For Centres that Input Data in ECFSTracker

Variables, definitions and inclusion criteria used by ECFSPR

Part 1 Variables: name; description; input ECFSTracker

i. **Core Data**: Demographics & Diagnosis Data (input only once, at enrolment).

ii. **Annual Data** (added to PAS*): Therapy, Nutrition, Lung Function, Microbiology, Hospitalisation, Complications, Transplant
*PAS: Patient Annual Summary submitted to the ECFSPR, Core Data + Annual Data

Part 2 Criteria, Definitions and References

i Diagnostic inclusion criteria; Reversal of diagnosis.

ii. Sweat Test: parameters; values to be reported.

iii. Nutrition: method; values to be reported; references.

iv. Spirometry: criteria; method; values to be reported; references.

v. Lung Clearance Index 2.5%: references

v. Chronic infection in the lower airways: definition; references.

vi ABPA: diagnositic criteria; references

vii Liver Disease: definitions.

viii Pancreatic status: criteria; definitions; (faecal fat & elastase, salt-loss syndrome)

ix. Salt-loss syndrome: definition; reference

x. Transplantation: what to report.

Variable	Description	Input ECFSTracker
Part 1.i. Core Data variables (new in blue)		
Centre Code (only the Service Desk, Country Coordinator and authorised centre users know both centre name and centre code)	Unique 4 digit code randomly created and assigned by ECFSTracker at centre set up; Represents the centre in the Registry; When the Annual Summary data is submitted to the Registry it is submitted under this number. The centre name is never transmitted.	Does not need to be input by the user.
Patient Code (ECFSPR ID)	Unique 6 digit code randomly assigned by ECFSTracker when a patient is included in the Annual Summary for the first time; If transferred to another centre the patient retains this number; When PAS* (Patient Annual Summary) is submitted to the Registry it is submitted under this number.	Is never input by the user.
Year of follow-up	The year of follow-up runs from 1 January to 31 December of the year that you are providing data for.	The follow-up year is selected by the user and "initialised" to allow annual data input.
Date of birth	You record the date of birth for the patient when you enrol him/her in the Registry. In the PAS* - Patient Annual Summary - submitted to the Registry, the day of birth for all patients is 15.	You only input the date of birth once at enrolment. It is copied (15/mm/yyyy) into the PAS* when you initialise the year.
Gender	Biological gender: when you enrol the patient we ask you to indicate if the patient is male or female.	You only input the gender once. It is copied into the PAS* when you initialise the year.
Status of patient	The Registry records the vital status of the patient at 31/12 of the year of follow up. Options: Unknown; Missing; Alive; Not seen during the year of follow-up; Lost to follow-up (recommended for 3rd year not seen and if no knowledge of status or whereabouts of patient); Died.	Select from options provided. n.b. If the patient no longer attends your centre but you think he/she attends another centre, inform the Service Desk, and do not use the options "not seen during the year of follow-up" or lost to follow-up; See Appendix 1. ii - "Patient Transfer".
Date of death	If the patient died during the year of follow up the date of death is required (if known).	Type or select from calendar.

Variable	Description	Input ECFSTracker
Cause of death	If the patient died, a cause of death is required, even if the cause is unknown. Options: Respiratory; Liver /GI (GI - Gastrointestinal); Trauma; Suicide; Transplantation; Non CF-related; **Cancer;** **Other CF-related;** Unknown; Missing/Not collected.	Select from options provided.
Diagnosis Confirmed	You must indicate if the patient has a confirmed CF diagnosis. See Part 2.i. ECFSPR Inclusion Criteria Options: No; Diagnosis to be confirmed; Yes; Missing.	Copied to the (PAS*) when you initialise the year; n.b. Only patients with a confirmed CF diagnosis are included in data analyses. We recommend you do not enrol patients until diagnosis is confirmed.
Age at diagnosis	In the Core Data, age (in decimal years, to max 2 decimals e.g 2.25 = 2 years and 3 months) or date of diagnosis is required.	The age at diagnosis is copied to the (PAS*) when you initialise the year; if you input a date of diagnosis in the Core Data, ECFSTracker calculates the age.
Neonatal screening	If neonatal screening was carried out, and the outcome, are recorded in the Core Data. Options: Unknown; Missing; Not done; Yes; If "Yes": performed, result positive; performed, result negative; performed, result unknown.	The neonatal screening result is copied to the (PAS*) when you initialise the year.

Variable	Description	Input ECFSTracker
Meconium Ileus	If the patient had Meconium Ileus at birth, this is recorded in the Core Data. Options: No; Unknown; Missing; Yes; If "Yes": Yes, operated; Yes, not operated; Yes, don't know if operated	The result is copied to the patient annual summary (PAS*) when you initialise the year.
Mutations	The mutations identified from genotyping, if any, are recorded in the Core Data. The legacy name (CFTR1&2 databases) is preferred. Options: Not done / missing; Unknown / Unidentified; Select mutation from list; If the mutation is not on the list you can type it;	The results are copied to the patient annual summary (PAS*) when you initialise the year.
Special case	More than 1 mutation on the same allele	Select "other (not in list)" then type the mutations in the box, separated by a semi-colon (;) e.g. Mut1Ab; Mut2Cd; Mut3Ef etc.
Date of genotyping results	Record the date of the mutation result in the Core Data, if you wish. It is not obligatory.	The mutation result date is not included in the PAS* submitted to the Registry.
Sweat Tests - Sweat Test 1	If sweat tests have been done indicate the sweat test type and results in the Core Data. You can record outcomes for 2 sweat tests. See Part 2.ii Sweat Test Options: Not done; Unknown; Missing; Titration; If Titration, type of electrolytes: Chloride; Other; Unknown Conductivity. Test result: enter value in mmol/L.	The Sweat test data is copied to the patient annual summary (PAS*) when you initialise the year. n.b. For the Registry, Titration/Chloride is preferred.

Variable	Description	Input ECFSTracker
NPD **Nasal Potential Difference**	If nasal potential difference has been measured you record the information in the Core Data; If NPD has been measured you must indicate if the result is CF-typical or not. Options: No; Unknown; Missing; Yes; If "Yes", is the value CF-typical?: No; Unknown; Yes; Indeterminate.	The NPD data is copied to the (PAS*) when you initialise the year. NPD measurement is rarely done. In the Core Data the default result is "missing". Don't change this unless you can do NPD measurements in your centre;
Date of NPD	If NPD has been measured, record the date of the NPD measurment in the Core Data.	The NPD date is copied to the patient annual summary (PAS*) when you initialise the year.
ICM **Intestinal current measurement**	If intestinal current measurements have been taken you can record the outcome in the Core Data; If IC has been measured you must indicate if the result is CF-typical or not. Options: No; Unknown; Missing; Yes; If "Yes", is the value CF-typical?: No; Unknown; Yes; Indeterminate.	The ICM data is copied to the (PAS*) when you initialise the year. ICM measurement is rarely done. In the Core Data the default result is "missing". Don't change this unless you do ICM measurements in your centre;
Date of ICM	If intestinal current has been measured record the date of measurement in the Core Data	The ICM date is copied to the (PAS*) when you initialise the year.

END OF CORE DATA VARIABLES

Variable	Description	Input ECFSTracker
Part 1.ii. Annual Summary variables (new in blue)		
Therapy Some therapies have **"continuous"** in the variable name. For the ECFSPR this means "*at least 3 consecutive months during the year of follow-up".* If "continuous" is not in the variable name, for the ECFSPR it means "*at any time during the year of follow-up* ".		
Continuous inhaled hypertonic NaCl	Did the patient take continuous hypertonic saline (any concentration) during the follow up year? Options: No; Unknown; Missing; Yes.	Select from options provided.
Continuous inhaled Mannitol	Did the patient take continuous inhaled Mannitol during the follow up year? Options: No; Unknown; Missing; Yes.	Select from options provided.
Continuous inhaled antibiotic	Did the patient take continuous inhaled antibiotics during the follow up year? Options: No; Unknown; Missing; Yes.	Select from options provided.
Continuous inhaled bronchodilators	Did the patient take continuous inhaled bronchodilators during the follow up year? Options: No; Unknown; Missing; Yes.	Select from options provided.
Oxygen therapy	Did the patient have oxygen therapy at any point during the year? Options: No; Unknown; Missing; Yes.	Select from options provided.

Variable	Description	Input ECFSTracker
NIPPV: continuous non-invasive positive pressure ventilation	NIPPV - Ventilation support only, as a marker of correct treatment of complete respiratory insufficiency. Did the patient have continuous NIPPV therapy during the year of follow-up? If yes, which kind? Options: No; Unknown; Missing; CPAP - continuous positive air pressure; BPAP (also BiPap) bilevel positive air pressure.	Select from options provided.
Use of rhDNase	Did the patient use rhDNase at any time during the year? Options: No; Unknown; Missing; Yes.	Select from options provided.
Continuous inhaled steroids	Was the patient taking inhaled steroids continuously during the follow-up year? Options: No; Unknown; Missing; Yes.	Select from options provided.
Continuous oral steroids	Was the patient taking oral steroids continuously during the follow-up year? Options: No; Unknown; Missing; Yes.	Select from options provided.
Continuous Azithromycin or other macrolide	Did the patient take azithromycin or other macrolides continuously (somminstration of any type) during the follow-up year? Options: No; Unknown; Missing; Yes.	Select from options provided.

Variable	Description	Input ECFSTracker
Use of ursodeoxycholic acid	Did the patient use ursodeoxycholic acid at any point during the year? Options: No; Unknown; Missing; Yes.	Select from options provided.
Pancreatic enzymes	Did the patient take pancreatic enzymes during the year? Options: No; Unknown; Missing; Yes.	Select from options provided.
Continuous proton pump inhibitors	Did the patient take protein pump inhibitors continuously during the year? Options: No; Unknown; Missing; Yes.	Select from options provided.
CFTR modulator therapy	Did the patient take CFTR modulators during the year of follow-up? If yes, which? Options: No; Unknown; Missing; Yes. If yes: Ivacaftor; Ivacaftor / Lumacaftor; Ivacaftor / Tezacaftor; Other.	Select from options provided.
Nutrition and Lung Function		
Weight	Weight measured at date of best FEV1 or, if no lung function test, last previously recorded weight of the year (removal of outer clothing, shoes and socks); See also Part 2.iii. Nutrition.	Type weight in kg, to max 2 decimals. If no measurement / unknown, leave the default for unknown value = '-1'.

Variable	Description	Input ECFSTracker
Height	Height measured at date of best FEV1 or, if no lung function test, last previously recorded height of the year (without shoes and socks); See also Part 2. iii Nutrition.	Type height in cm, to max 2 decimals. If no measurement / unknown, leave the default for unknown value = '-1'.
FEV1	Value of FEV1, in litres (up to 2 decimals), of the highest FEV1% predicted of the year, in accordance with local reference values; Record value, if you have it, for patients who died during the year of follow-up; For patients who have had a lung transplant during the year of follow-up, record the best FEV1 from before transplant; See also Part 2. iv Spirometry and Part 2. x Transplantation.	Type value in litres, to max 2 decimals; If no test / unknown, leave the default '-1'.
FVC	Value of FVC in litres (up to 2 decimals) at date of recorded FEV1. See also Part 2. iv Spirometry and Part 2 x Transplantation.	Type value in litres, to max 2 decimals; If no test / unknown, leave the default '-1'.
Date of FEV1/growth measurement	Date of recorded FEV1 (of highest FEV1% predicted of the year), or, if no lung function, the date of the last recorded weight/height of the year; See also Part 2. iii Nutrition and Part 2. iv Spirometry.	Type or select from calendar; If no lung function and no growth measurements, leave blank; Date unknown: 01/07; Only day unknown: 15; Only month unknown: 7; Born after 01/07 of year of follow up: use date of diagnosis or 31/12.
Lowest LCI 2.5%	Value of lowest LCI (Lung Clearance Index) 2.5% of the year. If a value is recorded, date and device should be recorded. The value should be between 2 and 25. See also Part 2. v Lung Clearance Index	Type the value.
Date of lowest LCI 2.5%	If a value is recorded for lowest LCI 2.5%, a date is required.	Type or select from calendar. Date unknown: 01/07; Only day unknown: 15; Only month unknown: 7; Born after 01/07 of year of follow up: use date of diagnosis or 31/12

Variable	Description	Input ECFSTracker
Device used for LCI measurement	If a value is recorded, the device used for the test should be recorded. Options: Unknown; Missing (variable not collected); Ecomedics Exhalyzer D N2-Washout; Ecomedics Exhalyzer SF6 (Tracergas 4 %); Innovision Innocor; NDD Easyone Pro; Other.	Select from options provided.
Microbiology, Intravenous antibiotics, Hospitalisation *Chronic infection: more than 50% of samples - sputum/other - should be positive (at least 4 samples from the year of follow-up). See also Part 2.vi Chronic infection in the lower airways.*		
Chronic Pseudomonas aeruginosa	Did the patient have chronic Pseudomonas aeruginosa during the year of follow up? See also Part 2.v Chronic Infection in the lower airways. Options: No; Unknown; Missing / variable not collected; Yes.	Select from options provided.
Chronic Staphylococcus aureus	Did the patient have chronic Staphylococcus aureus infection during the year of follow up? See also Part 2.v Chronic Infection in the lower airways. Options: No; Unknown; Missing / variable not collected; Yes.	Select from options provided.
Chronic Burkholderia cepacia complex	Did the patient have chronic Burkholderia cepacia complex during the year of follow up? See also Part 2.v Chronic Infection in the lower airways. Options: No; Unknown; Missing / variable not collected; Yes.	Select from options provided.

Variable	Description	Input ECFSTracker
Haemophilus influenzae	Did the patient have Haemophilus influenzae at any time during the year of follow up? Options: Never during the year of follow-up; Unknown; Missing / variable not collected; Yes, at least once during the year of follow-up.	Select from options provided.
Nontuberculous mycobacteria	Did the patient have nontuberculous mycobacteria at any time during the year of follow up? Options: Never during the year of follow-up; Unknown; Missing / variable not collected; Yes, at least once during the year of follow-up.	Select from options provided.
Stenotrophomonas maltophilia	Did the patient have Stenotrophomonas maltophilia at any time during the year of follow up? Options: Never during the year of follow-up; Unknown; Missing / variable not collected; Yes, at least once during the year of follow-up.	Select from options provided.
Achromobacter Species	Did the patient have Achromobacter Species at any time during the year of follow up? Options: Never during the year of follow-up; Unknown; Missing / variable not collected; Yes, at least once during the year of follow-up.	Select from options provided.
MRSA - methyicillin-resistant Staphylococcus aureus	Did the patient have methycillin-resistant Staphylococcus aureus at any time during the year of follow-up? Options: Never during the year of follow-up; Unknown; Missing / variable not collected; Yes, at least once during the year of follow-up.	Select from options provided.

Variable	Description	Input ECFSTracker
IV antibiotics at home and in hospital	Total no. of days on intravenous antibiotics for CF-related reasons, in hospital and at home, during the year of follow-up.	Record number of days, include first and last day.
IV antibiotics - hospital only	No. of days on intravenous antibiotics for CF-related reasons, in hospital, during the year of follow-up.	Record number of days, include first and last day.
Hospitalisation	Record the number of days spent in hospital during the year of follow-up, for any reason, not just those related to CF. Routine check-up days however should not be included.	Record number of days, include first day (but not last day if discharged in the morning).
Complications of CF		
ABPA - Allergic bronco-pulmonary aspergillosis	Was the patient diagnosed with or treated for allergic bronco-pulmonary aspergillosis at any time during the year of follow-up? See Part 2.vi. ABPA for the diagnostic criteria Options: Never during the year of follow-up; Unknown; Missing / variable not collected; Current ABPA (diagnosed or still treated).	Select from options provided.
DIOS - Distal intestinal obstruction syndrome	Did the patient have distal intestinal obstruction syndrome at any time during the year of follow-up? Options: No; Unknown; Missing / variable not collected; Yes.	Select from options provided.
Salt loss syndrome	Did the patient have salt-loss syndrome at any time during the year of follow-up? See diagnostic criteria for SLS in Part 2.viii. Pancreatic Status. Options: No; Unknown; Missing / variable not collected; Yes.	Select from options provided.

Variable	Description	Input ECFSTracker
Diabetes: treated	Was the patient treated for diabetes and, if yes, what kind of treatment? Options: No; Unknown; Missing / variable not collected; Yes. If yes: Treated with daily insulin; Treated with oral hypoglycaemic agents; Only dietary advice; Therapy unknown.	Select from options provided.
Pneuomothorax	Did the patient have a pneumothorax during the year of follow up? If yes, specify if a chest drain was required. Options: No; Unknown; Missing / variable not collected; Yes. If yes: Chest drain required; Observation only; Unknown.	Select from options provided.
Liver disease	Did the patient have liver disease during the year of follow-up and, if yes, what kind? If the patient has had a liver transplant, the data should reflect the post-transplant situation. For definitions see Part 2.vii. Liver Disease Options: No; Unknown; Missing / variable not collected; Yes. If yes: Cirrhosis, no portal hypertension/hypersplenism; Cirrhosis, portal hypertension/hypersplenism; Cirrhosis, portal hypertension unknown; Liver disease without cirrhosis; Variaceal bleeding.	Select from options provided.

Variable	Description	Input ECFSTracker
Haemoptysis (major haemoptysis only)	Did the patient have a major haemoptysis during the year of follow up (>250ml)? Options: Never during the year of follow-up; Unknown; Missing / variable not collected; Yes, at least once during the year of follow-up.	Select from options provided.
Occurrence of malignancy (this year)	Was the patient diagnosed with cancer during the year of follow-up? **If yes, indicate what type, if known.** **Options:** **Colorectal;** **Small bowel;** **Lymphoid leukaemia;** **Testicular;** **Breast;** **Thyroid;** **Type unknown;** **Other.**	Select from options provided.
Pancreatic status: faecal elastase	Was faecal elastase measured during the year of follow-up? If yes, indicate the result. See also Part 2.viii Pancreatic status Options: Not done; Unknown; Missing/variable not collected; Yes: If yes: <200 µg/g once; < 200µg/g twice; ≥ 200 µg/g once; ≥ 200 µg/g twice.	Select from options provided.

Variable	Description	Input ECFSTracker
Pancreatic status: faecal fat	Was faecal elastase measured during the year of follow-up? If yes, indicate the result. Options: Not done; Unknown; Missing/variable not collected; Yes: If yes: high once; high twice; normal once; normal twice.	Select from options provided.
Transplantation		
Has the patient had a lung transplant?	Every year we collect information about patients who have had a lung transplant during their life time (not just those transplanted during follow-up year), and the year of the latest transplant. If a transplant was already recorded for a patient in a previous year, it is automatically copied to all future PASs - it does not need to be input every year. If a patient has a subsequent transplant of the same organ, that it is a re-transplant, and the new year, are recorded. Options: No; Unknown; Missing (variable not collected); Yes.	First transplant during the year of follow-up: select option and type the year; Previous transplant: will be copied to the PAS when the year is initialised; 2nd Transplant: select "re-transplant" and input the new year of transplant.
Year of latest lung transplant	If "yes" is selected for transplant, the year of the latest transplant is required.	Type the year
Has the patient had a re-transplant?	If the patient has had a subsequent transplant we collect this information, and the year.	Select the option and update the year field.
Has the patient had a liver transplant?	As for lung transplant	As for lung transplant
Year of latest liver transplant	As for lung transplant	As for lung transplant
Has the patient had a re-transplant	As for lung transplant	As for lung transplant

Variable	Description	Input ECFSTracker
Has the patient had a kidney transplant?	As for lung transplant	As for lung transplant
Year of latest kidney transplant	As for lung transplant	As for lung transplant
Has the patient had a re-transplant	As for lung transplant	As for lung transplant
Has the patient had another organ	Indicate if the patient has had a transplant of an organ other than lung, liver or kidney.	As for lung transplant
Year of latest other transplant	As for lung transplant	As for lung transplant
Has the patient had a re-transplant	As for lung transplant	As for lung transplant

Part 2: Criteria, Definitions and References

2.i. Diagnostic Inclusion Criteria and Reversal of Diagnosis

1. Two sweat tests value > 59 mmol/L chloride: CF diagnosis accepted.
2. One sweat test value > 59 mmol/L chloride + DNA Analysis/Genotyping – two identified disease-causing CF mutations: CF diagnosis accepted.
3. If the sweat value is less than or equal to 60 mmol/L chloride, or not reported, then at least 2 of the following must be fulfilled:
   a. DNA Analysis/Genotyping: two identified disease causing CF mutations;
   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.
4. Diagnosis reversal*
   CF diagnosis should be reversed if any of the following cases are true:
   a. DNA Analysis: unable to identify any disease causing CF mutations;
   b. NPD (nasal potential difference) and/or ICM (intestinal current measurement): result not consistent with a diagnosis of CF;
   c. Normal values from repeated sweat testing (confirm with the clinical team).

   *See also ECFSPR Standard Operating Procedure regarding reversal of diagnosis and previously submitted date on the homepage of data collection software ECFSTracker.

2.ii Sweat Test: parameters; values to be reported.

1. Diagnostic standards: the quantity of sweat should indicate an adequate rate of sweat production;
2. a. The sweat sample should be processed immediately after sweat collection;
   b. Chloride concentration measurement is the preferred analysis;
   c. Chloride value: report the Chloride value in millimols per litre (mmol/L); If duplicate tests were completed on the same day, **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;***
3. The ECFSPR considers only Titration/Chloride values in analyses.

References:

ECFS Standards of Care (2013) and ECFS Best Practice Guidelines Update (2018)

Part 2: Criteria, Definitions and References

2.iii Nutrition: method; values to be reported; References.

Weight and height should be measured in accordance with the guidelines of EuroCareCF;
Z-scores for height, weight and BMI will be calculated using the CDC reference values [Kuczmarski et al (2002)].

1. **Weight**: removal of outer clothing, shoes and socks;
2. **Height:** without shoes and socks; stadiometer - top of head in contact with head board, slight pressure;
3. **Date:** the recorded height and weight should be the measurements taken t**he same day as the best FEV1**.
   If spirometry was not done the l**ast weight and height measurements of the year, and the date they were measured**, should be recorded.

References:

a) 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

b) 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.

c) Public Use File BGS98, German National Health Interview and Examination Survey 1998, Robert-Koch-Institut, Berlin, Germany, 2000

d) 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

e) 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

2.iv Spirometry: criteria; method; values to be reported; References.

The ECFS Patient Registry collects data on spirometry values in order 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 /extracted by the National Registries.**
All spirometry tests should be carried out in accordance with the ATS/ERS guidelines: **(www.thoracic.org/statements/resources/pfet/PFT2.pdf).**

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

1. **Pre-test:**

a) **Date of birth, gender and height** should be recorded for calculation of predicted values;

b) **All recorded spirometry tests should be pre-bronchodilator* values**
   i. short-acting bronchodilators: at least 4 hours pre-test
   ii. long-acting bronchodilators: at least 12 hours pre-test

*In accordance with the official criteria of PortCF.

Part 2: Criteria, Definitions and References

2. **Values to report:**

a) FEV1 value to report: Value of FEV1, in litres (up to 2 decimals), of the highest FEV1% predicted of the year, in accordance with local reference values;

b) The FEV1 and FVC measurements must be reported in litres (L), to max 2 decimal points;

c) The FVC measurement is the FVC on the date of recorded FEV1 and it must be greater than or equal to the FEV1 measurement;

d) For the reported spirometry value, 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;

e) Only tests deemed valid according to ATS/ERS guidelines to be reported

3. **Calculation of percent of predicted values:**

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

n.b. The ECFSPR Definitions Group considered the issue of race-specific reference values. The decision was to not record race for European patients and therefore not to calculate race-specific values.

References:

a) 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).

b) Miller et al. Standardisation of spirometry. Eur Respir J 2005; 26: 319–338.

c) Miller et al. General considerations for lung function testing. Eur Respir J 2005; 26: 153–161.

d) Cystic Fibrosis Foundation Patient Registry User Guide, Version 4.0. 2006.

e) 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.

2.v. Chronic infection in the lower airways: definition, References.

1. 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.
   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.

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

Part 2: Criteria, Definitions and References

References:

a) 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

b) 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.

c) 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.

2.vi. Allergic Bronchopulmonary Aspergillosis (ABPA): diagnostic criteria, References

Diagnostic criteria

1. Acute or subacute clinical deterioration (cough, wheeze, exercise intolerance, exercise-induced
2. Total IgE > 500 IU/ml;
3. Positive skin prick test for Aspergillus antigen (> 3 mm), or positive specific IgE for A. fumigatus.
4. Either:
   a). Precipitins to A. fumigatus, or in vitro demonstration of IgG antibody to A. fumigatus;
   or
   b). 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:

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. Allergic bronchopulmonary aspergillosis in cystic fibrosis-state of the art: Cystic Fibrosis Foundation Consensus Conference. Clin Infect Dis. 2003 Oct 1;37 Suppl 3:S225-64

2.vii. 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.

2.viii. Pancreatic Status: Definition and References

1. **Definition of pancreatic insufficiency** (two determinations are mandatory)

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

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

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

Part 2: Criteria, Definitions and References

2. **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:

a) 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.

b) 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.

2.ix Salt Loss Syndrome: definition and Reference

Primary metabolic alkalosis with blood pH > 7.45, serum sodium < 130 mmol/l and serum chloride < 90 mmol/l.

References:

c) Reference: 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.

2.ix. Transplantation: Indications

1. *For patients who had a transplant during the year of follow up:
   Use the best FEV1 before transplantation;
   Record therapy, complications and microbiology from before transplantation.

2. For patients who had a transplant before the current follow-up year:
   Record all information available.

* If a patient is transferred to a different hospital for transplant, and that hospital submits data to the ECFSPR, the transplant centre must not re-register the patient. The Core Data will be transferred through the data collection software and the transplant centre should record pre-transplant data.