Cystic Fibrosis Foundation Patient Registry Data Entry Guidelines

Patient Demographics Form

Record all known demographics information about the patient. This form can be edited at any time. For patients seen at multiple centers (i.e., shared patients), all centers with access to the patient’s record have the ability to update this form.

Be sure to update and fill-in any data that changes or becomes available after the original creation of this form.

Demographics

**Last Name**: Record the patient’s current last name. *Note: Be sure to check that the patient’s last and first name are not transposed.*

**First Name**: Record the patient’s first name. *Note: Be sure to check that the patient’s last and first name are not transposed.*

**Middle Name**: Record the patient’s middle initial or name.

**Last Name at Birth (if different)**: Record the patient’s last name at the time of his/her birth only if it is different from his/her current last name (i.e., the patient’s maiden name or the patient’s birth parents’ name if known).

**Last 4 digits of SSN**: If the patient has a Social Security Number and consents to provide it, record the last four digits of the patient’s SSN.

**Date of Birth**: Record the patient’s date of birth in the format mm/dd/yyyy.

**State of Birth**: Record the state in which the patient was born. If the patient was born outside of the U.S., select *Foreign Birth*. If the state of birth is not known, select *Unknown*.

**Gender**: Record the biological sex of the patient. If the patient is transgendered, select the biological sex, not the gender with which the patient identifies, even if gender reassignment surgery has been performed.

**Current Zip Code**: Record the zip code where the patient currently resides. If the patient’s place of residence is outside of the U.S., including Puerto Rico, report “Foreign”.

**Emergency Phone**: Enter the phone number at which the patient/family of the patient can be reached using the format ###-###-####.

**Is patient residing in the US permanently?** Select Yes, No, or Unknown.

**Email**: Enter the email address at which the patient/family of the patient can be reached.

Race/Ethnicity Information

**Race**: Record all races that apply, according to what the patient considers himself/herself.

- **White**: A person having origins in any of the original peoples of Europe, the Middle East, or North Africa.
- **Black or African American**: A person having origins in any of the Black racial groups of Africa.
- **American Indian or Alaska native** A person having origins in any of the original peoples of North and South America (including Central America) and who maintains tribal affiliation or community attachment.
- **Asian** A person having origins in any of the original peoples of the Far East, Southeast Asia, or the Indian subcontinent, including Cambodia, China, India, Japan, Korea, Malaysia, Pakistan, the Philippine Islands, Thailand, and Vietnam.
- **Native Hawaiian or Other Pacific Islander** A person having origins in any of the original peoples of Hawaii, Guam, Samoa, or other Pacific Islands.
- **Some other race** Select only if the patient's race cannot be described by any of those listed.
- **Two or more races** If selected, check all races that apply. Refer to previous descriptions to determine which may apply.
  - White
  - Black or African American
  - American Indian or Alaska Native
  - Asian
  - Native Hawaiian or Other Pacific Islander

**Is Patient of Hispanic Origin?** Select Yes, No, or Unknown. If the patient is of Cuban, Mexican, Puerto Rican, South or Central American, or other Spanish culture or origin, regardless of race, select Yes.

**Death Information**

This section only appears after first saving a new patient's demographics information. If the patient is currently living, leave this section blank.

**Date of Death:** Record the patient's date of death. If you know the patient is deceased but you do not know the date of death, estimate the date and check the box “Check if death date is approximate”. **Note:** Be sure to record all encounters that occurred prior to death.

**Primary Cause of Death:** Select the option that best describes the patient's cause of death. Only one may be selected. If you are uncertain of the cause, review the cause of death with your director or email RegHelp@cff.org and we will assist in determining the appropriate reason to select.

- Respiratory/cardiorespiratory
- Liver Disease/Liver Failure
- Trauma
- Suicide
- Transplant related: Bronchiolitis obliterans
- Transplant related: Other Includes rejection, infection, or cancer related to immunosuppressive therapy.
- Other Select only if the cause of death is not listed.
- Unknown If you are uncertain of the cause of the patient's death, review the cause of death with your director or email RegHelp@cff.org and we will assist in determining the appropriate reason to select. If a cause is still not established or is not known, record Unknown.

**Additional Information**

Please use this field to record any additional information about this patient Record any additional patient information.
CF Diagnosis/Possible CF Diagnosis Form

Record all known information about the patient’s diagnostic history. This form can be edited at any time. For patients seen at multiple centers (i.e., shared patients), all centers with access to the patient’s record have the ability to update this form.

Be sure to update and fill-in any data that changes or becomes available after the original creation of this form.

CF Diagnosis

Diagnosis Information

History of Patient Diagnosis

This section is a repeat group. As a result, multiple histories of patient diagnosis can be created, edited, and deleted as appropriate (i.e., if a patient’s diagnosis changes, a new entry can be created to reflect this change and incorrectly or incompletely entered histories of diagnosis can be edited or deleted). Only edit or delete an already existing entry in the event of incorrectly or incompletely entered information. Otherwise, create a new history by clicking “Create Next History of Patient Diagnosis” at the bottom of the section.

If the patient’s diagnosis has changed as a result of clinical presentation, genotyping, etc., the order of diagnosis history entries should reflect the sequence of diagnoses as they occurred. If you are unable to save a newly created history of diagnosis entry or edits to an already existing one, please contact reghelp@cff.org.

For more information about how to create, edit, and delete repeat group entries, visit https://portcf2dev.cff.org/RegistryLaunch/Documents/Manual/Manual.html#_Entering_and_Altering.

Date of Diagnosis (may be earlier than first sweat test): Record the date of diagnosis using the format mm/dd/yyyy. This should be the date on which the diagnosis was made by genotyping and/or sweat test. If diagnosis was confirmed by genotyping, the date the results were received should be entered. If the patient was diagnosed prenatally, the date the CF-specific prenatal testing results should be entered. If unsure of the patient’s date of diagnosis, contact the facility where the patient was diagnosed to obtain this information. If the exact date of diagnosis is not known, enter an approximated date and check the box “Date is an approximation” above.

Diagnosis Select the appropriate patient diagnosis.

- Cystic Fibrosis
- CFTR-related metabolic syndrome
- CFTR-related disorder
- CF, CRMS, and CFTR-related disorder all ruled out
Patient was diagnosed with CF after false negative results by newborn screening: Select Yes, No, or Unknown. If the patient was diagnosed after a positive newborn screening result or did not receive newborn screening, select No. If the newborn screening status or the newborn screening results of the patient are not known, select Unknown.

Diagnosis suggested by the following: Select all that apply. Symptoms, signs, and laboratory results listed are available for selection based on the reported diagnosis according to the options listed by diagnosis below.

**Diagnosis of Cystic Fibrosis**
- Acute or persistent respiratory abnormalities
- CBAVD (absent vas deferens) or related abnormalities
- Digital clubbing
- DNA Analysis
- Edema
- Electrolyte imbalance
- Failure to thrive/malnutrition
- Family History Diagnosis of CF in one or more first or second degree relatives.
- Infertility/GU abnormalities
- Liver problems
- Meconium ileus/other intestinal obstruction If selected, specify type below.
  - Please specify selection of meconium ileus:
    - Meconium ileus with perforation
    - Meconium ileus without perforation.
    - Other neonatal bowel obstructions Enter only if neither apply or if there are/were other, additional neonatal bowel obstructions.
- Nasal polyps/sinus disease
- Newborn (neonatal) screening Select if the patient ever had a positive newborn screening result.
- Prenatal screening (CVS, amnio)
- Rectal prolapse
- Steatorrhea/abnormal stools/malabsorption
- Other, specify: Select only if symptom/sign(s) is not listed. If selected, specify in the adjacent field all unlisted signs and/or symptoms.

**Diagnosis of CFTR-related metabolic syndrome (CRMS)**
- Elevated immunoreactive trypsinogen (IRT) at CF newborn screening
- Less than 2 identified disease causing mutation
- Non-diagnostic sweat chloride value (<60 mmol/L)
- Other, specify: Select only if symptom/sign(s) is not listed. If selected, specify in the adjacent field all unlisted signs and/or symptoms.

**Diagnosis of CFTR-related disorder**
- CBAVD (absent vas deferens) or related abnormalities
- Less than 2 identified disease causing mutation
- Non-diagnostic sweat chloride value (<60 mmol/L)
- Pancreatitis (not explained by other etiologies)
- Persistent respiratory colonization/infection with typical CF pathogen(s) (e.g., Pseudomonas aeruginosa)
- Pulmonary mycobacterial infection
- Other, specify: Select only if symptom/sign(s) are not listed. If selected, specify in the adjacent field all unlisted signs and/or symptoms.

**Diagnosis of CF, CRMS and CFTR-related disorder all ruled out**
- DNA Analysis
- Repeat Normal Sweat Testing
- Transepithelial potential differences
- Other, specify: Select only if symptom/sign(s) are not listed. If selected, specify in the adjacent field all unlisted signs and/or symptoms.

**Date & value of documented positive quantitative pilocarpine iontophoresis sweat test (Chloride)**

This section is a repeat group. As a result, multiple sweat test entries can be created, edited, and deleted as appropriate (i.e., if a patient undergoes multiple sweat tests, separate entries can be created and completed to record all results and incorrectly or incompletely entered sweat test results can be edited or deleted). Only edit or delete an already existing entry in the event of incorrectly or incompletely entered information. Otherwise, create a new entry by clicking “Create Next Sweat Test Measurement” at the bottom of the section.

If a sweat test was **not** performed on the patient, do not fill out this section. Record all results of all sweat tests performed at or verified by a CF Foundation-accredited Care Center.


**Date of Test** Record the date of the sweat test using the format mm/dd/yyyy. If two or more sweat tests are performed on different dates, record them individually by creating additional segments of sweat test measurement. To enter an estimated date when only the year is known, enter July 1 (07/01) as the date. If the date of a sweat test is known but the result is not, do not record the date.

**Value (mmol/L)** Report all sweat test values in millimols per liter (mmol/L). If the amount of sweat collected is not enough for an accurate result (“quantity not sufficient”), check the adjacent box, “QNS”. If the sweat test value is ≤ 60 mmol/L, then the field below must also be completed. **Note: The range for sweat values is between 1 and 160. Anyone who has a sweat test value that is above 160 must be re-tested.**

If sweat test value <= 60 CF diagnosis was suggested by (select all that apply) Only complete this section if the patient’s sweat test value was less than or equal to 60 mmol/L.

- DNA Analysis/Genotyping
- Transepithelial (Nasal) Potential Difference
- Clinical Presentation (pancreatic fxn tests, Microbiology, etc.)
- Unknown It is not known what indicated a diagnosis of CF.

**Parents’ Information**

Record the patient's biological parents’ heights using inches or centimeters. If either parent’s height is unavailable, select “Not Available”. Measured heights are preferable, but if not available, record reported parental heights.
Not Available: Check if parental heights are not available.

Mother height: Selecting the appropriate unit of measure, enter the patient’s mother’s height in centimeters or inches. If recorded in inches, the value will automatically be converted to centimeters when saved.

Father height: Selecting the appropriate unit of measure, enter the patient’s father’s height in centimeters or inches. If recorded in inches, the value will automatically be converted to centimeters when saved.

Birth Measurements

Baby delivered: Select the term at which the patient was delivered as a baby.

- **Full term** (>= 37 weeks gestational age)
- **Premature** (< 37 weeks gestational age) If the patient was delivered premature, gestational age in weeks must be reported to save the form.
- **Unknown** The gestational age of the patient at delivery is not known.

Specify gestational age: If the patient was delivered premature, you must select the specific gestational age at which the patient was delivered from the options: <=24, 25, 26, 27, 28, 29, 30, 31, 32, 33, 34, 35, 36. If the exact gestational age at which the patient was delivered is unknown, provide your/the patient’s best estimate.

**Birth length:** Selecting the appropriate unit of measure, enter length at birth of the patient in centimeters or inches. If recorded in inches, the value will automatically be converted to centimeters when saved.

**Birth weight:** Selecting the appropriate unit of measure, enter the weight at birth of the patient in kilograms or pounds. If recorded in pounds, the value will automatically be converted to kilograms when saved.

Genotype Information

Has this patient been genotyped? Select Yes or No. If Yes, report the most recent genotyping results. Only documented genotyping results should be entered. If the patient has been re-genotyped (e.g., through the Cystic Fibrosis Mutation Analysis Program), enter the most recent date of genotyping and most recent genotype results. Even if no new mutations are found, update the data of genotyping to reflect that the patient was retested.

**Date of genotyping** Enter the date of the most recent genotyping. If the exact date of genotyping is not known, enter an approximated date and check the adjacent box, “Date is an approximation”.

Select Mutation 1 Select the appropriate mutation from the list provided. All mutations are listed by their legacy names. Visit [http://cftr2.org/browse.php](http://cftr2.org/browse.php) to find legacy names for mutations reported in the new nomenclature (protein and nucleotide). If you are unable to find the patient’s mutation in the list provided and have verified that is not listed under a different name, select Other and specify the mutation in the adjacent “Other Genotype” field. If the lab does not identify a mutation, report Unknown. **Note: be sure to click “Ok” at the top right of the mutation list window in order for the mutation selected to populate “Genotype: “.**

Other Genotype: This field will become available and is a required field if Other is selected in the mutation list window. Record the name of the patient’s mutation as reported in their genotype results here.

Poly T tract (select one of the following): Indicate the poly-T variant of the allele from the options: 5T, 7T, 9T, not 5T and Unknown. These options will only become available if one of the following

Poly TG repeats (select one of the following): Indicate the poly-TG repeat status of the allele from the options: 9, 10, 11, 12, 13, and Other/Unknown/Not done. These options will only become available if one of the following mutations is selected: TG12/T5, TG12, TG10, TG11, R117H, I148T, R117C, M470V, I1027T, L997F, F1052V, R1162L, G622D, or 5T.

Select Mutation 2 Refer to guidelines for “Select Mutation 1” and its subcategories for instructions on how to complete this section.

Select Mutation 3 If applicable, enter the patient’s third mutation and refer to guidelines for “Select Mutation 1” and its subcategories for instructions on how to complete this section. Poly-T tact and poly-TG repeat variant cannot be reported for mutations recorded in this field.

Additional information about genotype not captured above: Record any additional genotype information.
**Encounter Form**

Record all information for clinical encounters with the patient including clinic visits, hospitalizations, and periods of home IV treatment. Encounters with physicians who are not associated with a CF Foundation-accredited Care Center should not be recorded with the exception of outpatient visits with a transplant center within the same hospital system or institution, which can be recorded in PortCF.

Encounters that occur outside of typical clinic visits, hospitalizations, and periods of home IV treatment (e.g., lab visits, visits to the PFT lab) should be recorded with a location of “Other”.

For periods of hospitalization or home IV treatment, a Care Episode should be created and filled out for each period of treatment (which can include multiple segments of hospitalization/home IV administration). Any information collected during a Care Episode that is not collected in the Care Episode Form (i.e., culture results, multiple PFT values, etc.), can be entered into an Encounter. The entry of such Encounters should be in addition to, not in place of, a Care Episode.

**Vital Signs/ Encounter Start**

**Encounter Date:** Record the date of the encounter with the patient using the format mm/dd/yyyy.

**Location:** Record the location of the encounter. If the encounter was during a hospital stay or a home IV, create a care episode for the associated period of hospitalization/Home IV treatment.

- Clinic
- Hospital
- Home IV
- Other Encounters that occur outside of typical clinic visits, hospitalizations, and periods of home IV treatment (e.g., lab visits, visits to the PFT lab) should be recorded with a location of “Other”.

**Start Date:** This field will become enabled if a location of Hospital or Home IV is selected for the encounter. Record the start date of the hospitalization or home IV treatment for the patient using the format mm/dd/yyyy. **Start Date** must be completed in order to save the encounter form as complete.

**End Date:** This field will become enabled if a location of Hospital or Home IV is selected for the encounter. Record the end date of the hospitalization or home IV treatment for the patient using the format mm/dd/yyyy. **End Date** is not a required field for an encounter to be saved as complete.

**Height:** Selecting the appropriate unit of measure, record the patient's height using centimeters or inches. If recorded in inches, the value will automatically be converted to centimeters when saved. **Note:** if the patient’s height has decreased more than 2% since the last height measurement, a height validation warning will appear the in “Errors and Warnings” box. Check the entered height measure if you receive this warning. Verify that the number and units of measurement are correct if you receive this message.

**Height Percentile:** Once the data is saved, the height percentile is calculated if the patient is under the age of 20 at the time of the encounter. CDC percentiles are used to calculate this value.
**Weight:** Selecting the appropriate unit of measure, record the patient's weight using kilograms or pounds. If recorded in pounds, the value will automatically be converted to kilograms when saved.

**Weight Percentile:** Once the data is saved, weight percentile is calculated if the patient is under the age of 20 at the time of the encounter. CDC percentiles are used to calculate this value.

**BMI Value:** Once the data is saved, BMI is calculated for patients age 20 and older at the time of the encounter.

**BMI Percentile:** Once the data is saved, BMI percentile is calculated if the patient is between the ages of 2 and 20 at the time of the encounter. CDC percentiles are used to calculate this value.

**Weight-for-Length Percentile:** Once the data is saved, weight-for-length percentile is calculated if the patient is under the age of 2 at the time of the encounter.

**Exacerbation Assessment**

What was your assessment regarding pulmonary exacerbation at this visit?: An assessment of the patient’s exacerbation status must be recorded. The guidelines below are recommended to determine exacerbation status when the treating physician has not provided an assessment.

<table>
<thead>
<tr>
<th>Absent</th>
<th>Symptoms</th>
<th>Treatment</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Patient is not experiencing any notable increase in pulmonary symptoms.</td>
<td>No treatment is recommended by treating physician for <strong>pulmonary symptoms</strong>.</td>
</tr>
<tr>
<td>Mild exacerbation</td>
<td>Patient is experiencing a mild increase in pulmonary symptoms.</td>
<td>Typically treated with minor changes to the patient’s chronic regime for <strong>pulmonary symptoms</strong> (e.g., greater attention to adherence, increased frequency/duration of airway clearance treatments, etc.). It may also include treatment with an oral and/or inhaled antibiotics.</td>
</tr>
<tr>
<td>Moderate exacerbation</td>
<td>Patient is experiencing a clinically significant increase in pulmonary symptoms, often including decrease in pulmonary function. Patient may also be experiencing constitutional symptoms (e.g., fatigue, poor appetite, weight loss, fever, chills, etc.).</td>
<td>Typically treated with oral and/or inhaled antibiotics for <strong>pulmonary symptoms</strong>. It may also be treated with IV antibiotics.</td>
</tr>
<tr>
<td>Severe exacerbation</td>
<td>Patient is experiencing a clinically significant increase in pulmonary symptoms, often including constitutional symptoms (e.g., fatigue, poor appetite, weight loss, fever, chills, etc.).</td>
<td>Typically treated by hospitalization or home IV treatment for <strong>pulmonary symptoms</strong>.</td>
</tr>
<tr>
<td>Don’t know/unable to answer</td>
<td>The patient's symptoms cannot be determined or are not known.</td>
<td>The treatment of the patient cannot be determined or is not known.</td>
</tr>
</tbody>
</table>
If you determined that an exacerbation was present, please select the treatment course prescribed to treat the exacerbation: If pulmonary exacerbation is reported to be present, specify the course of treatment prescribed, if any. If the patient is hospitalized or prescribed home IV treatment, start a Care Episode – this treatment does not need to be noted as None of the above. Select all that apply.

- Increased airway clearance, exercise, and/or bronchodilators
- Oral NON-quinolone antibiotic (e.g. azithromycin, Bactrim, Augmentin, etc.)
- Oral quinolone antibiotic (e.g. ciprofloxacin (Cipro), levofloxacin)
- Inhaled antibiotic
- Inhaled antibiotic PLUS Oral NON-quinolone antibiotic
- Inhaled antibiotic PLUS an oral quinolone antibiotic
- None of the above

Select only if the prescribed treatment for the current exacerbation is not listed above. If selected, specify any unlisted treatment(s).

Social Worker Consultation

Patient consulted with a Social Worker at this visit: Check if the patient was evaluated by one of the care team’s social workers. Consultations by phone can be reported. DO NOT check if the patient was referred but refused services.

Nutritional

Patient was seen by a Dietitian/Nutritionist at this visit: Check if the patient was evaluated by one of the care team’s dietitians or nutritionists. Consultations by phone can be reported. DO NOT check if the patient was referred but refused services.

Pulmonary

Patient was seen by a Respiratory Therapist/Physical Therapist at this visit: Check if the patient was evaluated by one of the care team’s respiratory or physical therapists. DO NOT check if the patient was referred but refused services.

Other

Notes, comments, or any additional information about the encounter with the patient can be recorded in the custom fields.

Record any notes about the encounter – Custom Field 1
Record any notes about the encounter – Custom Field 2
Record any notes about the encounter – Custom Field 3

Microbiology

Bacterial Culture

Report all cultures performed. If a respiratory culture was not performed or if the laboratory report reads “insufficient specimen”, do not include a microbiology tab, for an encounter cannot be saved as complete with a microbiology tab included unless the “Bacterial culture done?” box is checked.

Bacterial culture done? Check if a bacterial culture was performed.

Date of Culture Record the date the culture was performed using the format of mm/dd/yyyy.
Type of specimen: Record the type of specimen cultured.
- sputum
- induced sputum
- throat/nasal
- bronchoscopy

Culture Results: Record culture results. Use only final laboratory reports to answer this question. If Microorganisms is selected, you will be able to specify those microorganisms cultured in the selection below.

- Microorganisms
- Normal flora Select if only normal flora, including Group A beta hemolytic strep (streptococcus pyogenes), is cultured.
- No growth/sterile culture

Staphylococcus aureus: Check if any form of Staph aureus was cultured. If checked, you must specify resistance. If small colony variant Staph aureus (SCVs) is cultured, check “Other bacterial or fungal species” and specify in the adjacent field.

- MRSA (methicillin resistant Staph aureus) Includes both MRSA (methicillin-resistant Staphylococcus aureus) and ORSA (oxacillin-resistant Staphylococcus aureus).
- MSSA (methicillin sensitive Staph aureus)

Haemophilus influenza (any species) Note: Haemophilus species other than Haemophilus influenzae should not be reported in PortCF. This includes Haemophilus Parainfluenzae

Pseudomonas aeruginosa If selected you must report mucoid status in order to save the encounter as complete. If both mucoid and non-mucoid types are cultured, select both.

- mucoid
- non-mucoid
- Mucoid status unknown Mucoid status is not known.
Susceptibility Testing

If susceptibility testing was not done or is reported as “indeterminant” by the lab for aminoglycosides, quinolones, or beta-lactams, report as “Testing Not Done”. Otherwise, record resistance to all drugs tested (“Yes” or “No”) for each of the categories.

If multiple strains of *P. aeruginosa* are cultured, fill out the susceptibility testing section according to the antimicrobial resistance pattern of the most resistant *P. aeruginosa* strain cultured. For example, if a culture grows two strains of *P. aeruginosa* and one strain is resistant to two classes of antibiotics and the other is resistant to only one, record the strain resistant to two classes.

For data entry purposes, if multiple *P. aeruginosa* strains are resistant to the same number of classes of antibiotics then use the following schema to determine which strain to record: strains resistant to Beta lactams > strains resistant to Quinolones > strains resistant to Aminoglycosides.

**Resistant to All Aminoglycosides Tested (e.g., tobramycin, gentamicin, amikacin)**
Select Yes, No, or Testing Not done. If intermediate resistance is reported, select Yes.

**Resistant to All Quinolones Tested (e.g., ciprofloxacin, levofloxacin, moxifloxacin)**
Select Yes, No, or Testing Not done. If intermediate resistance is reported, select Yes.

**Resistant to All Beta Lactams Tested (e.g., ceftazidime, imipenem, meropenem, piperacillin/tazobactam (Zosyn), ticarcillin/clavulanic acid (Timentin), aztreonam)**
Select Yes, No, or Testing Not done. If intermediate resistance is reported, select Yes.

**Burkholderia species** If reported, you must specify the species and report whether or not the *Burkholderia* species was confirmed at the CFF reference lab in order to save the encounter as complete.

- *B. gladioli*
- *B. cenocepacia*
- *B. multivorans*
- *Burkholderia – other* If selected, select all that apply.
  - *B. cepacia*
  - *B. stabilis*
  - *B. vietamiensis*
  - *B. dolosa*
  - *B. anthina*
  - *B. ambifaria*
  - *B. pyrrocinia*
  - *B. ubonensis*
  - *B. arboris*
  - *B. latens*
  - *B. lata*
  - *B. metallica*
  - *B. seminalis*
  - *B. contaminans*
  - *B. diffusa*
Was the identification of *Burkholderia* species confirmed at the CFF reference lab? Select Yes, No, or Unknown. If any *Burkholderia* species is selected, this must be reported in order to save the encounter as complete.

**Other Microorganisms:** Report any microorganisms cultured that are not listed above. Select all that apply.

- *Alcaligenes (Achromobacter) xylosoxidans* Includes any *Achromobacter* sp. in cases where *Achromobacter xylosoxidans* is suspected but the lab cannot confirm.
- *Stenotrophomonas (Xanthomonas)/Maltophilia*
- *Other types* If selected, select all that apply.
  - Acinetobacter baumannii
  - Acinetobacter species - other*
  - Agrobacterium species
  - Bordetella species
  - Brevundimonas species
  - Chryseobacterium species
  - Cupriavidus metallidurans
  - Cupriavidus pauculus
  - Cupriavidus respiraculi
  - Delftia acidovordans
  - Delftia species – other*
  - Enterobacter species
  - Exophilia dermatitidis
  - Herbaspirillum frisingense
  - Herbaspirillum seropedicae
  - Inquilinus limosus
  - Klebsiella pneumonia
  - Klebsiella species – other*
  - Pseudomonas mendocina
  - Pseudomonas pseudoalcaligenes
  - Pseudomonas putida
  - Pseudomonas stutzeri
  - Pseudomonas species – other*
  - Ralstonia insidiosa
  - Ralstonia picketti
  - Ralstonia species – other*
  - Serratia marcescens
  - Streptococcus milleri

**Fungal/Yeast:** Report any fungi and/or yeast cultured. Select all that apply.

- Aspergillus (any species)
- Candida (any species)
- Scedosporium species

Specify if additional species were found
• Other bacterial or fungal species Check only if bacterial or fungal species cultured are not listed. If checked, specify in the adjacent box all unlisted species (e.g., small colony variant Staph aureus (SCVs), E. coli, etc.) cultured.

Mycobacterial Culture

Was Mycobacterial culture done? Check if a mycobacterial culture was performed. DO NOT check if the specimen tested was a from a throat swab unless a mycobacterial species was cultured.

Date of culture Record the date the culture was performed using the format mm/dd/yyyy.

Type of specimen If reporting the positive results of a mycobacterial culture performed on a throat swab specimen, select sputum as the specimen type and complete the section as normal.

• sputum
• induced sputum
• bronchoscopy

AFB Smear

• Positive
• Negative
• Not Done

Culture Results: Record culture results. Use only final laboratory reports to answer this question. If Microorganisms is selected, you will be able to specify those microorganisms cultured in the selection below.

• Microorganisms
• Normal flora
• No growth/sterile culture

Mycobacterial Species: Select all species cultured.

• Mycobacterial tuberculosis
• Mycobacterium abscessus/chelonea
• Mycobacterium avium complex (MAC) Includes M. avium subsp. avium, M. avium subsp. hominissuis, M. avium subsp. paratuberculosis, and M. intracellulare.
• Mycobacterium fortuitum group
• Mycobacterium gordonae
• Mycobacterium kansasii
• Mycobacterium marinum
• Mycobacterium terrae
• Other Select only if unlisted mycobacterial species are cultured. If selected, you must specify any unlisted mycobacterial species cultured.
  • Specify: You must specify any unlisted mycobacterial species cultured if Other is selected in order to save the encounter as complete.

Medications
At the top of the Pulmonary Medications section is the “Upload Medications” link. Clicking this will automatically add any medications and dosage/frequency information currently entered on the patient’s most recent Medications tab. When using this functionality, be sure to update the medications selected as necessary to reflect any changes in regimen including any changes to dosage and newly prescribed or discontinued medications.

Not on Medications

This patient is not on any of the pulmonary medications below Check if the patient is not currently prescribed any pulmonary medications.

Pulmonary Medications

PortCF is not designed to measure patient adherence and as a result is concerned with prescribing practices, not patient adherence, of pulmonary medications.

Record all medications prescribed, not just those considered chronic unless they have been prescribed solely for the acute treatment of a pulmonary exacerbation.

Only report medication use that can be confirmed. For example, medications prescribed as part of an open-label clinical trial, when you it is certain that the patient received the medication, should be reported. In such circumstances, record medication use as close to the start date as possible. In contrast those medications prescribed as part of a phase II or phase III clinical trial, when you are not certain if the patient received the medication or placebo, should not be reported.

Medications prescribed for the eradication of a newly acquired microorganism (such as P. aeruginosa) should be reported. When reporting antibiotics prescribed for the eradication of a microorganism, select Other regimen (different dose or freq) when this frequency option is available. Medications prescribed for eradication between Encounters should be reported on the encounter associated with the culture that identified the microorganism for eradication.

For medications prescribed between encounters, enter the newly prescribed medication on the patient’s next encounter.

For medications prescribed with an alternating month treatment schedule, report the medication prescribed even if the patient is on an “off” month at the time of the encounter.

Chronic antibiotics (i.e. not prescribed to treat exacerbation) – inhaled and/or oral

- **Tobramycin solution for inhalation (i.e. TOBI)**: Select if TOBI® or a generic equivalent (300mg/5mL) is prescribed. Any other unlisted inhaled tobramycin solution should be recorded as *Other inhaled aminoglycoside*. Report even if patient is on an “off” month at the time of the encounter.
Frequency: Select the prescribed dosage and frequency from the options provided. This field is required if Tobramycin solution for inhalation (i.e. TOBI) is selected. If prescribed for eradication, report Other regimen (different dose or freq).
- 300 mg BID alternate month schedule
- 300 mg BID continuous
- Other regimen (different dose or freq) Includes prescription for the eradication of microorganisms.

- Tobi Podhaler (Tobramycin Inhalation Powder): Select if TOBI® Podhaler™ is prescribed. Any other unlisted inhaled tobramycin solution should be recorded as Other inhaled aminoglycoside. Report even if patient is on an “off” month at the time of the encounter.
  - Frequency: Select the prescribed dosage and frequency from the options provided. This field is required if Tobi Podhaler (Tobramycin Inhalation Powder) is selected. If prescribed for eradication, report Other regimen (different dose or freq).
    - Four 28mg capsules BID every other month
    - 300 mg BID continuous
    - Other regimen (different dose or freq) Includes prescription for the eradication of microorganisms.

- Bethkis: Select if Bethkis® or a generic equivalent (300mg/4mL) is prescribed. Any other unlisted inhaled tobramycin solution should be recorded as Other inhaled aminoglycoside. Report even if patient is on an “off” month at the time of the encounter. If prescribed for eradication, report Other regimen (different dose or freq).
  - Frequency: Select the prescribed dosage and frequency from the options provided. This field is required if Bethkis is selected.
    - 300 mg BID alternate month schedule
    - Other regimen (different dose or freq) Includes prescription for the eradication of microorganisms.

- Other inhaled aminoglycoside (e.g. gentamicin, amikacin, or tobramycin preparation): Select if an inhaled aminoglycoside has been prescribed including any unlisted inhaled tobramycin solutions. Report even if patient is on an “off” month at the time of the encounter. If prescribed for eradication, report Other regimen (different dose or freq).
  - Frequency: Select the prescribed frequency from the options provided. This field is required if Other inhaled aminoglycoside is selected.
    - Alternate Month
    - Continuous
    - Other regimen (different dose or freq) Includes prescription for the eradication of microorganisms.

- Colistin: Record even if patient is on an "off" month at the time of the encounter.
  - Frequency: Select the prescribed frequency from the options provided. This field is required if Colistin is selected. If prescribed for eradication, report Other regimen (different dose or freq).
    - Alternate Month
    - Continuous
    - Other regimen (different dose or freq) Includes prescription for the eradication of microorganisms.

- Aztreonam – Inhaled: Record even if patient is on an "off" month at the time of the encounter.
  - Frequency: Select the prescribed frequency and dosage from the options provided. This field is required if Aztreonam - Inhaled is selected. If prescribed for eradication, report Other regimen (different dose or freq).
    - 75 mg TID Alternate Month Schedule
- 75 mg TID Continuous Schedule
- Other regimen (different dose or freq) Includes prescription for the eradication of microorganisms.

- **Chronic oral macrolide antibiotic**
  - azithromycin (Zithromax)
  - clarithromycin (Biaxin)

- **Other chronic oral antibiotic**: If select, select all that apply. You must specify the antibiotic type if Other chronic oral antibiotic is selected in order to save the encounter form as complete.
  - Quinolone (Cipro, Levaquin, gatifloxacin, etc.)
  - Cephalosporin (cephalexin, Keflex, cefixime, etc.)
  - Sulfax (Bactrim, Septra, etc.)
  - Amoxicillin (Augmentin, etc.)
  - Tetracycline (doxycycline, Vibramycin, minocycline, etc.)
  - Other: Select only if the prescribed chronic oral antibiotic is not listed.

**CFTR Modulators**

- **Ivacaftor (i.e. Kalydeco, VX-770)**
  - Frequency: Select the prescribed dosage from the options provided. This field is required if Ivacaftor is selected.
    - 150 mg BID
    - Other regimen (different dose or freq)

**Other Medications**

- **Dornase alfa (i.e. Pulmozyme)**
  - Frequency: Select the prescribed dosage and frequency from the options provided. This field is required if Dornase alfa is selected.
    - 2.5 mg QD
    - 2.5 mg BID
    - Other regimen (different dose or freq)

- **Acetylcysteine or Mucomist**

- **High-dose ibuprofen (e.g. 25-30 mg/kg)**: If selected, record the total mg/dose prescribed, not the number of mg per kg.

- **Hypertonic saline**
  - Concentration (%): Select the prescribed concentration. Choose from the following options in the adjacent dropdown list: 3, 4, 5, 6, 7, 8, 9, 10. This is a required field if Hypertonic saline is selected.
  - Frequency: Select the prescribed frequency from the options provided. This field is required if Hypertonic saline is selected.
    - QD
    - BID
    - Other

**Bronchodilators (oral):**

- **Beta agonist (e.g. Proventil Repetabs, Volmax, etc.)**
- **Theophylline product (e.g. Theodur, Slo-bid, Uniphyll)**
Bronchodilator (inhaled):

- Short acting beta agonist (e.g. albuterol, Proventil, Ventolin, Xopenex, etc.)
- Long acting beta agonist (e.g. salmeterol, Sevevent, Foradil, Brovana, etc.)
- Short acting anticholinergic (e.g. ipratropium, Atrovent)
- Long acting anticholinergic (e.g. tiotropium, Spiriva, etc.)
- Combination beta agonist and anticholinergic (e.g. Combivent, DuoNeb, etc.)

Corticosteroids:

- Oral (e.g. prednisone)
- Inhaled (e.g. fluticasone, Flovent, budesonide, Pulmicort, etc.)
- Inhaled in combination with a bronchodilator (e.g. Advair, Symbicort)

Other

- Leukotriene modifiers (e.g. montelukast, Singulair, zafirlukast, Accolate, zileuton, Zyflo, etc.)
- Mast cell stabilizers (e.g. cromolyn, Intal, nedocromil, Tilade, etc.)
- Antifungals (e.g. itraconazole, Sporanox) Do not select if the antifungals prescribed are topical agents for the treatment of skin conditions or agents used for the treatment of oral thrush.

Drug Intolerance/Allergies: Record any drug intolerances and/or allergies the patient has. **Note: This is not an all-inclusive list and the patient may have allergies and drug intolerances to medications other than those listed here.**

- Dornase alfa (i.e. Pulmozyme)
- Tobramycin solution for inhalation (i.e. TOBI)
- Aztreonam
- Colistin
- Macrolide antibiotics
- High-dose ibuprofen
- Hypertonic saline

GI/Nutrition/Endocrine Medications

Unlike pulmonary medications, report enzyme use according to what the patient reports taking on a typical day, not what is prescribed/recommended, when this information is available.

**This patient is on enzyme medications:** Select Yes or No. If yes, record enzyme use as reported by the patient.

**Enzymes**

The guidelines for entry of the fields *Number of capsules per largest meal of the day* and *Total capsules per day* apply to all listed enzymes according to dosage. If a listed enzyme is selected, the completion of these fields is required in order to save the encounter as complete.

**Enzyme Reporting**

Updated 10/08/2014
Number of capsules per largest meal of the day: Choose from the following options in the adjacent dropdown list: 1, 2, 3, 4, 5, 6, 7, 8, 9, 10, 10+. If patient is takes enzymes in .5 capsule increments, round up to the next whole number (i.e., if the patient takes 4.5 capsules of Zenpep 15 per largest meal, report 5 capsules).

Total capsules per day: Record in the adjacent box the total number of capsules taken per day. This should be the total number of capsules taken by the patient on a typical day.

Creon

- **Creon 1203** Refer to guidelines for Enzyme Reporting and it subcategories for instructions on how to complete this section. *5 capsule size is available in the drop down list for this dosage. Do not round up to 1 capsule if patient is prescribed .5 capsule per largest meal.
- **Creon 1206** Refer to guidelines for Enzyme Reporting and it subcategories for instructions on how to complete this section. *5 capsule size is available in the drop down list for this dosage. Do not round up to 1 capsule if patient is prescribed .5 capsule per largest meal.
- **Creon 1212** Refer to guidelines for Enzyme Reporting and it subcategories for instructions on how to complete this section.
- **Creon 1224** Refer to guidelines for Enzyme Reporting and it subcategories for instructions on how to complete this section.
- **Creon 1236** Refer to guidelines for Enzyme Reporting and it subcategories for instructions on how to complete this section.

Pancreaze

- **Pancreaze MT4** Refer to guidelines for Enzyme Reporting and it subcategories for instructions on how to complete this section. *5 capsule size is available in the drop down list for this dosage. Do not round up to 1 capsule if patient is prescribed .5 capsule per largest meal.
- **Pancreaze MT10** Refer to guidelines for Enzyme Reporting and it subcategories for instructions on how to complete this section.
- **Pancreaze MT16** Refer to guidelines for Enzyme Reporting and it subcategories for instructions on how to complete this section.
- **Pancreaze MT20** Refer to guidelines for Enzyme Reporting and it subcategories for instructions on how to complete this section.

Ultresa

- **Ultresa 14** Refer to guidelines for Enzyme Reporting and it subcategories for instructions on how to complete this section. *5 capsule size is available in the drop down list for this dosage. Do not round up to 1 capsule if patient is prescribed .5 capsule per largest meal.
- **Ultresa 20** Refer to guidelines for Enzyme Reporting and it subcategories for instructions on how to complete this section.
- **Ultresa 23** Refer to guidelines for Enzyme Reporting and it subcategories for instructions on how to complete this section.

Updated 10/08/2014
Pertzye (Pancrecarb)

- **Pertzye 4000** Refer to guidelines for Enzyme Reporting and it subcategories for instructions on how to complete this section. *5 capsule size is available in the drop down list for this dosage. Do not round up to 1 capsule if patient is prescribed .5 capsule per largest meal.*
- **Pertzye 8000** Refer to guidelines for Enzyme Reporting and it subcategories for instructions on how to complete this section.
- **Pertzye 16000** Refer to guidelines for Enzyme Reporting and it subcategories for instructions on how to complete this section.

Zenpep

- **Zenpep 3** Refer to guidelines for Enzyme Reporting and it subcategories for instructions on how to complete this section. *5 capsule size is available in the drop down list for this dosage. Do not round up to 1 capsule if patient is prescribed .5 capsule per largest meal.*
- **Zenpep 5** Refer to guidelines for Enzyme Reporting and it subcategories for instructions on how to complete this section. *5 capsule size is available in the drop down list for this dosage. Do not round up to 1 capsule if patient is prescribed .5 capsule per largest meal.*
- **Zenpep 10** Refer to guidelines for Enzyme Reporting and it subcategories for instructions on how to complete this section.
- **Zenpep 15** Refer to guidelines for Enzyme Reporting and it subcategories for instructions on how to complete this section.
- **Zenpep 20** Refer to guidelines for Enzyme Reporting and it subcategories for instructions on how to complete this section.
- **Zenpep 25** Refer to guidelines for Enzyme Reporting and it subcategories for instructions on how to complete this section.

Viokace

- **Viokace 10** Refer to guidelines for Enzyme Reporting and it subcategories for instructions on how to complete this section. *5 capsule size is available in the drop down list for this dosage. Do not round up to 1 capsule if patient is prescribed .5 capsule per largest meal.*
- **Viokace 20** Refer to guidelines for Enzyme Reporting and it subcategories for instructions on how to complete this section.

Other enzymes

Please specify if other enzymes: Record any enzymes and their corresponding dosages (the number of capsules per largest meal of the day and the total number of capsules taken per day) not listed above in the “Enzymes” section.

Acid Blocker

Acid Blocker (Daily use. Check all that apply since last visit)
- *H2 Blocker* (e.g. Zantac, Pepcid, etc.)
- *Proton Pump Inhibitor* (e.g. Prilosec, Nexium, etc.)
- *Unknown* It is now known whether or not the patient uses acid blockers.

**GI Other**

**Ursodeoxycholic acid** Check if patient is prescribed ursodeoxycholic acid.

**PFTs**

**Pulmonary**

**PULMONARY FUNCTION TESTS (PFTs)**

Only report measurements from pulmonary function tests (PFTs) that have been performed according to the current standards issued by the American Thoracic Society (ATS). Do not enter PFTs results from PFTs not meeting ATS standards. For patients, including those under age 6, who perform PFTs but are unable to meet ATS standards, include this tab and report “Unable to perform test”.

PFT measurements from outside laboratories are acceptable if a Care Center physician reviews the measurements and verifies their validity before they are entered into PortCF.

**Pre-bronchodilator** PFT measurements should be always be entered unless the only measurements taken/available are post-bronchodilator measurements (i.e., if pre- and post-bronchodilator measurements are taken, enter pre-bronchodilator values).

For patients less than six years old, PFT results should be entered if they meet the above criteria. However, predicted values will not be calculated due to age.

For patients who are unable to perform PFTs, include this tab and report “Unable to perform test”. This is only necessary for patients 6 years or older.

The date of encounter and patient's height and unit of measure must be entered in the General Encounter Start for predicted and percent predicted values to be calculated.

**Unable to perform test:** Check if the patient is unable to perform PFTs or is unable to perform them according to ATS standards. If selected, you must specify the reason test results are not recorded or why the patient was unable to perform the test.

Reason why PFTs have not been done: If "Unable to perform test" is checked, you must enter in the adjacent field the reason why PFTs were not done/are not recorded.

**FVC – measure (L):** Record the FVC measure in liters (L) up to the hundredths place. This is a required field if “Unable to perform test” is not reported.
• **Predicted Value:** Once the raw FVC value is saved, if the patient is age 6 or older and height is entered for the encounter, the predicted value is calculated and displayed. The author of the equation along with the variables (i.e., race, gender, age) used to calculate the predicted value will be displayed under this value.

• **% predicted:** Once the raw FVC value is saved, if the patient is age 6 or older, the percent predicted will be calculated and displayed.

• **Relative change (to previous measure):** Once the raw FVC value is saved, the relative change (to previous measure) – percent – from the last entered measure will be calculated and displayed, if applicable. The number of days since the last measure (L) will be displayed under this value.

**FEV₁ – measure (L):** Record the FEV₁ measure in liters (L) up to the hundredths place. The FEV₁ value cannot be larger than the FVC value. If the FEV₁ measure entered is larger than the FVC entered, you will be unable to save the form until the measure is corrected. This is a required field if “Unable to perform test” is not reported.

• **Predicted Value:** Once the data is saved, if the patient is age 6 or older and height is entered for the encounter, the predicted value is calculated. The author of the equation along with the variables (i.e. race, gender, age) used to calculate the predicted value will be displayed under this value.

• **% predicted:** Once the raw FEV₁ value is saved, if the patient is age 6 or older, the percent predicted percentile is calculated.

• **Relative change (to previous measure):** Once the raw FEV₁ value is saved, the relative change (to previous measure) – percent – from the last entered FEV₁ measure (L) will be calculated and displayed, if applicable. The number of days since the last measure (L) will be displayed under this value.

**FEF25-75 – measure (L):** Record the FEF25-75 measure in liters (L) up to the hundredths place.

• **Predicted Value:** Once the data is saved, if the patient is age 6 or older and height is entered for the encounter, the predicted value is calculated. The author of the equation along with the variables (i.e. race, gender, age) used to calculate the predicted value will be displayed under this value.

• **% predicted:** Once the data is saved, if the patient is age 6 or older, the percent predicted percentile is calculated.

**CF specific FEV₁ percentile (ages 6-21):** Once the raw FEV₁ value is saved, if the patient is age 6 to 22 at the time of the encounter and a height is entered for the encounter, the CF-specific FEV₁ percentile will be calculated.

**GI/Nutrition**

**Assessment of oral intake:** Select Done or Not done. Includes any conscious effort to assess the patient’s caloric intake.

**Is patient currently receiving supplemental feeding?** Select Yes, Not, or Unknown. If Yes, record all forms of supplemental feeding the patient received at home since last clinic encounter.

- **Feeding:** If “Is patient currently receiving supplemental feeding?” is reported as Yes, you must specify the type of supplemental feeding in order to save the encounter as complete. If the patient receives tube feedings, distinguish which type.

  - **oral supplementation (Scandishakes, Pediasure, Instant Breakfast, etc.):** Oral supplementation includes additions to meals and snacks such as half ’n half and other
high calorie foods consumed on a regular basis. Vitamin and appetite stimulant use should not be reported as oral supplementation.

- nasogastric tube (NG)
- gastrostomy tube/button (G-tube)
- jejuna tube (J-tube)
- total parenteral nutrition (TNP)

**CF-specific vitamins (i.e. with additional vitamins A, D, E, and K):** Select Yes or No. Report according to vitamin use reported by the patient. Yes should only be selected if the patient is taking prescribed CF-specific vitamins (e.g., aquadeks) or is taking vitamin supplements of A, D, E, and K per prescription.

**Infants under 2 years of age**

Complete this section for patients under the age of 2.

**Salt supplementation:** Select Yes or No. Report according to salt supplement use reported by the patient.

**Select type of feeding:** Report the type of feeding accounting for the majority of the patient’s consumption. If *Breast milk plus formula, Formula exclusively, Other food, or Unknown* is reported, the type and caloric density must be specified in order to save the form as complete.

- *Breast milk*
- *Breast milk plus formula*
- *Formula exclusively*
- *Other food*
- *Unknown* Feeding type is not known.

**If receiving any formula feeding, select type of formula and caloric density:** Report the type of formula accounting for the majority of the patient’s consumption.

- *Cow’s milk*
- *Soy milk*
- *Predigested*
- *Other* Select only if the formula type is not listed. If selected, specify the formula type in the adjacent field.

**Caloric density:** Report the caloric density of formula accounting for the majority of the patient’s consumption.

- 20 cal/oz
- 22 cal/oz
- 24 cal/oz
- 27 cal/oz
- 30 cal/oz
- *Other, specify* Select only if the caloric density is not listed. If selected, you must specify the caloric density in order to save the encounter as complete. If the caloric density is not known, type “Caloric density unknown”.

**Complications**

Report all complications the patient was experiencing at the time of the Encounter, being treated for, or which have occurred since the last Encounter entered into PortCF.
At the top of the Complications Form is the “Upload Complications” link. Clicking this will automatically add all complications and dosage/frequency information currently entered into the most recent Complications Form for the patient. When using this functionality, be sure to update the complications selected as necessary to reflect any changes including the addition of new active complications or removal of resolved complications.

**Patient does not have any complications** Check if patient has no complications.

**CFRD Status**

*Impaired Glucose Tolerance (FBG < 126, 2-h PG 140-199)*

*CFRD with or without fasting hyperglycemia* Includes chronic and intermittent CFRD (with or without fasting hyperglycemia) regardless of whether or not the patient is receiving treatment. Does not include impaired glucose tolerance (i.e., glucose between 140-200 mg/dL on an oral glucose tolerance test).

**CFRD secondary complications:** If *CFRD with or without fasting hyperglycemia* is reported, select all complications secondary to CFRD.

- Retinopathy
- Microalbuminuria
- Chronic renal insufficiency
- Chronic renal failure requiring dialysis
- Peripheral neuropathy

**Hepatobiliary**

*Gall stones*

*Gall stones, requiring surgery/procedure*

*Liver disease, cirrhosis*  
Please specify complications related to cirrhosis: If *Liver disease, cirrhosis* is reported, select all complications secondary to cirrhosis.

- Esophageal varices
- Gastric varices
- GI bleed related to varices
- Splenomegaly
- Hypersplenism (i.e., WBC < 3.0 or platelets < 100,000)
- Ascites

*Liver disease, non-cirrhosis* Includes viral hepatitis.

**Hepatic Steatosis**

*Liver disease, other*  If selected, specify the unlisted liver disease type in the adjacent field. If a patient has elevated Liver Function Test (AST, ALT, and/or GGTP) values greater than 2 times the specified Upper Limit of Normal (ULN), report “Elevated LFT’s”.

**Bone/Joints**

*Arthritis/Arthropathy*

*Bone fracture*
Osteopenia Diagnosed by bone densitometry (DXA).
Osteoporosis Diagnosed by bone densitometry (DXA).

Pulmonary

Allergic Bronchial Pulmonary Aspergillosis (ABPA) ABPA, for data entry purposes, should be considered resolved when the disease is dormant for more than one year without therapy.
Asthma Diagnosed by physician.
Hemoptysis, massive Expectoration of greater than or equal to 1 cup (240ml) of bright red blood in a 24 hour period.
Pneumothorax requiring chest tube

GI

Distal intestinal obstruction syndrome (DIOS, Meconium ileus equiv.) Previously called meconium ileus equivalent in older children and adults. If the physician is confident of the diagnosis, i.e., typical symptoms that respond to treatment, then report DIOS whether hospitalized or not.
Fibrosoing colonopathy/colic stricture (report incidence only) Report incidence only requiring surgery in the current year.
GERD (Gastro-Eosophageal Reflux Disease) Heartburn symptoms responsive to medications.
GI Bleed req hosp non variceal Any acute GI bleed not related to esophageal or gastric varices.
History of intestinal or colon surgery
Pancreatitis
Peptic ulcer disease Report only peptic ulcers confirmed by radiologic studies or endoscopy.
Rectal prolapse

Other Complications

Absence of Vas Deferens
Anxiety Disorder Clinical diagnosis of moderate to severe anxiety disorder of any type. Do not report mild or situational episodes of anxiety.
Cancer confirmed by histology
Depression Clinical diagnosis of moderate to severe depression. Do not report mild or situational episodes of depression.
Hearing loss Verified by a hearing test.
Hypertension
Kidney Stones
Nasal polyps requiring surgery
Renal failure requiring dialysis (cause other than CFRD)
Sinus Disease (symptomatic)

Complications Not Listed Above

Enter additional complications Record any unlisted complications.

Lab

Blood Counts
WBC count x1,000/microL (Typical clinical value: 3.0 to 30.0)
Platelet Count x1,000/microL (typical clinical value: 100 to 500)
Hemoglobin (grams per deciliter)

**Serum Creatinine**

Serum Creatinine Level (mg/dL)

**Liver Function Tests (LFT)**

Alanine Aminotransferase (ALT or SGPT), IU/L
GGTP (gamma glutamyl transpeptidase), IU/L

**Glucose Test**

Random blood glucose (mg/dL)
Fasting blood glucose (mg/dL)

**If OGTT performed:**

OGTT Fasting glucose level (mg/dL)
2 hour (mg/dL)

**Hemoglobin A1C (Hgb A1C)**

Hgb A1C value, %

**Fecal Elastase**

Fecal Elastase Value (microg/g of stool)

**ACT/Exercise**

**Primary Airway Clearance Technique (ACT)**

Record the primary Airway Clearance Technique (ACT) the patient reports using, not what is prescribed/recommended, when this information is available. Only one form of ACT may be selected.

- Positive Expiratory Pressure (PEP)
- Postural drainage with clapping (CPT)
- Forced expiratory techniques (e.g. autogenic drainage, huff cough, active cycle breathing)
- Oscillating PEP (e.g. Flutter, acapella, IPV)
- High frequency chest wall oscillation (e.g. Vest)
- Exercise
- None
• Other Select only if the primary form of ACT used is not listed. If selected, specify the form used in the adjacent field.
  o Specify if other technique: Specify the primary Airway Clearance Technique used if Other is selected.

Secondary Airway Clearance Technique (ACT)

Record any other Airway Clearance Technique(s) (ACT) the patient reports using. Multiple forms of ACT, or none, may be selected.

• Positive Expiratory Pressure (PEP)
• Postural drainage with clapping (CPT) Includes the use of a hand-held percussor since no “Other, specify” field is available for secondary forms of ACT.
• Forced expiratory techniques (e.g. autogenic drainage, huff cough, active cycle breathing)
• Oscillating PEP (e.g. Flutter, acapella, IPV)
• High frequency chest wall oscillation (e.g. Vest)
• Exercise
Care Episode Form

All periods of hospitalization and home IV treatment, even those not occurring at your Care Center when possible, should be recorded in a Care Episode with the exception of IV treatment for Non-Tuberculous Mycobacteria (NTM). If a patient has a clinic visit and is admitted into the hospital from that visit or is prescribed home IV treatment, report data for the clinic visit in an Encounter Form and data for the hospitalization/home IV in a Care Episode.

Care Episode

Care Episode Segment

This section is a repeat group. As a result, multiple segments of care can be created, edited, and deleted as appropriate (i.e., you can record periods of hospitalization and/or home IV administration of an episode of care as individual segments). Separate segments should be created if the location of care (hospital or home IV) or reasons for hospitalization or home IV treatment change.

Only edit or delete an already existing Care Episode Segment in the event of incorrectly or incompletely entered information. Otherwise, create a new entry by clicking “Create Next Segment” at the bottom of the section.

For more information about how to create, edit, and delete repeat groups on the Care Episode Form, visit https://portcf2dev.cff.org/RegistryLaunch/Documents/Manual/Manual.html#_Entering_and_Altering.

Start Date Record the start date of the period of hospitalization or home IV treatment using the format mm/dd/yyyy.

End Date Record the end date of the period of hospitalization or home IV treatment using the format mm/dd/yyyy.

Location Record the type of treatment/location of treatment for the segment of care.

- Hospital
- Home IV

Reasons Select all reasons for the initiation of hospitalization/home IV treatment.

- Pulmonary Exacerbation
- Pulmonary Complication Other Than Exacerbation Includes massive hemoptysis, pneumothorax, asthma, and/or ABPA flare.
- GI Complications
• Transplant related
• Sinus infection
• Non-transplant surgery Includes outpatient surgeries.
• Other Select only if the reason(s) for hospitalization/home IV treatment are not listed. If selected, specify any unlisted reason(s) for hospitalization or home IV treatment.
  o Please specify reason: You must specify the reason for hospitalization or home IV treatment if Other is selected in order to save the Care Episode as complete.

Care Episode Measurements

Import data at the beginning of the care episode from the encounter: Dates for encounters that occurred in the 3 months prior to the start date of the first segment of a Care Episode will populate in the drop down menu. By selecting a date from this menu, the “At the beginning of Care Episode” measurement fields will populate with the corresponding values from the selected encounter. Care Episode values should be directly entered in the event of a lack of relevant measurements being available in existing Encounters.

Import data at the end of the care episode from the encounter: Dates for encounters that occurred between the start date of the Care Episode and the current date will populate in the drop down menu. By selecting a date from this menu, the “At the end of Care Episode” measurement fields will populate with the corresponding values from the selected encounter. Care Episode values should be directly entered in the event of a lack of relevant measurements being available in existing Encounters.

At the beginning of Care Episode/At the end of the Care Episode:

FVC – measure (L) Refer to the guidelines for entering FVC measurements found in the Encounter Form section. This field is required in order to save the encounter form as complete if “Check if data were impossible to measure” is not checked.

FEV₁ – measure (L) Refer to the guidelines for entering FEV₁ measurements found in the Encounter Form section. This field is required in order to save the encounter form as complete if “Check if data were impossible to measure” is not checked.

FEF25-75 – measure (L) Refer to the guidelines for entering FEF25-75 measurements found in the Encounter Form section.

Height Record the patient’s heights using inches or centimeters. If recorded in inches, it will automatically be converted to centimeters when saved. This field is required in order to save the encounter form as complete if “Check if data were impossible to measure” is not checked.

Weight Record the patient’s weight using pounds or kilograms. If recorded in pounds, it will automatically be converted to kilograms when saved.

Date recorded Record the date the above values were measured using the format mm/dd/yyyy. This field is required in order to save the encounter form as complete if “Check if data were impossible to measure” is not checked.

Check if data were impossible to measure: Check if measurements were not collected/impossible to collect.

Comments Any additional notes/comments relevant to the Care Episode can be recorded here. This field is not specific to the start or end of the Care Episode.
Phone Note

Any notable events, communications with the patient, or changes to a patient’s regimen that do not occur at the time of an Encounter or Care Episode can be recorded in a phone note, including but not limited to, phone conversations, voicemails, emails, and Emergency Room visits.

Note

**Note Date** Record the date of correspondence with the patient using the format mm/dd/yyyy.

**Reason for the note** Select the method of communication/interaction with the patient.

- *Phone Call*
- *Electronic message*
- *Other* Patient was in contact with the center by means other than those listed above, or the patient received emergency care but was not admitted. This includes known changes in medications when not made at the time of a phone call or electronic message with the patient.

**Comments** Record any comments about the interaction with the patient.

Medications

Refer to the guidelines for entering medications found in the Medication section of the Encounter Form.

Symptoms

Refer to the guidelines for entering pulmonary exacerbation assessments and treatment found in the Vital Signs/Encounter Start section of the Encounter Form.
Annual Review

Annual Reviews are to be completed once per year for every patient seen at a Care Center and include a number of pre-populated fields based on data entered throughout the year. Any care known to have been received by the patient should inform the completion of this form regardless of where care was received.

For patients seen at multiple centers (i.e., shared patients), each center will enter their own Annual Review for the patient.

Annual Review

Patient Statistics

Number of Encounters recorded by Center: The number of Encounters recorded for the current reporting year at your center are calculated and displayed here.
Number of Encounters recorded by other Care Centers: The number of Encounters recorded for the current reporting year at any other center the patient was seen at are calculated and displayed here.
Number of Care Episodes recorded by Care Center: The number of Care Episodes recorded for the current reporting year at your center are calculated and displayed here.
Number of Care Episodes recorded by Other Care Centers: The number of Care Episodes recorded for the current reporting year at any other center the patient was seen at are displayed here.

Demographics Update

Current Zip: Record the zip code of the patient’s current primary residence. Record the last known zip code if the zip code of the patient’s current primary residence is not known.
Patient is alive/Patient died on: mm/dd/yyyy This is a calculated field. If the patient does not have a death date entered “Patient is alive” will be displayed. If a death date has been entered for the patient, “Patient died on: mm/dd/yyyy” will be displayed and the date will be populated according to the death date entered.

Pulmonary

Did this patient use oxygen therapy during the reporting year?: If the patient used varying dosages of oxygen therapy during the reporting year, report the dose used by the patient the majority of the time when using oxygen therapy. Oxygen therapy includes extra oxygen delivered to the patient through nasal cannula, a face mask, or transtracheally.

- Yes, Continuously Oxygen therapy was used continuously (i.e., during the day and night regardless of physical activity).
- Yes, Nocturnal and/or with exertion Oxygen therapy was used at night and/or during times of physical exertion.
- Yes, During exacerbation Oxygen therapy was only used during a period(s) of pulmonary exacerbation.
- Yes, prn Oxygen therapy used pro re nata (PRN), as needed.
- No Oxygen therapy was not used at any point during the reporting year.
• Unknown Select if it is not known whether or not the patient used oxygen therapy during the reporting year.

Did this patient use non-invasive ventilation during the reporting year (i.e., assisted breathing, BiPap, CPAP, etc.)? Select Yes, No, or Unknown.

Was a Chest X Ray performed during the reporting year? Select Yes, No, or Unknown. If a CT scan was performed in lieu of a chest x-ray, select Yes.

Did the patient receive an influenza vaccination this season (Sept through Jan)? Select Yes, No, Allergic/Refused, or Unknown.

Mycobacterial culture:
According to the Encounters a mycobacterial culture has been performed for the patient in this reporting year. This response is populated according to data entered in the current reporting year at your center. If a mycobacterial culture was recorded during the reporting year, Yes will automatically be selected. If no mycobacterial cultures are recorded during the reporting year, No will automatically be selected.

Please check to confirm that information about mycobacterial culture above is correct. If it is incorrect, please return to the encounter forms and enter correct information. Please note that it is important to enter mycobacterial tests even if no mycobacterial organisms were found. Check the adjacent box if the automatically populated response to “According to the Encounters a mycobacterial culture has been performed for the patient in this reporting year” is accurate. If incorrect, correct data entered into PortCF.

Was treatment INITIATED for a pulmonary mycobacterial infection during this reporting year? This question cannot be answered and is read-only unless a mycobacterial culture(s) was performed during the reporting year and the culture result was Microorganisms (not Normal flora or No growth/sterile culture). If microorganisms were cultured, select Yes, No, or Unknown.

Was an IgE screening for ABPA performed in this reporting year? Select Yes, No, or Unknown.

Did this patient smoke cigarettes during the reporting year? Includes the smoking of any tobacco products, not just cigarettes (e.g., cigars, hookah). If the patient smoked with varying frequency during the reporting year, report the frequency at which the patient smoked for the majority of the reporting year.

• No
• Occasionally
• Yes, Regularly, less than 1 ppd
• Yes, Regularly, 1 ppd or more
• Declined to answer
• Not Known
• Not Applicable

Does anyone in the patient’s household smoke cigarettes? Select Yes, No, or Unknown. Includes the smoking of any tobacco products, not just cigarettes (e.g., cigars, hookah).

During the reporting year, how often was this patient exposed to secondhand smoke? Includes the smoke of any tobacco products, not just cigarettes (e.g., cigars, hookah). If the patient was exposed to secondhand smoke at any point during the reporting year, report the frequency of secondhand smoke exposure the patient experienced the majority of the reporting year.

• Daily
• Several Times Per Week
• Several Times Per Month or less
• Never
• Declined to answer
• Not Known

Growth and Nutrition

Fat soluble vitamin levels measured? Select Yes, No, or Unknown.
Has this patient been on growth hormone in the reporting year? Select Yes, No, or Unknown.
Was a DEXA scan for bone density performed in this reporting year? Please enter findings of osteoporosis or osteopenia into the complications section of last patient encounter. Select Yes, No, or Unknown.

Results of DEXA Scan: If z-scores (non-diagnostic) are reported, report the diagnosis of the treating physician.

• Normal
• Osteopenia
• Osteoporosis
• Other Select only if the DXA scan results are not listed.
• Unknown The results of the DXA scan are not known.

Update on CFRD Status

CFRD status from most recent encounter ([cannot be determined], [does not indicate CFRD], [indicates impaired glucose tolerance], [indicates CFRD]). This response is populated according to the CFRD status indicated on the patient’s most recent Encounter form in the reporting year. If the patient’s CFRD status was not reported on an Encounter in the year, complete this section according to the patient’s clinical diagnosis even if it is not supported by lab values.

• Normal Glucose Metabolism (includes normal, random, fasting, or OGTT) Select if the patient is assumed to have a normal glucose metabolism even if the patient did not undergo OGTT or glucose testing during the year.
• Impaired Glucose Tolerance (FBG < 126, 2-h PG 140-199)
• CF-related diabetes with or without fasting hyperglycemia (2-h PG >= 200)

Was a retinal eye exam performed by an ophthalmologist in this reporting year? Select Yes, No, or Unknown.
Was a spot urine sent for albumin/creatinine ratio in this reporting year? Select Yes, No, or Unknown.
Was the patient prescribed treatment for CFRD? Select Yes, No, or Unknown.
Select all that apply:

• Dietary change
• Oral hypoglycemic agents
• Intermittent insulin (with illness, steroids, etc.)
• Chronic insulin

Did the patient experience any episodes of severe hypoglycemia (became unconscious or required help to resolve) during the reporting year? Select Yes, No, or Unknown.

Transplantation

What is the transplantation status of the patient currently? If the patient had transplantation in previous years please select or keep “Had transplantation” option:
- Not pertinent
- Accepted, on waiting list
- Evaluated, final decision pending Patient has been evaluated for an organ transplant and a final decision as to whether or not they will be placed on the transplant waiting list has not yet been made.
- Evaluated, rejected Patient has been evaluated for an organ transplant and was declined for organ transplant/placement on the transplant waiting list.
- Had transplantation Patient has had an organ transplant. Select if the patient has ever received an organ transplant.

**Transplant:** Select what type of transplant(s) the patient received. Once checked, specify the number of transplants received this year and the date of the last transplant performed. If the transplant(s) occurred outside of the reporting year, select "0".

- Lung: Bilateral
- Heart/Lung
- Lung: Lobar/Cadaveric
- Lung: Lobar/living donor
- Liver
- Kidney
- Other: Select only if the type(s) of organ transplanted are not listed. If selected, specify the transplant type(s).  
  - Specify transplant type: You must specify any unlisted organs transplanted if Other is selected in order to save the Annual Review Form as complete.

**Were there post-transplant complications?** Check if the patient had any post-transplant complications. If checked, you must indicate the specific complication(s) below in order to save the form as complete.

Select those that apply: Select all that apply.

- Bronchiolitis obliterans syndrome
- Lympho-proliferative disorder
- Other: Select only if the post-transplant complication(s) experienced are not listed. If selected, specify any unlisted transplant complications.
  - Specify other complication: You must specify any unlisted transplant complications if Other is selected in order to save the Encounter Form as complete.

**Clinical Trials**

Has this patient participated in any interventional (drug) studies? Select Yes, No, or Unknown.
Has this patient participated in any observational studies? Select Yes, No, or Unknown. This does not include participation in the Cystic Fibrosis Foundation Patient Registry.

**Health Insurance Coverage**

It is important for us to have accurate numbers of patients who have specific types of coverage: If the patient had health insurance for a majority (more than six months) of the reporting year, select all that apply.

- Health Insurance Policy (e.g. Private Insurance)
- Medicare
- Medicaid
- State special needs program, e.g., BCMH, CCS, CRS, GHPP, etc.
• **TriCare or other military health plan**
• **Indian Health Service**
• **Other** Select only if the patient’s insurance type is not listed. If selected, specify any unlisted insurance coverage.
  o **Specify if other insurance**: Specify insurance coverage if Other is selected.

**Patient has no health insurance** Check only if the patient did not have any health insurance for the majority (more than six months) of the reporting year.

**Was patient covered under parents’ health insurance plan?** Select Yes, No, or Unknown. Select Yes if the patient was covered under their parents’ health insurance plan at any point during the reporting year.

**Did patient receive free medicine or co-pay/deductible assistance from a Patient Assistance Program?** Select Yes, No, or Unknown. Select Yes if the patient received any free medications or co-pay/deductible assistance from a patient assistant program including the CFF’s Patient Assistance Fund at any point during the reporting year.

**Socio-Economic Status**

**Educational levels**: If the patient is adopted, report the adoptive parent’s education information. If the patient’s parents are of the same sex, record both parents' information in the mother/father education sections, regardless of sex, with the highest education level of the two parents entered under “Mother of patient”.

**Patient** Select the highest level of education completed by the patient.

- Less than High School
- High School diploma or equivalent
- Some college
- College Graduate
- Masters/Doctoral level degree
- Unknown/Not applicable The education level of the patient is not known or is not applicable (i.e., the patient is under the age 18 or is still in high school at the end of the reporting year).

**Father of patient** Select the highest level of education completed by the father of the patient.

- Less than High School
- High School diploma or equivalent
- Some college
- College Graduate
- Masters/Doctoral level degree
- Unknown/Not applicable The education level of the father of the patient is not known or is not applicable (i.e., the patient’s father is under the age 18 or is still in high school at the end of the reporting year).

**Mother of patient** Select the highest level of education completed by the mother of the patient. If patient’s parents are of the same sex, enter the education information for the parent with the **highest education level**, if different, here.

- Less than High School
- High School diploma or equivalent
- Some college
- College Graduate
- Masters/Doctoral level degree
• **Unknown/Not applicable** The education level of the mother of the patient is not known or is not applicable (i.e., the patient’s mother is under the age 18 or is still in high school at the end of the reporting year).

**Spouse of patient** Select the highest level of education completed by the spouse of the patient. If patient does not have a spouse, select **Unknown/Not applicable**.

- Less than High School
- High School diploma or equivalent
- Some college
- College Graduate
- Masters/Doctoral level degree
- **Unknown/Not applicable** The education level of the spouse of the patient is not known or is not applicable (i.e., the patient’s spouse is under the age 18 or is still in high school at the end of the reporting year, or the patient’s does not have a spouse).

**What was the total combined income of the household before taxes where the patient resided for the majority of the reporting year?** Select the total combined income of the household in which the patient resided for the majority of the reporting year before taxes. If the patient lives in two separate households equally, report the total combined income of the highest-earning household.

- <$10,000
- $10,000 to $19,999
- $20,000 to $29,999
- $30,000 to $39,999
- $40,000 to $49,999
- $50,000 to $59,999
- $60,000 to $69,999
- $70,000 to $79,999
- $80,000 to $89,999
- >$90,000
- **Unknown or Prefer not to Answer** The annual combined income of the patient’s household is not known or the patient/guardian(s) decline to disclose annual combined income.

**How many people currently live in the patient’s household (including the patient)?** Select the number of people that live in the patient’s household. This includes anyone that currently resides in the patient’s household a majority of the time. If the patient lives in two separate households equally, report the total number of people living in the household for which income was report.

- 1
- 2
- 3
- 4
- 5
- 6
- 7
- 8
- 9
- 10
- 11
- **12 or more**
- **Unknown** The number of people currently residing in the patient’s household is not known.
Age 18 and Older

This section will be enabled for patients age 18 and older.

Marital Status Select the patient’s marital status at the end of the reporting year.

- Single (never married)
- Living Together
- Married
- Separated
- Divorced
- Widowed
- Unknown The marital status of the patient is not known.

Employment (Select all that apply) Select the employment status of the patient for the majority of the reporting year.

- Part Time The patient worked between 1 and 30 hours per week for a business.
- Full time homemaker
- Full time employment The patient worked 30 hours or more per week for a business.
- Unemployed The patient is jobless, looking for jobs, and available for work.
- Student
- Disabled
- Retired

Unknown Check if the employment status of the patient is not known.

Pregnancy

This section will be enabled for female patients who are or will be age 14 or older by the end of the reporting year.

Was patient pregnant during the reporting year? Select Yes, No, or Unknown.

If “Yes”, indicate outcome:

- Live Birth
- Still Birth Baby is born with no signs of life at 28 weeks gestation or later.
- Spontaneous Abortion Also known as spontaneous miscarriage or miscarriage; spontaneous loss of pregnancy before 28 complete weeks of gestation.
- Therapeutic Abortion Any intentional termination of a pregnancy.
- Undelivered The patient is pregnant and has not yet delivered/will not deliver before the end of the reporting year.
- Unknown The outcome of the pregnancy is not known.

Age 2 and Younger

This section will be enabled for patients who were under the age of 2 during the reporting year.

Be sure to answer all questions.
Did the patient attend day care during this reporting year? Select Yes, No, or Unknown.
Did the family receive genetic counseling this reporting year? Select Yes, No, or Unknown.
Was the patient given palivizumab (Synagis) this season (Sept through January)? Select Yes, No, or Unknown. Select Yes even if injections were missed or discontinued.

Other

Please use this field to record any additional information about this patient: Record any additional information relevant to the reporting year here.