

# Variable list NCFR 2022

The variable list of the Dutch Cystic Fibrosis Registration (NCFR) is revised and updated every year. Below are the variables collected in 2022. These are divided into two categories: core data and annual data. Core data is collected once during the first year of participation in the registration. Annual data is collected every year.

## Core data:

### Basics

Description	Variables	Comments
Date of birth	Year of birth Month of birth	By default, birthday is set to the 15th of the month
Gender	Male/female/other	
Ethnicity	Ethnicity (multiple choice)	
Height of parents	Mother's height in cm Father's height in cm	Only entered for children
CFTR-mutations	First mutation (text) Second mutation (text)	By legacy name

### Diagnosis

Description	Variables	Comments
Has the CF diagnosis been confirmed?	Yes / No, CF-SPID / No, no CF-SPID either	
Age at diagnosis	Age in years	
Sweat test	Done yes/no Date of execution Type of titration test Result	
Neonatal screening	Done yes/no Result positive/negative	
Meconium ileus	Yes/no Operated or not operated on	
Intestinal current measurements (ICM)	Done yes/no ICM result: CF diagnosis confirmed/not confirmed	
Symptoms around CF diagnosis	Airway symptoms Electrolyte imbalance Growth and/or nutritional problems Family history Liver problems Nasal polyps/sinusitis Prenatal screening Rectal prolapse CBAVD	

# Annual data

## Basics

Description	Variables	Comments
Patient status	Seen all year/ transfer/ deceased/ lost to follow-up	
Transfer	To which centre When	
Deceased	Cause Date	
Seen in hospital	Number of outpatient visits Number of days hospitalized	Including video consultations

## Complications

Description	Variables	Comments
Glucose tolerance test	Done yes/no Type of test Result: normal/ impaired glucose tolerance/ diabetes	
Diabetes	And/nee Treatment: insulin/ oral agents/ dietary advice	
ABPA	Yes/no	
Pneumothorax	Yes/no With or without drain	
Liver disease	No With or without cirrhosis With or without hypertension	
Haemoptysis	Lung haemorrhage yes/no	At least 250ml in one day
Faecal elastase	Never performed More/ less than 200ug Once or twice	
Malignancy	No/ yes, this year/ yes, before this year Type of malignancy	
Other complications	Arthritis of arthropathy GOD Depression GERD Gastrointestinal bleeding Hearing loss Nasal polyps Sinusitis Osteopenia Osteoporosis Bone fracture Pancreatitis Rectal prolapse Renal failure	For each of these complications, indicate: Yes/No

## Treatment

Description	Variables	Comments
Chronic inhalation of hypertonic saline (NaCl)	Yes/no	At least three months
Dornase alfa (pulmozyme)	Yes/no	At least three months
Chronic inhalation of antibiotics	Tobramycin solution Colistin solution Dry powder (any brand) Aztreonamlysin solution Levofloxacin solution Other inhalation antibiotics	At least three months
IV antibiotics	Number of days with IV: in hospital/ at home/ in total Number of courses	Number of courses as a surrogate for exacerbations
Chronic use of azithromycin or other macrolide	Yes/no	At least three months
Chronic inhalation of bronchodilators	Yes/no Short-acting/ long-acting/ combination	At least three months
Corticosteroids	Inhalation and/or oral	At least three months
Ursodeoxycholic acid	Yes/no	At least three months
Pancreatic enzymes	Yes/no	At least three months
Proton pump inhibitors	Yes/no	At least three months
Oxygen supplementation	Oxygen treatment Non-invasive ventilation: no/ yes, CPAP/ yes, BiPAP	At least three months
Consultation with dietitian	Yes/no With/ without protein, fat and calorie breakdown	
Food supplements	Oral dietary supplement(s) Gastric tube Gastric stoma	

## CFTR modulators

Description	Variables	Comments
CFTR modulators this year	Yes/no	
Ivacaftor	If no, latest sweat test: result and date Starting date Sweat test before start, result and date Sweat test after start, result and date Stop date Possible restart	
Lumacaftor/ivacaftor	Starting date Sweat test before start, result and date Sweat test after start, result and date Stop date Possible restart	
Tezacaftor/ivacaftor	Starting date Sweat test before start, result and date Sweat test after start, result and date Stop date Possible restart	
Elexacaftor/tezacaftor/ivacaftor	Starting date Sweat test before start, result and date	

	Sweat test after start, result and date Stop date Possible restart	
Other modulator	Starting date Sweat test before start, result and date Sweat test after start, result and date Stop date Possible restart	In the case of blinded study, this is entered as 'other modulator'

## Lung function / growth measurement

Description	Variables	Comments
Lung function measurement	Number of measurements this year Date of best FEV1 Age at measurement	If no lung function measurement is done, then date of last growth measurement is recorded
FEV1 – of best FEV1% predicted	FEV1 in litres %predicted Z-score	Scores calculated according to GLI
FVC – of best FEV1% predicted	FVC in litres %predicted Z-score	Scores calculated according to GLI
Growth measurement	Height Weight BMI Z-scores children	
LCI measurement	Done yes/no Date Apparatus LCI value	

## Microbiology

Description	Variables	Comments
Number of cultures in the year	Number	Including BAL and cough, nose or throat swabs
Pseudomonas aeruginosa	No/ yes, chronic/ yes, not chronic	Chronic based on Leeds criteria or doctor determination
MSSA	No/ yes, chronic/ yes, not chronic	
MRSA	No/ yes, chronic/ yes, not chronic	
Burkholderia cepacia complex	No/ yes, chronic/ yes, not chronic	
Haemophilus influenzae	No/ yes, chronic/ yes, not chronic	
Stenotrophomonas maltophilia	No/ yes, chronic/ yes, not chronic	
Achromobacter species	No/ yes, chronic/ yes, not chronic	
Fungal infection	Cultured yes/no Culture positive/negative Aspergillus fumigatus	

NTM	Scedosporium species Cultured yes/no Culture positive/negative M. abscessus M. avium complex Other variant (not specified)
Sars-CoV-2	No/ yes, symptomatic/ yes, asymptomatic Vaccination this year yes/no

## Transplantation

Description	Variables	Comments
Liver transplantation	Yes/no Year of transplantation Waiting list yes/no	
Lung transplantation	Yes/no Year of transplantation Waiting list yes/no	
Kidney transplantation	Yes/no Year of transplantation Waiting list yes/no	
Other organs	Yes/no Year of transplantation Waiting list yes/no	

## Socioeconomic

Description	Variables	Comments
Work	Unemployed Full-time job Part-time job Stay-at-home parent Student Unable to work Retired Person under 18 years of age	
Pregnancy this year	Yes/no Ongoing on 31/12 or ended before 31/12 Outcome of pregnancy	
Pregnancy ever	Yes/no Born alive/ abortion/ stillborn	