Diabetes	– Insulin	No, but if prescribed, should be added.	Yes (see also Complications - below)
		– Oral hypoglycaemic agents		
	Other	– Hormonal contraception	No	No, none of these 4 variables are in the PA
		– Calcium supplementation	No	
		– Antidepressants	No	
		– Bisphosphonate	No	

Section	Sub-Section, Group	Variable	Mandatory in Encounter	Mandatory in Annual Summary
Regular medications - *cont.*	CFTR modulators			
	Sweat test	– Sweat chloride taken before start of therapy? – Date of sweat chloride before start – Value of sweat chloride before start – Sweat chloride taken during therapy – Date of sweat chloride during therapy – Value of sweat chloride during start	No, but if measured, should be added Yes, if measured Yes, if measured No, but if measured, should be added Yes, if measured Yes, if measured	Yes, Sweat chloride before start; Yes, lowest Sweat chloride during therapy; No - sweat test dates are not in the PAS
	Therapy	– Ivacaftor – Lumacaftor / Ivacaftor – Tezacaftor / Ivacaftor – Elexacaftor / Tezacaftor / Ivacaftor – Other	No, but if prescribed, should be added.	Yes, selection necessary for all types
	Dosage, frequency, dates	– Dosage – Frequency – Start date – Stop date – Continuing	No No Yes Yes, if stopped Yes, if continuing	No No Yes, start date is mandatory Yes, if stopped Yes (confirmed in PAS through *Issues*)
Pulmonary Exacerbations	N/A			
	Dates	– Start date – Stop date – Continuing	No, but PEx treated with IV antibiotics should be recorded (mandatory in PAS)	1. PEx treated with IV antibiotics 1. Yes, PEx treated with IV antibiotics; 2. Yes, number of PEx treated with IV antibiotics (n.b. NOT total duration of PEx episodes, but number of episodes).
	Treatment	– Treated with oral antibiotics – Treated with inhaled antibiotics – Treated with Intravenous antibiotics – Hospitalised	No, but recommended	
Acute Care	Oral antibiotic			
	Dates	– Start date – Stop date – Continuing	No, but recommended	No, oral antibiotics for acute care are not in the PAS
	Antibiotic name	– Flucloxacillin – Unknown – Other (type name if selected)	No, but recommended	

Section	Sub-Section, Group	Variable	Mandatory in Encounter	Mandatory in Annual Summary
Acute Care *- cont.*	**Inhaled antibiotic**			
	Dates	– Start date – Stop date – Continuing	No, but recommended	No, inhaled antibiotics for acute care are not in the PAS
	Antibiotic name	– Amikacin – Aztreonam – Colistin – Flucloxacillin – Tobramycin – Gentamicin – Vancoymycin – Unknown – Other (type name if selected)	No, but recommended	
	Intravenous antibiotic			
	Dates	– Start date – Stop date – Continuing	No, but recommended	Yes, number of days of IV antibiotics for CF-related reasons, at home or in hospital, is mandatory in the PAS.
	Antibiotic name	– Amikacin – Aztreonam – Cefoxitin – Ceftazidime – Colistin – Flucloxacillin – Fosfomycin – Tobramycin – Gentamicin – Meropenem – Teicoplanin – Vancoymycin – Cephradine – Unknown – Other – Other (type name if selected)	No	No, antibiotic name is not in the PAS

Section	Sub-Section, Group	Variable	Mandatory in Encounter	Mandatory in Annual Summary
Acute Care - Intravenous Antibiotic - *cont.*	Indication	– Pulmonary exacerbation – Eradication therapy – Other CF-related – Non CF-related – Unknown	No but recommended - number of days of IV antibiotics for CF-related reasons, at home or in hospital, is mandatory in the PAS.	Yes, number of days of IV antibiotics for CF-related reasons is mandatory in the PAS.
	Administered	– In hospital – At home		Yes, number of days of IV antibiotics for CF-related reasons at home and in hospital is mandatory in the PAS.
	Hospitalisation			
	Dates	– Date of admission – Date of discharge – Continuing	No, but recommended - total number of days in hospital for any reason is mandatory in the PAS.	Yes, total number of days in hospital for any reason is mandatory in the PAS.
	Reason	– CF pulmonary exacerbation – CF-related hemoptysis – CF-related distal intestinal obstruction – Elective surgery – CF-related pneumothorax – CF-related end stage lung disease – Other CF-related – Other non-CF-related – Unknown	No, but recommended - total number of days in hospital for any reason is mandatory in the PAS.	Yes, total number of days in hospital for any reason is mandatory in the PAS.
	Admittance	– Clinic – Emergency room – Unknown	No	No, not in the PAS
	Planned	– No – Unknown – Yes – No – Unknown – Yes	No	No, not in the PAS
	ICU	– ICU start date – ICU end date – Continuing	No, but recommended - total number of days in hospital for any reason is mandatory in the PAS.	Yes, total number of days in hospital for any reason is mandatory in the PAS, but ICU days not specified

Section	Sub-Section, Group	Variable	Mandatory in Encounter	Mandatory in Annual Summary
Microbiology	Sample			
		– Sample taken? If yes, select type – OP Swab – Spontaneous sputum – Induced sputum – Bronchoscopy – Pharyngeal aspiration – BAL - Bronchoalveolar lavage – Other – Sample date	No	No, type and date of sample are not in the PAS
	Pathogens			
	Bacteria	– Has the patient tested positive for the following? – (Options: *No, Yes, Unknown, Pending results*) – Mucoid Pseudomonas aeruginosa – Non-mucoid pseudomonas aeruginosa – Pseudomonas aeruginosa mucoid unknown – MSSA - Methicillin sensitive Staphlycoccus aureus – MRSA - Methicillin resistant Staphlycoccus aureus – Burkholderia cepacia complex – Stenotrophomonas maltophilia – Achromobacter spp – Haemophilus influenzae (any species)	No, but should be recorded if tested positive (mandatory in PAS). No data recorded for a pathogen in any encounter in the YFU = no infection in the PAS; It is possible to indicate chronicity in the Encounter, even if no/insufficient samples. This is done with a tick box. This will be copied to the PAS.	Mandatory for all pathogens except other bacteria or fungus; It is possible to indicate chronicity in the Encounter, even if no/insufficient samples. This is done with a tick box. This will be copied to the PAS.
	Other	– Other bacteria (not NTM) – Text box for Other	No	No
	Non-tuberculous mycobacteria	– Was sputum/BAL cultured for NTM	No but strongly recommended	Yes
		– (Options: *No, Yes, Unknown)*		
		– Non-tuberculous mycobacteria detected?	Yes, if *Yes* to main question	Yes
		– If detected, Select (more than 1 possible) from:		
		– Mycobacterium avium complex	Yes, if NTM detected	Yes
		– Mycobacterium abscessus complex	Yes, if NTM detected	Yes
		– Other	Yes, if NTM detected	Yes

Section	Sub-Section, Group	Variable	Mandatory in Encounter	Mandatory in Annual Summary
Microbiology - cont.	Fungi	– Sputum/BAL/Throat swab investigated for fungi?	No but strongly recommended	Yes
		– (Options: *No, Yes, Unknown)*		
		– Fungi detected?	Yes, if *Yes* to main question	Yes
		– If detected, Select (more than 1 possible) from:		
		– Aspergillus fumigatus	Yes, if fungi detected	Yes
		– Scedosporium species	Yes, if fungi detected	Yes
		– Other	Yes, if fungi detected	Yes
		– Text box for Other	No	No
Complications of CF	N/A			
		– Options: *No (or not done), Yes, Unknown.* – For some complications, if Yes is selected, additional questions must be answered.		
	ABPA	– Allergic Bronchopulmonary Aspergillosis (ABPA) – If *Yes* to ABPA, select from: – ABPA or – ABPA requiring treatment	No, but recommended	Yes
	Diabetes	– CF-related diabetes – If *Yes* to CF-related diabetes, select from: – Treated with daily insulin – Treated with oral hypoglycemic agents – Only dietary advice – Therapy unknown	No, but recommended	Yes
		– DIOS (distal intestinal obstruction syndrome)	No, but recommended	Yes
	Glucose tolerance test	– Glucose tolerance test done? – If Yes to glucose tolerance test, select from: – Normal – Impaired glucose tolerance – Impaired fasting hyperglycemia – Diabetic – Other	No, but recommended	Glucose tolerance test is not in the PAS but *Diabetes* is.
	Pneumothorax	– Pneumothorax – If pneumothorax *Yes* , select from: – Chest drain – Observation only – Therapy unknown	No, but recommended	Yes

Section	Sub-Section, Group	Variable	Mandatory in Encounter	Mandatory in Annual Summary
Complications of CF - *cont.*	Liver disease	– Liver - GI disease	No, but recommended	Yes
		– If *Yes* to Liver - GI disease, select from:		
		– Cirrhosis with portal hypertension/hypersplenism		
		– Cirrhosis without portal hypertension/hypersplenism		
		– Cirrhosis portal hypertension unknown		
		– Liver disease without cirrhosis		
		– Variaceal bleeding		
		– Major hemoptysis ≥ 250ml/day since last encounter:	No, but recommended	Yes
		– Is the patient being treated for NTM?	No, but recommended	Yes
	Pancreatic status	– Pancreatic status: faecal elastase	No, but recommended	Yes for both faecal elastase and faecal fat.
		– < 200µg		
		– ≥ 200µg		
		– Pancreatic status: faecal fat		
		– High		
		– Normal		
		– Pancreatitis	No, but recommended	No, Pancreatitis is not in the PAS
	Malignancy	– Malignancy diagnosed since last encounter?	No, but recommended	Yes
		– If yes to malignancy, select from:		
		– Colorectal cancer		
		– Small bowel cancer		
		– Lymphoid leukaemia		
		– Testicular cancer		
		– Breast cancer		
		– Thyroid cancer		
		– Type unknown		
		– Other		
		– Sinusitis	No, but recommended	No
		– Osteoporosis	No, but recommended	No
		– Osteopenia	No, but recommended	No
		– Gastroesophageal reflux	No, but recommended	No
		– Arthritis	No, but recommended	No
		– Anxiety	No, but recommended	No
		– Depression	No, but recommended	No
		– Salt loss syndrome	No, but recommended	Yes

Section	Sub-Section, Group	Variable	Mandatory in Encounter	Mandatory in Annual Summary
Complications of CF - *cont.*	Comorbidities	– Comorbidities status – If yes to comorbidities, tick box for following: – Cardiac arrhythmias – Cardiovascular disease (including heart failure) – Cholesterolemia – Chronic pain – Constipation – Eye disease – Cataract – Gall stone – Hearing loss – Arterial hypertension with cholesterolemia – Arterial hypertension without cholesterolemia – Arterial hypertension, cholesterolemia unclear – Infertility – Micro gall bladder – Nasal polyp – Rectal prolapse – Renal disease – Nephrolithiasis – Renal disease - proteinuria – Renal disease - proteinuria requiring dialysis – Renal disease - GFR < 50% – Renal disease - microalbuminuria – Renal disease - requiring dialysis – Splenomegaly – Tinnitus – Other	No	No
Liver Chemistries	N/A			
		– Date of lab sample DDMMYYYY – ALT (alanine transaminase) result – AST (aspartate transaminase) result – GGT (gamma-glutamyl transferase) result – ALP (alkaline phosphatase) result – Bilirubin result	No	No

Annual Summary Variables - Included for Information

Demographics

CF centre code;
Centre Patient code (optional);
Year of follow-up;
Year and month of birth;
Sex (Previously "Gender") - Male, Female, Other/Prefer not to say;
Ethnicity;
Vital Status of patient;
Cause of death;
Date of death.

Diagnosis

Age at diagnosis;
Sweat test type and values (x2);
First & second variants in trans (possible to record complex variants in cis);
Meconium Ileus;
Neonatal screening;
Nasal Potential Difference Measured? (NPD);
CF-typical NPD measurement Yes/No;
Date of NPD measurement;
Intestinal current value measured? (ICM);
CF-typical IC measurement Yes/No;
Date of IC measurement.

Complications

ABPA (Allergic bronchopulmonary aspergillosis)*
Distal intestinal obstruction syndrome (DIOS) this year;
Salt loss syndrome this year;
Diabetes this year;
Pneumothorax this year;
Liver disease this year;
Haemoptysis major volume of expectorate > 250ml in a day;
Occurrence of malignancy - diagnosed this year;
Pancreatic status: faecal elastase;
Pancreatic status: faecal fat;
Pregnancy this year;
Pregnancy stopped this year - reason for stop;
Pregnancy ongoing at 31/12.

Maintenance Therapy

Inhaled continuous (≥ 3 months) hypertonic saline ≥3% ;
Inhaled continuous (≥ 3 months) Mannitol;
Inhaled antibiotic this year - continuous (≥ 3 months) or on/off for a total of (≥ 6months);
Inhaled continuous (≥ 3 months) bronchodilators, long-acting or short-acting or both;
Oxygen therapy ≥ 3 months during the year of follow-up (inc. 24h/day, night time, exercise).
Does not need to be continuously but should be from a single prescription);
Use of continuous (≥ 3 months) non-invasive positive pressure ventilation (NIPPV);
Use of continuous (≥ 3 months) rhDNase this year;
Use of continuous (≥ 3 months) Inhaled steroids;
Use of continuous (≥ 3 months) Oral steroids;
Use of continuous (≥ 3 months) azithromycin (or other macrolide) this year;
Use of continuous (≥ 3 months) ursodeoxycholic acid this year;
Use of continuous (≥ 3 months) pancreatic enzymes this year;
Use of continuous (≥ 3 months) proton pump inhibitors (PPI);

** Originally removed from the variables collected from 2022 onwards but reinstated in January 2023 at start of the 2022 data collection period (as requested by various members of the ECFSPR Steering Commmittee*

Use of CFTR Modulator Therapy (data for each of the following are collected: Ivacaftor, Lumacaftor/ Ivacaftor, Tezacaftor/Ivacaftor, Elexacaftor/Tezacaftor/Ivacaftor, Other CFTR modulator;
Start and stop dates x 2 for each CFTR modulator;
Sweat chloride values - before start and during (lowest of the year) for each CFTR modulator.

Lung function and nutrition follow-up

Value of FEV1 in litres of highest FEV1% predicted of the year;
Value of FVC in litres (from same spirometry as recorded FEV1);
Height measured at date of best FEV1 (or if no available FEV1, last height of the year);
Weight measured at date of best FEV1 (or if no available FEV1, last weight of the year);
Date of recorded FEV1 or if no FEV1 recorded, date of recorded height and weight);
Lowest LCI 2.5% of the year;
Type of device used for LCI measurement;
Date of lowest LCI 2.5% this year;
Value of lowest LCI 2.5% this year.

Microbiology (*positive-chronic* or *positive-at least once/not chronic* for all pathogens)

Pseudomonas aeruginosa;
MSSA - Methicillin-sensitive Staphylococcus aureus;
MRSA - Methicillin-resistant Staphylococcus aureus;
Burkholderia cepacia complex;
Stenotrophomonas maltophilia;
Achromobacter spp;
Haemophilus influenza;
Nontuberculous mycobacteria cultured;
Mycobacterium abscessus complex;
Mycobacterium avium complex;
Other mycobacteria;
Nontuberculous mycobacteria treated this year;
Fungi investigated;
Aspergillus fumigatus;
Scedosporium spp.

Hospitalisation, Pulmonary Exacerbations, IV Antibiotics

Total days on iv antibiotics at home and in hospital this year (CF-related reasons);
Total days on iv antibiotics in hospital this year (CF-related reasons);
Total days in hospital this year (any reason);
PEx treated with intravenous antibiotics yes/no etc.
Number of PExs treated with intravenous antibiotics during the year.

Transplant

Liver transplant at any time;
Year of latest liver transplant (before or during this year);
Lung transplant at any time;
Year of latest lung transplant (before or during this year);
Kidney transplant at any time;
Year of latest lung transplant (before or during this year);
Other transplant at any time;

Year of latest other transplant (before or during this year).

End

CRITERIA AND REFERENCES USED BY THE ECFS PATIENT REGISTRY

1 Diagnosis / Reversal of Diagnosis: Criteria, References

A. Diagnosis accepted if:

Two sweat tests value > 59 mmol/L chloride;
OR

ii. One sweat test value > 59 mmol/L chloride + DNA Analysis/Genotyping – two identified disease-causing CF variants in trans
OR

iii. Sweat chloride value is less than or equal to 60 mmol/L or not reported **AND at least 2 of the following:**

a. DNA Analysis/Genotyping: two identified disease-causing CF variants in trans;
b. NPD (Transepithelial (Nasal) Potential Difference) or ICM (Intestinal current measurement): result consistent with a diagnosis of CF;
c. Clinical presentation: typical features of CF.

B. Diagnosis reversal* - CF diagnosis should be reversed if any of the following is true:

i. DNA Analysis has not identified any CF disease-causing variants;
ii. NPD (nasal potential difference) &/or ICM (intestinal current measurement): result not consistent with diagnosis of CF;
iii. Normal values from repeated sweat testing when (never prescribed CFTR modulators & confirm with the clinical team).

**See also ECFSPR SOP (Standard Operating Procedure) 214 Reversal of Diagnosis - regarding reversal of diagnosis and previously submitted data (find it on the homepage of the data collection software ECFSTracker, together with other useful information).*

References

1) ECFS best practice guidelines: the 2018 revision
2) European Cystic Fibrosis Society Standards of Care: Best Practice guidelines (2013)

2 Demographics: Ethnicity, References

i. Ethnicity categories are based on the available Global Lung Function Initiative equations and are: Caucasian; Black; North East Asian; South East Asian; Mixed / Other (including Indian sub-continent); Unknown; Missing.

ii. **Group country/region**

Caucasian: Europe, Israel, Australia, USA, Canada, Mexican Americans, Brazil, Chile, Mexico, Uruguay, Venezuela, Algeria, Tunisia;
Black: African-American;
South East Asian: Thailand, Taiwan and China (including Hong Kong) south of the Huaihe River and Qinling Mountains;
North East Asian: Korea and China north of the Huaihe River and Qinling Mountains;
Mixed / Other: Indian Sub-continent and individuals not represented by one of the groups or who are of mixed ethnicity.

iii. In addition to the countries listed, it is reasonable to apply the same ethnicity to others with geographic or ethnic proximity. Thus the Caucasian equations could be used for any person having origins in any of the original peoples of Europe, the Middle East or North Africa and for any non-indigenous person of South America. Similarly, the South East Asian equations may be reasonably extended throughout that region.

References

1) Global Lung Function Initiative equations described by Quanjer PH et al. (Multi-ethnic reference values for spirometry for the 3-95-yr age range: the global lung function 2012 equations. Eur Respir J 2012; 40: 1324–1343).
https://erj.ersjournals.com/content/40/6/1324

3 Sweat Test: Parameters, Values to be reported, References

i. Diagnostic standards: the quantity of sweat should indicate an adequate rate of sweat production;

ii. a. The sweat sample should be processed immediately after sweat collection;
b. Chloride concentration measurement is the preferred analysis for Diagnostic sweat tests. **For sweat tests in relation to CFTR modulator therapy, Chloride is the only accepted value;**
c. Chloride value: report the Chloride value in millimols per litre (mmol/L). If duplicate tests were completed on the same day, for Diagnostic sweat tests, **report the highest positive value;**
d. A sweat chloride value >59 mmol/L is consistent with a diagnosis of CF;
e. A sweat chloride value <30 mmol/L makes the diagnosis of CF unlikely (However, specific CF causing mutations can be associated with a sweat test below 30 mmol/L).
n.b. *The acceptable range for Chloride values is 1-160 mmol/L.* ***Anyone who has a Chloride value above 160 mmol/L should be re-tested;***

iii. As already mentioned above, the ECFSPR will consider only Titration/Chloride values in analyses.

References (links)

1) ECFS best practice guidelines: the 2018 revision
2) European Cystic Fibrosis Society Standards of Care: Best Practice guidelines (2013)

CRITERIA AND REFERENCES USED BY THE ECFS PATIENT REGISTRY

4 Nutrition: Method, Values and Dates to be reported, References.

i. The height and weight reported to the ECFSPR should be from the same day as the reported FEV1 (of hightest FEV1% predicted of the year);
If spirometry was not done the last weight and height measurements of the year, and the date they were measured, should be recorded.

ii. If spirometry was not done, the last weight and height measurements of the year, and the date they were measured, should be recorded.

iii. Height and weight should be measured in accordance with EuroCareCF guidelines:
Weight: removal of outer clothing, shoes and socks;
Height: removal of shoes and socks, stadiometer - top of head in contact with head board, slight pressure.

iv. Z-scores for height, weight & BMI are calculated with the CDC reference values [Kuczmarski et al (2002)].

References

1) Kromeyer-Hauschild K, Wabitsch M, Kunze D, Geller F, Geiss HC, Hesse V et al. Percentiles of body mass index in children and adolescents evaluated from different regional German studies. Monatsschr Kinderheilkd 2001; 149:807-818.
2) Lai H-C, Corey M, FitzSimmons S, Kosorok MR, Farrell M. Comparision of growth status of patients with cystic fibrosis between the United States and Canada. Am J Clin Nutr 1999; 69:531-538.
3) Public Use File BGS98, German National Health Interview and Examination Survey 1998, Robert-Koch-Institut, Berlin, Germany, 2000.
4) Wiedemann B, Paul KD, Stern M, Wagner TO, Hirche TO, on behalf of the German CFQA Group. Evaluation of body mass index percentiles for assessment of malnutrition in children with cystic fibrosis. Eur J Clin Nutr 2007; 61, 759-768.
5) Kuczmarski RJ, Ogden CL, Guo SS et al. 2000 CDC Growth Charts for the United States: methods and development. Vital Health Stat 2002; 11(246): 1-190.

5 Spirometry: Criteria, Method, Values to be reported, References.

- The ECFS Patient Registry collects data on spirometry values to obtain standardised data for comparison with other centres/countries and for use in specific epidemiological studies.
- **n.b.** Some of the conditions for this (see below) may not be met at every clinical visit for all patients and, for the ECFSPR, only spirometry tests fulfilling the criteria should be recorded by centres or submitted by the National Registries.
We recommend a**ll spirometry tests are carried out in accordance with the ATS/ERS guidelines** (www.thoracic.org/statements/resources/pfet/PFT2.pdf) although
- a decision was taken by the ECFSPR Definitions group in 2022 to accept lung function values from spirometry tests carried out also post-bronchodilator.*

For the spirometry values reported to the ECFSPR the following criteria should be met:

i. **Pre-test preparation**
a. All recorded spirometry tests should be pre-bronchodilator (but see above*);
i. short-acting bronchodilators: at least 4 hours pre-test;
ii. long-acting bronchodilators: at least 12 hours pre-test.
b. Date of birth, gender and height should be recorded for calculation of predicted values. In addition, the ECFS Patient Registry asks for the weight and height to be measured at the same time, and recorded.

ii. **Values to report:**
a. FEV1 in litres of best FEV1% predicted, pre- or post-bronchodilator (to 2 decimals), in accordance with local reference values;
b. The FEV1 and FVC measurements must be reported in litres (L), to max 2 decimal points;
c. FVC in litres: must be the FVC measured from the same test as recorded FEV1 (of the best FEV1% predicted of the year, pre- or post-bronchodilator) and it must be greater than or equal to the FEV1 measurement.
d. For the reported spirometry values, the date of the test and the patient's height and weight at that date should also be recorded in order to calculate the percent of predicted values and other values;
e. Only tests deemed valid according to ATS/ERS guidelines to be reported.

iii. **Calculation of percent of predicted values:**
a. A common set of reference values - the Global Lung Function Initiative equations (See (1) below) - is used for calculations.

References

1) Global Lung Function Initiative equations described by Quanjer PH et al. (Multi-ethnic reference values for spirometry for the 3-95-yr age range: the global lung function 2012 equations. Eur Respir J 2012; 40: 1324–1343).
2) Miller et al. Standardisation of spirometry. Eur Respir J 2005; 26: 319–338 / Graham et al. Standardisation of spirometry 2019 update. Am J Respir Crit Care Med.2019 Oct 15;200(8):e70-e88.
3) Miller et al. General considerations for lung function testing. Eur Respir J 2005; 26: 153–161.
4) Rosenfeld et al. Task Force to Evaluate Choice of Spirometric Reference Equations for the National Patient Registry: Summary and Recommendations. Cystic Fibrosis Foundation Registry Committee; 2005.

CRITERIA AND REFERENCES USED BY THE ECFS PATIENT REGISTRY

6 Chronic infection in the lower airways: Definition, References.

i. Chronic Pseudomonas aeruginosa infection: A patient should be considered chronically infected if the modified Leeds criteria are met - (a) below - and/or anti-pseudomonas antibodies are detected - (b) below.

ii. A patient should be defined as chronically infected if he/she fulfils the criteria now, or has done so in recent years, and the physician has no reason to think that the status has changed.

a. Modified Leeds criteria - chronic infection: >50% of the samples (sputum/other) collected during the last 12 months should be positive; at least 4 samples collected.
b. Significantly raised levels of anti-pseudomonas antibodies according to local laboratories.

iii. Chronic infection with other gram-negative or gram-positive bacteria should meet the same criteria as described above.

References

1) Lee TWR, Brownlee KG, Conway SP, Denton M, Littlewood JM. Evaluation of a new definition for chronic Pseudomonas aeruginosa in cystic fibrosis patients. J Cystic Fibrosis
2) Proesmans M, Balinska-Miskiewiscz, Dupont L et al. Evaluating the "Leeds criteria" for Pseudomonas aeruginosa infection in a cystic fibrosis centre. Eur Resp J 2006;27:937-943.
3) Doring G, Conway SP, Heijerman HG, et al. Antibiotic therapy against Pseudomonas aeruginosa in cystic fibrosis: a European consensus. Eur Respir J 2000;16:749-767.

7 Allergic Broncho-pulmonary aspergillus - ABPA

i. Diagnostic criteria:

a. Acute or subacute clinical deterioration (cough, wheeze, exercise intolerance, exercise-induced asthma, change in pulmonary function, or increased sputum production) not attributable to another etiology.
b. Total IgE > 500 IU/ml.
c. Positive skin prick test for Aspergillus antigen (> 3 mm) or positive specific IgE for A. fumigatus.
d. Precipitins to A. fumigatus or in vitro demonstration of IgG antibody to A. fumigatus;
OR
new or recent abnormalities on chest radiography (infiltrates or mucus plugging) or chest CT (characteristic changes) that have not cleared with antibiotics and standard physiotherapy.

References

Allergic bronchopulmonary aspergillosis in cystic fibrosis--state of the art: Cystic Fibrosis Foundation Consensus Conference. Stevens DA, Moss RB, Kurup VP, Knutsen AP, Greenberger P, Judson MA, Denning DW, Crameri R, Brody AS, Light M, Skov M, Maish W, Mastella G; Participants in the Cystic Fibrosis Foundation Consensus Conference. Clin Infect Dis. 2003 Oct 1;37 Suppl 3:S225-64.

8 Liver Disease: Definitions

The ECFSPR has adopted the definitions for Liver Disease used by the Cystic Fibrosis Registry in the UK. These definitions discriminate patients with severe liver disease (with portal hypertension) from milder cases (cirrhosis without portal hypertension).

- **Cirrhosis with Hypertension**: scarring of the liver related to underlying CF, typically in a biliary pattern. Severe liver disease may include portal hypertension and/or hypersplenism;
- **Cirrhosis without Hypertension**: scarring of the liver related to underlying CF;
- **Liver disease without cirrhosis**: this includes fatty liver or viral hepatitis but not biliary cirrhosis.

CRITERIA AND REFERENCES USED BY THE ECFS PATIENT REGISTRY

9 Pancreatic Status: Pancreatic Insufficiency, References

i. **Indicator of Pancreatic Insufficiency - Faecal Fat** (2 determinations are mandatory)

a. Young children: Stool fat (van de Kamer) > 4-5 g/d;

b. Children older than 10 years and adults: Stool fat (van de Kamer) >7g/d and/or faecal pancreatic elastase-1 < 200 ug/g.

Please note

- Faecal fat excretion values of infants below 3 months are contradictory.
- Other than pancreatic causes of steatorrhoea must have been excluded.

ii. **For the ECFSPR, pancreatic status will be assessed as follows:**

- Pancreatic insufficiency: Faecal elastase <200 µg/g (twice), and faecal fat high* (twice);
- Pancreatic sufficiency: Faecal elastase ≥200 µg/g (twice) and Faecal fat normal* (twice).

**see no. 1 Definition, above*

References

1) Sinaasappel M, Stern M, Littlewood J, Wolfe S, Steinkamp G, Heijerman HGM, Robberecht E, Döring G. Nutrition in patients with cystic fibrosis. A European consensus. J Cystic Fibrosis 2002; 1:51-75.
2) Walkowiak J, Nousia-Arvanitakis S, Henker J, Stern M, Sinaasappel M, Dodge JA. Invited review: Indirect pancreatic function tests in children. J Pediatr Gastroenterol Nutr 2005; 40:107-114.

10 Salt Loss Syndrome: Definition and Reference

All 3 of the following to be present:

'- Primary metabolic alkalosis with blood pH > 7.45;
- Serum sodium < 130 mmol/l;
- Serum chloride < 90 mmol/l.

Reference

1) Fustik S, Pop-Jordanova N, Slaveska N, Koceva S, Efremov G. Metabolic alkalosis with hypoelectrolytemia in infants with cystic fibrosis. Pediatr int 2002; 44: 289-92.

11 Transplantation: Indications

i. For patients who had a transplant during the year of follow up:

a. Use the best FEV1 before transplantation;

b. Record therapy, complications and microbiology from before transplantation.

ii. For patients who had a transplant before the current follow-up year:

a. Record all available information.