



Variables, Inclusion Criteria and References used by the ECFSPR (valid from 2024 YFU)

New variables introduced for the 2024 follow-up year are blue

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.

Maintenance Therapy

- Inhaled continuous (≥ 3 months) hypertonic saline $\geq 3\%$;
- Inhaled continuous (≥ 3 months) Mannitol;
- Inhaled antibiotic this year - continuous (≥ 3 months) or on/off for a total of (≥ 6 months);
- 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;

Diagnosis

- Age at diagnosis;
- Sweat test type and values (x2);
- First & second genetic CFTR 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.

- 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);
- Use of CFTR Modulator Therapy (data for each of the following are collected: Ivacaftor, Lumacaftor/ Ivacaftor, Tezacaftor/Ivacaftor, Elexacaftor/Tezacaftor/Ivacaftor, **Vanzacaftor/Texacaftor/Deutivacaftor**, Other CFTR modulator);
- Start and stop dates x 2 for each CFTR modulator (pause of <30 days not to be recorded);
- Sweat chloride values - before start & during (lowest of year) for each CFTR modulator;
- **Principal reason for stopping CFTRm therapy (for each stop).**



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 LCI2.5 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 except Nontuberculous mycobacteria and Fungi)*

- Pseudomonas aeruginosa;
- MSSA - Methicillin-sensitive Staphylococcus aureus;
- MRSA - Methicillin-resistant Staphylococcus aureus;
- Burkholderia cepacia complex;
- Stenotrophomonas maltophilia;
- Achromobacter spp;
- Haemophilus influenza;
- Nontuberculous mycobacteria (tested for);
- 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 PEx episodes treated with intravenous antibiotics during the year.

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 (from 2024 YFU change to ICD10 codes C00 - C96)
- Pancreatic status: faecal elastase;
- Pancreatic status: faecal fat;
- Pregnancy this year;
- Pregnancy stopped this year - reason for stop;
- Pregnancy ongoing at 31/12.
- **BMD (Bone mineral density): osteopenia or osteoporosis in year of follow-up;**
- **BMD Diagnosis/status: was the diagnosis/status based on a DEXA scan?**
- **At least 1 episode of acute pancreatitis this year;**
- **Arterial hypertension treated with medication during the year of follow-up**
- **Hypercholesterolaemia treated with medication during the year of follow-up**

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).