

1. Patient personal details

Insurance number:.....
Name:.....
Date of Birth:.....
Gender: female / male
Race: Asian/Indian / White / Black / N/A
Date of visit:
Year of follow-up:.....
Diagnosis of CF (year):.....

CF centre:
Doctor code:
Blood sample code:
Date of blood sampling:
Stool sample code:
Date of stool sampling:

2. Social background

Family members living together with the patient
Siblings
Social services input yes / no / no data
Kindergarten/School/other
Adherence good yes / no / no data

3. Diagnosis of cystic fibrosis

Two sweat tests value > 60 mmol/L chloride yes / no / no data

One sweat test value > 60 mmol/L chloride and two disease causing CF mutations identified by genotyping yes / no / no data

Type of the sweat test: chloridometer/iontophoresis-nonselective condutance/no data

At least 2 of these criteria are fulfilled:

- two disease causing CF mutations identified by genotyping
- Transepithelial (Nasal) Potential Difference – study consistent with a diagnosis of CF;
- Clinical Presentation – typical features of CF.
- no data

4. Diagnosis reversal

yes / no / no data

If yes, please specify:

- two CF mutations can not be identified by genotyping

- Transepithelial (Nasal) Potential Difference – study not consistent with CF
- Confirmed repeated normal sweat testing

5. Medical history (current or previous)

A) General information about the medical history

Alcohol consumption: yes / no / no data
if yes: frequency: occasionally / monthly / weekly / daily
amount (g/occasion):.....
duration (years).....

total alcohol consumption in the last 2 weeks:

if not: Alcohol consumption earlier yes / no/ no data
if yes: frequency: occasionally / monthly / weekly / daily
amount (g/occasion):.....
duration (years).....
How long ago did you stop drinking alcohol?.....

Guide for estimation of the amount:
1 dl beer (4.5 vol. %) = ~3.5 g alcohol
1 dl wine (12.5 vol. %) = ~10 g alcohol
1 dl hard drink (50 vol. %) = ~40 g alcohol

Smoking: yes / no/ no data
if yes: amount (cigarettes/day):.....
duration (years).....

if not: Smoking earlier yes / no/ no data
if yes: amount (cigarettes/day):.....
duration (years).....
How long ago did you stop smoking?

do parents smoking? yes / no/ no data
if yes: inside/outside/no data
do other household members smoking? yes / no/ no data
if yes: inside/outside/no data

Drug abuse: yes / no / no data
Prescribed medication should not be included here.
if yes: name of drug:..... dose:.....
duration (years).....

Heart disease: yes / no / no data

Renal disease: yes / no / no data

Liver disease: yes / no / no data

Diabetes mellitus: yes / no / no data

if yes: type: Type I. / Type II. / Type III. c / MODY/undetermined

date of diagnosis (year):.....

GI problems: yes / no / no data

Disease of the pancreas: yes / no / no data

if yes: acute pancreatitis / chronic pancreatitis / autoimmune pancreatitis / pancreatic cancer / congenital anatomical malformation / other

if other: please describe:.....

If acute pancreatitis:

Number of acute episodes before current:.....

Date of first episode (year):.....

If chronic / autoimmune pancreatitis:

Date of diagnosis:.....

Date of first episode (year):.....

Number of acute episodes before current:.....

If pancreatic cancer:

Date of diagnosis:.....

Diagnosed with chronic pancreatitis? yes / no / no data

If yes, date of diagnosis:.....

Number of acute episodes before current:.....

Date of first episode (year):.....

If congenital anatomical malformation:

Date of diagnosis:.....

Date of first episode (year):.....

Number of acute episodes before current:.....

If cystic lesions/malformation:

Date of diagnosis:

Number of cysts:

Diameters of cysts:

Other:.....

Porthacath: yes / no / no data

if yes, please describe type:.....

Gastrostomy: yes / no / no data

if yes, please describe type:.....

Malignancies: yes / no / no data

if yes, please describe type:.....

Allergies: yes / no / no data

if yes, please describe type:.....

Transplantation

Did the patient have a transplantation surgery? yes / no/ no data

liver transplant yes / no/ no data

year of latest:.....

lung transplant yes / no/ no data

year of latest:.....

other, please specify:..... yes / no/ no data

year of latest:.....

referred awaiting assessment yes / no/ no data

date:.....

assessed awaiting decision yes / no/ no data

date:.....

Decision on suitability for listing yes / no/ no data

date:.....

B) Exacerbation history

Oral antibiotics taken

name:..... start date:..... stop date:.....

name:..... start date:..... stop date:.....

name:..... start date:..... stop date:.....

Inhaled antibiotics

name:..... start date:..... stop date:.....

name:..... start date:..... stop date:.....

name:..... start date:..... stop date:.....

Intravenous antibiotics

name:..... start date:..... stop date:.....

name:..... start date:..... stop date:.....

name:..... start date:..... stop date:.....

Hospitalisations:

date:..... reason:.....

date:..... reason:.....

date:..... reason:.....



C) Microbiology (Have these organisms ever been isolated?)

Mucoid Pseudomonas aeruginosa yes / no/ no data
Sample Type oropharyngeal swab / sputum / bronchoalveolar lavage /Unknown
Sample Date:.....

Non-mucoid Pseudomonas aeruginosa yes / no/ no data
Sample Type oropharyngeal swab / sputum / bronchoalveolar lavage /Unknown
Sample Date:.....

Staphylococcus Aureus yes / no/ no data
Sample Type oropharyngeal swab / sputum / bronchoalveolar lavage /Unknown
Sample Date:.....

Methicillin-Resistant Staphylococcus aureus yes / no/ no data
Sample Type oropharyngeal swab / sputum / bronchoalveolar lavage /Unknown
Sample Date:.....

Burkholderia cepacia complex yes / no/ no data
Sample Type oropharyngeal swab / sputum / bronchoalveolar lavage /Unknown
Sample Date:.....

Non-tuberculous Mycobacteria no / MAC / abscessus / other/ no data
Sample Type oropharyngeal swab / sputum / bronchoalveolar lavage /Unknown
Sample Date:.....

Stenotrophomonas maltophilia yes / no/ no data
Sample Type oropharyngeal swab / sputum / bronchoalveolar lavage /Unknown
Sample Date:.....

Achromobacter species yes / no/ no data
Sample Type oropharyngeal swab / sputum / bronchoalveolar lavage /Unknown
Sample Date:.....

Haemophilus influenzae yes / no/ no data
Sample Type oropharyngeal swab / sputum / bronchoalveolar lavage /Unknown
Sample Date:.....

D) Complications (Have these complications ever been occurred?)

Allergic bronchopulmonary aspergillosis (ABPA) yes / no/ no data
Distal intestinal obstruction syndrome (DIOS) yes / no/ no data
Pneumothorax requiring chest drain yes / no/ no data
Liver disease yes / no/ no data
Haemoptysis more than 250ml yes / no/ no data
Malignancy diagnosed yes / no/ no data

Sinusitis	yes / no/ no data
Osteoporosis	yes / no/ no data
Osteopenia	yes / no/ no data
Gastroesophageal reflux	yes / no/ no data

6. Diseases are running in the family currently and/or previously

Cystic fibrosis in family history: yes / no / no data
if yes, please describe type:..... relationship to patient:.....

Diabetes mellitus in family history: yes / no / no data
if yes, please describe type:..... relationship to patient:.....

Pancreas disorders in family history: yes / no / no data

acute pancreatitis:	yes / no	if yes: relationship to patient:.....
chronic pancreatitis:	yes / no	if yes: relationship to patient:.....
autoimmune pancreatitis:	yes / no	if yes: relationship to patient:.....
pancreatic cancer:	yes / no	if yes: relationship to patient:.....
congenital anatomical malformation:	yes / no	if yes: relationship to patient:.....
other (please describe):.....		if yes: relationship to patient:.....

Other GI disease in family history: yes / no / no data
if yes, please describe type:..... relationship to patient:.....

Heart disease in family history: yes / no / no data
if yes, please describe type:..... relationship to patient:.....

Renal disease in family history: yes / no / no data
if yes, please describe type:..... relationship to patient:.....

Liver disease in family history: yes / no / no data
if yes, please describe type:..... relationship to patient:.....

Any other disease in the family history:

7. Current Medications

Maintenance antibiotic therapy: yes / no/ no data

if yes, please specify:

<u>Nebulised Tobramycin</u>	no / continuous / alternating monthly / unknown
<u>Nebulised Colomycin</u>	no / continuous / alternating monthly / unknown
<u>Nebulised Aztreonam</u>	no / continuous / alternating monthly / unknown
<u>Tobramycin Dry Powder Inhaler</u>	no / continuous / alternating monthly / unknown
<u>Colomycin Dry Powder Inhaler</u>	no / continuous / alternating monthly / unknown
<u>Azithromycin Maint. Therapy</u>	no / continuous / alternating monthly / unknown

Anti Staphylococcal Maint. Therapy no / continuous / alternating monthly / unknown

Name of other nebulised / dry powder antibiotic:.....

Respiratory therapy: yes / no/no data

if yes, please specify:

Use of rhDNase yes / no / no data

if yes, please specify name:.....dose:.....

Use of Hypertonic Saline - Percent yes / no / no data

if yes, please specify name:.....dose:.....

Inhaled Short Acting Bronchodilators yes / no / no data

if yes, please specify name:.....dose:.....

Inhaled Long Acting Bronchodilators yes / no / no data

if yes, please specify name:.....dose:.....

Inhaled Steroid Alone yes / no / no data

if yes, please specify name:.....dose:.....

Inhaled Steroid with Long-acting Muscarinic Antagonist (LAMA) yes / no / no data

if yes, please specify name:.....dose:.....

Inhaled Long-acting Beta-agonist (LABA) yes / no / no data

if yes, please specify name:.....dose:.....

Inhaled nasal steroid yes / no / no data

if yes, please specify name:.....dose:.....

Oral steroid yes / no / no data

if yes, please specify name:.....dose:.....

Oxygen Therapy yes / no / no data

Non-invasive positive pressure ventilation yes / no / no data

Nutrition / GI therapy:

Pancreatic enzyme replacement yes / no / no data

if yes, please specify name:.....dose:.....

Ursodeoxycholic Acid yes / no / no data

if yes, please specify name:.....dose:.....

Proton Pump Inhibitors yes / no / no data

if yes, please specify name:.....dose:.....

Enteral Feeding yes / no / no data
if yes, please specify name:.....dose:.....

Vitamin A Supplements yes / no / no data
if yes, please specify name:.....dose:.....

Vitamin E Supplements yes / no / no data
if yes, please specify name:.....dose:.....

Vitamin D Supplements yes / no / no data
if yes, please specify name:.....dose:.....

Vitamin K Supplements yes / no / no data
if yes, please specify name:.....dose:.....

Endocrine therapy:

Hormonal contraception yes / no / no data
if yes, please specify name:.....dose:.....

Calcium supplementation yes / no / no data
if yes, please specify name:.....dose:.....

Bisphosphonate yes / no / no data
if yes, please specify name:.....dose:.....

Insulin yes / no / no data
if yes, please specify name:.....dose:.....

Other therapy:

CFTR modifier therapy yes / no / no data

Ivacaftor yes / no / no data

Lumacaftor yes / no / no data
if other, please specify name:.....dose:.....

8. General state at registration

A) Status

Heart rate (/minute)	
Blood pressure (Hgmm)	
Respiratory rate (/minute)	
Oxygen saturation (%) on air	
Weight (kg)	
Height (cm)	
Height and weight Z-score	
BMI	
BMI age percentile	
BMI Z-score	
Body temperature (°C) axillary	

B) Respiratory function tests

	Value	%
FEV1		
FVC		
FEV1/FVC		
TLC		
FRC		
RV		
TLCO		
KCO		

Appetite: good / retained / bad/ no data

Weight loss: yes / no / no data

if yes: duration (weeks):.....
amount (kg):.....

Stool: normal / diarrhea / constipation / fatty / putrid / undigested food / bloody / mucus

9. Laboratory parameters

A) Biochemistry

Sodium (mmol/l)	
Potassium (mmol/l)	
Calcium (mmol/l)	
Iron (umol/l)	
Transferrin (ug/l)	
Ferritin (mg/dl)	
Total protein (g/l)	
Albumin (g/l)	
Cholesterol (mmol/l) fasting	

Triglyceride (mmol/l) fasting	
ASAT/GOT (U/l)	
ALAT/GPT (U/l)	
Gamma GT (U/l)	
Total bilirubin (umol/l)	
Direct/Conjugated bilirubin (umol/l)	
Alkaline phosphatase (U/l)	
Lactate dehydrogenase (U/l)	
Cholinesterase (U/l)	
Blood urea nitrogen (mmol/l)	
Creatinine (umol/l)	
C-reactive protein (mg/l)	
Amylase (U/l)	
Lipase (U/l)	

B) Hematology

White blood cell (WBC) count (G/l)	
Red blood cell (RBC) count (T/l)	
Hemoglobin (g/l)	
Hematocrit (%)	
Thrombocyte (G/l)	

C) Blood gas on air

arterial/capillary/venous

pH	
pO ₂	
pCO ₂	
BXS	
HCO ₃	
Sat	

D) Carbohydrate metabolism

serum glucose 0' (mmol/l)	
serum glucose 30' (mmol/l)	
serum glucose 60' (mmol/l)	
serum glucose 90' (mmol/l)	
serum glucose 120' (mmol/l)	
insulin 0' (pmol/l)	
insulin 60' (pmol/l)	
insulin 120' (pmol/l)	
Hemoglobin A1C (%)	

C-peptide	
Glutamic Acid Decarboxylase Autoantibodies	

E) Immunology and gluten sensitivite

IgA (g/l)	
IgM (g/l)	
IgG (g/l)	
EmA	
tTGA	

F) Virology

HIV	
CMV	
EBV	
Hepatitis A	
Hepatitis B	
Hepatitis C	

G) Others

faecal elastase	
sweat chloride (mmol/l)	

H) Additional Microbiology

10. Imaging examinations

yes / no/no data

Abdominal ultrasonography:

yes / no/ no data

Description:

.....
.....

Chest X-ray:

yes / no/no data

Description:

.....
.....

11. Final report

Consent form is filled

I declare that the patient received the necessary information and signed the consent form.

Notes:.....