CF Foundation Patient Registry Variable List

The CF Foundation Patient Registry variable list is organized into the following sections:

- Demographics
- Diagnosis
- Clinic Visits
- Clinical Measurements
- Microbiology
- Treatments
- Complications
- Care Episodes (Hospitalizations/ Home IV)

Notes:

- Any variable that includes a date or other identifying information is considered protected health information (PHI, highlighted in yellow) and will require an Information Use Agreement in order to be included for approved projects.
- CFF provides encounter-level OR annualized data.
- Variables added after 2018 are in purple.

Please submit all data requests through the online portal: Patient Registry Data Requests | CF Foundation

Questions? Please contact datarequests@cff.org.
I. Variable type: DEMOGRAPHICS

- Birth characteristics
  - Date of birth (mm/dd/yyyy) [PHI]
  - Year of birth
  - Patient's sex
  - State of birth
  - Height of baby in cm
  - Weight of baby in kg
  - Baby delivered (full term, premature, unknown)
  - Specify gestational age if under 37 weeks
  - Patient attended day care during this reporting year
- Race / Ethnicity
  - Race includes 'White'
  - Race includes 'Black or African American'
  - Race includes 'American Indian or Alaska Native'
  - Race includes 'Asian'
  - Race includes 'Native Hawaiian or Other Pacific Islander'
  - Race includes 'Some other race'
  - Patient is Hispanic
- Mortality and Transplant
  - Date of Death (if died) [PHI]
  - Year of death (if died)
  - Reason for death (if died)
  - Age at death [PHI]
  - Date of first lung transplant [PHI]
- Health insurance
  - Health insurance policy (i.e. private insurance)
  - Medicare
  - Medicaid
  - State special needs program (BCMH, CCS, CRS, GHPP...)
  - TriCare or other military health plan
  - Indian Health Service
  - Other health insurance
  - Patient has no health insurance for the entire year
  - Patient covered on their parents' insurance plan
  - Receives free medicine/assistance from patient assistance program
- Residence
  - Patient resides in the US permanently
  - Patient state of residence [PHI]
  - Program state used for resident state [PHI]
  - Current zip code [PHI]
- Socioeconomic characteristics
  - Patient's education
  - Spouse's education
  - Total combined household income before taxes
  - Number of people currently living in household
  - Marital Status
  - Employed part time
  - Full time homemaker
• Employment
  o Employed full time
  o Unemployed Student
  o Disabled
  o Retired
  o Employment unknown

• Smoking
  o Patient smokes cigarettes during the reporting year
  o How often was this patient exposed to secondhand smoke
  o Patient lives in household where someone smokes

• Parent characteristics
  o Patient's mother's height in cm
  o Patient's father's height in cm
  o Father's education
  o Mother's education

• Research involvement
  o Patient participated in interventional (drug) studies
  o Patient participated in observational studies

• Patient pregnancy
  o Patient was pregnant during the reporting year
  o Outcome of patient's pregnancy

Return to first page
II. **Variable type: DIAGNOSIS**

- **Diagnosis date**
  - Diagnosis date [PHI]
  - Patient diagnosis
  - Age at diagnosis
  - Year of CF diagnosis
  - First year in the CFF Patient Registry

- **Sweat test**
  - Date of last sweat test [PHI]
  - Year of last sweat test
  - First reported sweat test date [PHI]
  - Latest sweat value reported
  - Highest sweat value reported

- **Genotyping**
  - Has this patient been genotyped
  - Date of genotyping [PHI]
  - Year of last genotyped
  - Mutation classification
  - Name of the first mutation
  - Mutation 1 Class
  - Poly T tract associated with mutation 1
  - Poly TG repeats mutation 1
  - Name of the second mutation
  - Mutation 2 Class
  - Poly T tract associated with mutation 2
  - Poly TG repeats mutation 2
  - Does patient have additional mutations?
  - Delta F508

- **Diagnosis characteristics**
  - Meconium ileus with or without perforation
  - Patient diagnosed after false-negative newborn screen
  - CF dx sugg by Acute or persistent respiratory abnormalities
  - CF dx sugg by CBAVD or related abnormalities
  - CF dx sugg by Digital clubbing
  - CF dx sugg by DNA Analysis
  - CF dx sugg by Edema
  - CF dx sugg by Electrolyte imbalance
  - CF dx sugg by Failure to thrive/malnutrition
  - CF dx sugg by Family history
  - CF dx sugg by Infertility/GU abnormalities
  - CF dx sugg by Liver problems
  - CF dx sugg by Meconium ileus/intestinal obstruction
  - CF dx sugg by Nasal polyps/sinus disease
  - CF dx sugg by Newborn (neonatal) screening
  - CF dx sugg by Prenatal screening (CVS, amnio)
  - CF dx sugg by Rectal prolapse
  - CF dx sugg by Steatorrhea/abnormal stool/malabsorption
  - CF dx sugg by Other
  - CF dx sugg by unknown
  - Sweat<60, dx sugg DNA Analysis/genotyping
- Sweat<60, dx sugg Transepithelial potential differences
- Sweat<60, dx sugg Clinical presentation
- Sweat<60, dx sugg unknown

Return to first page
### III. Variable type: CLINIC VISITS

- **Visit details**
  - Site ID
  - Date of Encounter [PHI]
  - Location where patient was seen
  - Encounter age [PHI]
  - Patient was seen by a Social Worker
  - Patient was seen by a Dietitian/Nutritionist
  - Patient was seen by a Pharmacist
  - Patient seen by a Respiratory Therapist
  - Patient seen by a Physical Therapist
  - Patient seen by a Mental Health Coordinator
  - Was there assessment of oral intake

- **Clinic visits - summarized by calendar year**
  - # of clinic visits in the review year
  - Primary Program ID
  - Non-primary Program reported in the year
  - Patient was seen by a Social Worker
  - Patient was seen by a Dietitian/Nutritionist
  - Patient was seen by a Pharmacist
  - Patient seen by a Respiratory Therapist
  - Patient seen by a Physical Therapist
  - Did the family receive genetic counseling this year
  - Was there assessment of oral intake
  - Patient seen by a Mental Health Coordinator
  - Eye Exam on Annual Form
  - Age on 12/31 of review year [PHI]
  - Any encounter data
  - Any annual data

- **Pulmonary exacerbation - encounter level**
  - Assessment of potential pulmonary exacerbation
  - Incr. airway clearance, exercise, and/or bronchodilators
  - Oral NON-quinolone antibiotic
  - Oral quinolone antibiotic (e.g. Cipro, levofloxacin)
  - Inhaled antibiotic
  - Inhaled antibiotic PLUS Oral NON-quinolone antibiotic
  - Inhaled antibiotic PLUS Oral quinolone antibiotic
  - Crackles on physical exam
  - None of the above

- **Pulmonary exacerbation - summarized by calendar year**
  - Any pulmonary exacerbation assessment in the year
  - Any pulmonary exacerbation assessment positive in the year
  - # encounters with pulmonary exacerbation assessments: absent
  - # encounters with pulmonary exacerbation assessments: mild exacerbations
  - # encounters with pulmonary exacerbation assessments: moderate exacerbations
  - # encounters with pulmonary exacerbation assessments: severe exacerbations
  - Encounter exacerbation treatment: Incr. airway clearance
  - Encounter exacerbation treatment: Oral NON-quinolone antibiotic
  - Encounter exacerbation treatment: Oral quinolone antibiotic
  - Encounter exacerbation treatment: Inhaled Antibiotic
  - Encounter exacerbation treatment: Inhaled + Oral NON-quinolone antibiotic
Encounter exacerbation treatment: Inhaled + oral quinolone antibiotic
Encounter exacerbation treatment: None of the above
# of encounters with crackles (rales) on physical exam

- Infant feeding - encounter level
  - Type of feeding
  - Infant formula type
  - Caloric density (20, 22, 24, 27 or 30 cal/oz)

- Infant feeding - summarized by calendar year
  - Type of infant feeding: breast milk
  - Type of infant feeding: breast milk plus formula
  - Type of infant feeding: formula exclusively
  - Type of infant feeding: other food
  - Type of infant feeding: unknown
  - Infant formula type: Cow's milk
  - Infant formula type: Soy milk
  - Infant formula type: Predigested
  - Infant formula type: Other
  - Infant formula caloric density: 20 cal/oz
  - Infant formula caloric density: 22 cal/oz
  - Infant formula caloric density: 24 cal/oz
  - Infant formula caloric density: 27 cal/oz
  - Infant formula caloric density: 30 cal/oz
  - Infant formula caloric density: other

Return to first page
IV. Variable type: CLINICAL MEASUREMENTS

- Anthropometric measures - encounter level
  - Patient's height
  - Original height as report (includes suspect values)
  - Height Percentile
  - Height Percentile Z score (age < 20)
  - Height Percentile based on WHO (age < 2 only)
  - Height-for-age Z score based on WHO (age < 2)
  - BMI Value
  - BMI Percentile
  - Weight-for-Length Percentile
  - Weight for length percentile based on WHO (age < 2 only)
  - Weight-for-height Z score based on WHO (age < 2)
  - Weight of Patient
  - Original weight as report (includes suspect values)
  - Weight Percentile
  - Weight Percentile Z score (age < 20)
  - Weight percentile based on WHO (age < 2 only)
  - Weight-for-age Z score based on WHO (age < 2)

- Anthropometric measures - summarized by calendar year
  - # height measurements in the review year
  - Patient's height
  - Height Percentile in the review year
  - Height Percentile Z score (age < 20)
  - Height percentile based on WHO (age < 2 only)
  - Height for Age z-score
  - BMI Value
  - BMI Percentile
  - Weight-for-Length Percentile
  - Weight for length percentile based on WHO (age < 2 only)
  - Weight-for-height Z score based on WHO (age < 2)
  - # weight measurements in the review year
  - Weight of Patient
  - Weight Percentile
  - Weight Percentile Z score (age < 20)
  - Weight percentile based on WHO (age < 2 only)
  - Weight for Age z-score

- Pulmonary function tests - encounter level
  - Was impossible to take PFTs
  - Measure of FVC
  - Predicted Value for FVC using GLI reference equations
  - Measure of FEV1
  - CF specific FEV1 percentile for kids
  - Predicted Value for FEV1 using GLI reference equations
  - Measure of FEF25-75
  - Predicted Value for FEF25-75 using GLI reference equations
  - Measure of FEV1/FVC
  - Predicted Value for FEV1/FEV1 using GLI reference equations
• Pulmonary function tests - summarized by calendar year
  o Any pulmonary data completed
  o # unable to perform pulmonary function test
  o # FVC measurements in the review year
  o Predicted Value for FVC using GLI reference equations
  o Predicted Value for FEF25-75 using GLI reference equations
  o Measure of FEV1/FVC
  o Predicted Value for FEV1/FVC using GLI reference equations
  o # FEV1 measurements in the review year
  o Predicted Value for FEV1 using GLI reference equations
  o # FEV25-75 measurements in the review year
  o Measure of FEV25-75
  o Post-Transplant Predicted Value for FEF25-75 using GLI reference equations
  o Post-Transplant Predicted Value for FEV1 using GLI reference equations
  o Post-Transplant Predicted Value for FEV1/FVC using GLI reference equations
  o Post-Transplant Predicted Value for FVC using GLI reference equations

• Lab values - encounter level
  o WBC count x1,000/µL
  o Platelet Count x1,000/µL
  o Hemoglobin (grams per deciliter)
  o Serum Creatinine level (mg/dL)
  o Alanine Aminotransferase (ALT or SGPT), IU/L
  o Alkaline phosphatase 2 hour blood glucose (mg/dL)
  o Hgb A1C value, %
  o Fecal elastase value (microg/g of stool)
  o Liver enzymes drawn (pre-2010) Aspartate aminotransferase
  o Liver Function Tests Albumin
  o Total bilirubin, mg/dL
  o GGTP (gamma glutamyl transpeptidase), IU/L
  o Random blood glucose (mg/dL)
  o Fasting glucose level (mg/dL)
  o OGTT Fasting glucose level (mg/dL)
  o 1 hour blood glucose (mg/dL)

• Lab values - summarized by calendar year
  o Any lab data completed
  o WBC count x1,000/µL
  o Platelet Count x1,000/µL
  o Hemoglobin (grams per deciliter)
  o Serum creatinine level (mg/dL)
  o Alanine Aminotransferase (ALT or SGPT), IU/L
  o Aspartate aminotransferase
  o Total bilirubin, mg/dL
  o GGTP (gamma glutamyl transpeptidase), IU/L
  o Liver enzymes drawn (pre-2010)
  o Random blood glucose (mg/dL)
  o 1 hour blood glucose (mg/dL)
  o Fasting glucose level (mg/dL)
  o OGTT fasting glucose level (mg/dL)
  o 2 hour blood glucose (mg/dL)
- HgbA1C value, %
- Fecal elastase value (microg/g of stool)
- Alkaline phosphatase
- Albumin

- Screening tests
  - Was chest x-ray performed in the reporting year
  - Was IgE screening performed in the reporting year
  - Liver function tests performed
  - Fat soluble vitamin levels measured?
  - Most recent fecal elastase value
  - Patient screened for anxiety disorder
  - Patient screened for symptoms of classic depression
  - DEXA scan for bone density performed in the reporting year
  - DEXA scan result is normal
  - DEXA scan result is osteopenia
  - DEXA scan result is osteoporosis
  - DEXA scan result is other
  - DEXA scan result is unknown
  - Colonoscopy performed
  - Colonoscopy_results

Return to first page
V. Variable type: MICROBIOLOGY

- **Bacteria - encounter level**
  - Was bacterial culture done?
  - **Date of bacterial culture [PHI]**
  - Type of specimen (for bacterial culture)
  - Culture results
  - Staphylococcus aureus
  - MRSA
  - Any Burkholderia Complex species
  - Resistant to all aminoglycosides tested
  - Resistant to all quinolones tested
  - Resistant to all beta lactams tested
  - Multidrug-Resistant P. aeruginosa (MDR-PA) Infection
  - Alcaligenes
  - Stenotrophomonas
  - MSSA
  - Haemophilus influenzae (any species)
  - Pseudomonas aeruginosa found in the culture
  - Mucoid Pseudomonas aeruginosa
  - Non-mucoid Pseudomonas aeruginosa
  - Mucoid Pseudomonas aeruginosa status unknown

- **Bacteria - summarized by calendar year**
  - Any respiratory microbiology data completed
  - Was bacterial culture done?
  - Number of cultures in the year
  - Number of Micro cultures in the year
  - Number of Myco cultures in the year
  - # respiratory culture type: sputum
  - # respiratory culture type: induced sputum
  - # respiratory culture type: throat/nasal
  - Any Burkholderia Complex species
  - Resistant to all aminoglycosides tested
  - Resistant to all quinolones tested
  - Resistant to all beta lactams tested
  - Resistant to at least 2 (beta lactams, aminoglycosides, quinolones)
  - Alcaligenes
  - Stenotrophomonas
  - MSSA
  - Haemophilus influenzae (any species)
  - Pseudomonas aeruginosa found in the culture
  - Mucoid Pseudomonas aeruginosa
  - Non-mucoid Pseudomonas aeruginosa
  - Mucoid Pseudomonas aeruginosa status unknown
  - # respiratory culture type: bronchoscopy
  - Any culture result: no growth/sterile
  - Any culture result: normal flora
  - Any culture result: microorganisms
  - Staphylococcus aureus
  - MRSA
• Bacteria - other frequency
  o First year Burkholderia species was reported
  o First year b. cepacia was confirmed at ref. lab

• Mycobacteria - encounter level
  o Was mycobacterial culture done?
    o Date of myco culture [PHI]
  o Type of specimen (for mycobacterial culture)
    o Acid-Fast Bacilli Smear
  o Myco culture results
  o Mycobacterial tuberculosis
  o Mycobacterium terrae
  o Mycobacterium other
  o Mycobacterium abscessus/chelonaes
  o Mycobacterium avium complex
  o Mycobacterium fortuitum group
  o Mycobacterium gordonae
  o Mycobacterium kansasii
  o Mycobacterium marinum

• Mycobacteria - summarized by calendar year
  o Was mycobacterial culture done?
  o # mycobacterial culture type: sputum
  o # mycobacterial culture type: induced sputum
  o # mycobacterial culture type: bronchoscopy
  o Any Acid-Fast Bacilli smear: Positive
  o Any Acid-Fast Bacilli smear: Negative
  o Mycobacterium terrae
  o Mycobacterium other
  o Mycobacterium abscessus/chelonaes
  o Mycobacterium avium complex
  o Mycobacterium fortuitum group
  o Mycobacterium gordonae
  o Mycobacterium kansasii
  o Mycobacterium marinum
  o Any Acid-Fast Bacilli smear: Not done
  o Any mycobacterial result: no growth/sterile
  o Any mycobacterial result: normal flora
  o Any mycobacterial result: microorganisms
  o Nontuberculous mycobacteria - pre 2010
  o Mycobacterial tuberculosis

• Fungus - encounter level
  o Aspergillus Candida Scedosporium species
  o Other bacterial or fungal microorganism species, specify

• Fungus - summarized by calendar year
  o Aspergillus Candida Scedosporium species
  o Other bacterial or fungal microorganism species, specify

Return to first page
VI. Variable type: TREATMENTS

- **Antibiotics - encounter level**
  - Tobramycin solution for inhalation (i.e. TOBI)
  - TOBI frequency
  - TOBI Podhaler
  - TOBI Podhaler Frequency
  - Other inhaled aminoglycoside (e.g. gentamicin, amikacin)
  - Other aminoglycoside - frequency
  - Colistin
  - Colistin Frequency
  - Aztreonam - Inhaled
  - Aztreonam frequency
  - Chronic oral macrolide antibiotic
  - Azithromycin
  - Clarithromycin
  - Sulfa
  - Amoxicillin
  - Tetracycline
  - Other oral antibiotic
  - Other chronic oral antibiotic
  - Quinolone
  - Cephalosporin
  - Bethkis
  - Bethkis frequency

- **Antibiotics - summarized by calendar year**
  - Tobramycin solution for inhalation (i.e. TOBI)
  - TOBI 300 mg BID alternate month schedule
  - TOBI 300 mg BID continuous
  - TOBI Other regimen (different dose or freq)
  - TOBI for Eradication
  - TOBI as needed
  - TOBI Podhaler
  - TOBI Podhaler Four 28mg capsules BID alternate month schedule
  - TOBI Podhaler Other regimen (different dose or frequency)
  - TOBI Podhaler for Eradication
  - TOBI Podhaler as needed
  - Other inhaled aminoglycoside (e.g. gentamicin, amikacin)
  - Aminoglycoside alternate month schedule
  - Aminoglycoside Continuous
  - Aminoglycoside Other regimen (different dose or frequency)
  - Aminoglycoside for Eradication
  - Aminoglycoside as needed
  - Colistin
  - Colistin alternate month schedule
  - Colistin Continuous
  - Colistin Other regimen (different dose or frequency)
  - Colistin for Eradication
  - Colistin as needed
  - Aztreonam 75 mg TID Alternate Month Schedule
  - Aztreonam 75 mg TID Continuous
- Aztreonam - other regimen
- Aztreonam for eradication
- Aztreonam as needed
- Aztreonam – Inhaled
- Chronic oral macrolide antibiotic
- Azithromycin
- Tetracycline
- Other oral antibiotic
- Clarithromycin
- Other chronic oral antibiotic
- Quinolone
- Cephalosporin
- Sulfa
- Amoxicillin
- Bethkis for Eradication
- Bethkis as needed
- Bethkis
- Bethkis 300 mg BID alternate month schedule
- Bethkis: other regimen (different dose of freq)

- Pulmonary meds - encounter level
  - Not on any chronic pulmonary medications
  - Dornase alfa (i.e. Pulmozyme)
  - Dornase frequency
  - Acetylcysteine or Mucomist
  - High-dose ibuprofen (e.g. 25-30 mg/kg)
  - Hypertonic saline
  - Leukotriene modifiers
  - Mast cell stabilizers
  - Antifungals
  - Oral (e.g. prednisone)
  - Inhaled (e.g. fluticasone, Flovent, budesonide)
  - Inhaled in combination wth bronchodilator (e.g. Advair)
  - Other Corticosteroids type
  - Other Corticosteroids Total (mg/dose)
  - Other Corticosteroids frequency
  - Hypertonic saline concentration (%)
  - Hypertonic Saline Frequency
  - Short acting anticholinergic
  - Long acting anticholinergic
  - Combination beta agonist and anticholinergic
  - Beta agonist
  - Theophylline product
  - Short acting beta agonist
  - Long acting beta agonist
  - Allergic/intolerant to Macrolide antibiotics
  - Allergic/intolerant to High-dose ibuprofen
  - Allergic/intolerant to Hypertonic saline
  - Allergic/intolerant to Aztreonam
  - Drug intolerance/allergies for Ivacaftor
  - Drug intolerance/allergies for Ivacaftor Lumacaftor
  - Drug intolerance/ allergies for Tezacaftor/Ivacaftor (i.e. Symdeko)
  - Drug intolerance/allergies for Elexa/Tez/Iva
- Allergic/intolerant to Dornase alfa
- Allergic/intolerant to Tobramycin solution for inhalation
- Allergic/intolerant to Colistin

- Pulmonary meds - summarized by calendar year
  - Not on any chronic pulmonary medications
    - Dornase alfa (i.e. Pulmozyme)
    - Dornase alfa 2.5 mg qd
    - Dornase alfa 2.5 mg BID
    - Dornase alfa: Other regimen (different dose or frequency)
  - Dornase alfa as needed
  - Acetylcysteine or Mucomist
  - High-dose ibuprofen (e.g. 25-30 mg/kg)
  - Oral (e.g. prednisone)
  - Inhaled (e.g. fluticasone, Flovent, budesonide)
  - Inhaled combination with bronchodilator (e.g. Advair)
  - Oral Corticosteroids Types – Prednisone
  - Oral Corticosteroids Types - Methylprednisolone (Medrol)
  - Oral Corticosteroids Types – Dexamethasone
  - Oral Corticosteroids Types – Other
  - Average Oral Corticosteroids Total (mg/dose)
  - Oral Corticosteroids Frequency - Short Term (Less Than Two Weeks)
  - Oral Corticosteroids Frequency - Chronic, Low Dose
  - Oral Corticosteroids Frequency - Chronic, High Dose
  - Oral Corticosteroids Frequency - Other
    - Leukotriene modifiers
    - Mast cell stabilizers
    - Antifungals
    - Hypertonic saline
      - Hypertonic saline concentration: 1
      - Hypertonic saline concentration: 2
      - Hypertonic saline concentration: 3
      - Hypertonic saline concentration: 4
      - Hypertonic saline concentration: 5
      - Hypertonic saline concentration: 6
      - Hypertonic saline concentration: 7
      - Hypertonic saline concentration: 8
      - Hypertonic saline frequency: QD
      - Hypertonic saline frequency: BID
      - Hypertonic saline frequency: Other
      - Hypertonic saline frequency as needed
    - Beta agonist
      - Theophylline product
      - Short acting beta agonist
      - Long acting beta agonist
      - Short acting anticholinergic
      - Long acting anticholinergic
      - Combination beta agonist and anticholinergic
      - Drug intolerance/allergies for Dornase alfa
      - Drug intolerance/allergies for tobramycin
      - Drug intolerance/allergies for Colistin
- Drug intolerance/allergies for Macrolide antibiotics
- Drug intolerance/allergies for High-dose ibuprofen
- Drug intolerance/allergies for Hypertonic saline
- Drug intolerance/allergies for Aztreonam
- Drug intolerance/allergies for Ivacaftor
- Drug intolerance/allergies for Ivacaftor Lumacaftor
- Drug intolerance/allergies for Elexa/Tez/Iva

- CFTR modulators - encounter level
  - On Ivacaftor medication (VX-770)
  - VX770 (Ivacaftor) dosage
  - Ivacaftor/Lumacaftor (i.e., Orkambi)
  - Ivacaftor/Lumacaftor (i.e., Orkambi) dosage
  - Triple Combo Combination Therapy, Elexacaftor / Tezacafor / Ivacaftor (i.e., Trikafta)
  - Triple Combo Dose
  - First date prescribed a modulator
  - First date prescribed Ivacaftor (i.e., Kalydeco)
  - First date prescribed Lumacaftor / Ivacaftor (i.e., Orkambi)
  - First date prescribed Tezacaftor / Ivacaftor (i.e., Symdeko)
  - First date prescribed Elexacaftor / Tezacafor / Ivacaftor (i.e., Trikafta)
  - First date taking Elexacaftor / Tezacafor / Ivacaftor (i.e., Trikafta)

- CFTR modulators - summarized by calendar year
  - On Ivacaftor medication (VX-770)
  - Ivacaftor - 150 mg BID
  - Ivacaftor - Other regimen
  - Ivacaftor/Lumacaftor (i.e., Orkambi)
  - Ivacaftor/Lumacaftor - Full dose BID
  - Ivacaftor/Lumacaftor - Half dose BID
  - Ivacaftor/Lumacaftor - Other regimen
  - Triple Combo Combination Therapy (i.e. Trikafta)
  - Triple Combo - Full Dose BID
  - Triple Combo Dose - Half Dose BID
  - Triple Combo Dose - Other

- Other treatments - encounter level
  - Patient is taking enzymes
  - Lipase Units per Largest Meal
  - Lipase Units per Day
  - H2 Blocker
  - Proton Pump Inhibitor
  - Acid blocker unknown
  - GI Other: Ursodeoxycholic acid
  - Salt supplementation
  - CF-specific vitamins (i.e. vitamins A, D, E and K)
  - Is patient currently receiving supplemental feeding
  - Supplemental feeding: total parenteral nutrition
  - Supplemental feeding: oral supplementation
- Supplemental feeding: nasogastric tube
- Supplemental feeding: gastrostomy tube/button
- Supplemental feeding: jejunal tube
- Enzyme use with supplemental feeding
- Primary Airway Clearance technique
- Forced expiratory techniques
- Oscillating PEP
- High frequency chest wall oscillation
- Exercise
- Positive Expiratory Pressure
- Postural drainage with clapping

• Other treatments – annual
  - Did patient use oxygen therapy during this year
  - Use non-invasive ventilation
  - Did the patient receive an influenza vaccination
  - Was the patient started on treatment for NTM
  - Patient given palivizumab (Synagis) this season
  - Patient on growth hormone in the reporting year

• Other treatments - summarized by calendar year
  - Not on any chronic pulmonary medications
  - Any medicine data completed
  - Patient is on enzymes
  - Patient received supplemental feeding during year
  - Primary airway clearance technique - most frequently used
  - Salt supplementation (encounter age < 2)
  - CF-specific vitamins (i.e. vitamins A, D, E and K)
  - Any airway clearance technique data completed
  - Heart/lung transplant in review year
  - Kidney Transplant in review year
  - Liver Transplant in review year
  - Bilateral lung transplant in review year
  - Lung Transplant: Lobar/living donor transplant in review year
  - Lung Transplant: Lobar/cadaveric transplant in review year
  - Other Transplant in review year
  - Supplemental feeding: oral supplementation
  - Supplemental feeding: nasogastric tube
  - Supplemental feeding: gastrostomy tube/button
  - Supplemental feeding: jejunal tube
  - Supplemental feeding: total parenteral nutrition
  - No enzymes recommended with tube feeding
  - Yes, enzymes mixed into the formula
  - Yes, enzymes administered directly through the tube (i.e., not into the formula)
  - Yes, enzymes taken by mouth prior to, during and/or after the feeding
  - Yes, formula infused through Relizorb (enzyme cartridge)
  - Enzymes, other
  - Exercise
  - No primary airway clearance technique
  - Other primary airway clearance technique: Positive Expiratory Pressure (PEP)
Postural drainage with clapping (CPT)
- Forced expiratory techniques (e.g. autogenic drainage)
- Oscillating PEP (e.g. Flutter, acapella, IPV)
- High frequency chest wall oscillation (e.g. Vest)
- Oscillating PEP (e.g. Flutter, acapella, IPV)
- High frequency chest wall oscillation (e.g. Vest)
- Airway clearance technique: Exercise Positive Expiratory Pressure (PEP)
- Postural drainage with clapping (CPT)
- Forced expiratory techniques (e.g. autogenic drainage)

Transplant

- Current transplant status (includes prior years)
- First year had a lung transplant Ever
- Bilateral lung transplant Ever
- Heart/lung transplant Ever
- Lung Transplant: Lobar/cadaveric transplant
- Ever Lung Transplant: Lobar/living donor transplant
- Ever Any lung transplant
- Ever Liver transplant
- Ever Kidney transplant
- Ever Other transplant
- Were there post transplant complications
  - Bronchiolitis obliterans syndrome
  - Lympho-proliferative disorder
  - Other transplant complications

Return to first page
VII. Variable type: COMPLICATIONS

- CF related diabetes - encounter level
  - CFRD Status
  - Retinopathy
  - Microalbuminuria
  - Chronic renal insufficiency
  - Chronic renal failure requiring dialysis
  - Peripheral neuropathy

- CF related diabetes - annual
  - CFRD status on annual form
  - Retinal eye exam performed by an ophthalmologist
  - Spot urine sent for albumin/creatinine ratio
  - Was the patient prescribed treatment for CFRD
  - Spot urine sent for albumin/creatinine ratio
  - Was the patient prescribed treatment for CFRD
  - Dietary change
  - Oral hypoglycemic agents
  - Intermittent insulin (with illness, steroids, etc.)
  - Chronic insulin
  - Any episodes of severe hypoglycemia
  - Retinopathy
  - Microalbuminuria
  - Chronic renal insufficiency
  - Chronic renal failure requiring dialysis
  - Peripheral neuropathy

- Liver disease - encounter level
  - Gall stones
  - Gall stones, requiring surgery/procedure
  - Liver disease, cirrhosis
  - Esophageal varices
  - Gastric varices
  - GI bleed related to varices
  - Splenomegaly
  - Hypersplenism
  - Aspects
  - Encephalopathy
  - Acute hepatitis
  - Acute hepatitis - infectious
  - Acute hepatitis - non-infectious
  - Acute hepatitis - unknown
  - Liver disease, non-cirrhosis
  - Hepatic Steatosis
  - Liver disease, other Acute liver failure
• Liver disease - summarized by calendar year
  o Gallstones
  o Gallstones requiring surgery/procedure
  o Liver disease, cirrhosis
  o Esophageal varices
  o Gastric varices
  o GI bleed related to varices
  o Splenomegaly
  o Hypersplenism
  o Ascites
  o Encephalopathy
  o Acute hepatitis
  o Acute hepatitis - infectious
  o Acute hepatitis - non-infectious
  o Acute hepatitis - unknown
  o Liver disease, non-cirrhosis
  o Hepatic steatosis
  o Liver disease, other
  o Acute liver failure
  o Liver enzymes elevated (for years 2003-2009)

• Other complications - encounter level
  o Patient does not have any complications
  o Arthritis/Arthropathy
  o Bone fracture
  o Osteopenia
  o Osteoporosis
  o Allergic Bronchial Pulmonary Aspergillosis
  o Asthma
  o Hemoptysis, massive
  o Hemoptysis type
  o Pneumothorax requiring chest tube
  o Distal intestinal obstruction syndrome
  o Fibrosing colonopathy/colonic stricture
  o GERD
  o GI Bleed requiring hospitalization, non-variceal
  o Pancreatitis
  o Peptic ulcer disease
  o Rectal prolapse
  o History of intestinal or colon surgery
  o Anxiety disorder
  o Cancer confirmed by histology
  o Depression
  o Hearing loss
  o Hypertension
  o Kidney Stones
  o Nasal polyps requiring surgery
  o Renal failure requiring dialysis
  o Sinus Disease
  o Absence of vas deferens
Other complications - summarized by calendar year

- No complications
- Arthritis/Arthropathy
- Bone fracture
- Osteopenia
- Osteoporosis
- Allergic Bronchial Pulmonary Aspergillosis
- Asthma
- Any Hemoptysis
- Hemoptysis - massive
- Hemoptysis - other
- Pneumothorax requiring chest tube
- Distal intestinal obstruction syndrome
- Fibrosing colonopathy/colonic stricture
- GERD
- GI Bleed requiring hospitalization, non variceal
- Pancreatitis
- Peptic ulcer disease
- Rectal prolapse
- History of intestinal or colon surgery
- Anxiety disorder
- Cancer confirmed by histology
- Depression
- Hearing loss
- Hypertension
- Kidney stones
- Nasal polyps requiring surgery
- Renal failure requiring dialysis
- Sinus Disease
- Absence of vas deferens
VIII. Variable type: CARE EPISODES (HOSPITALIZATIONS/ HOME IV)

- Care episodes
  - Care center ID
  - Start date for care episode [PHI]
  - End date for care episode [PHI]
  - Location of care episode
  - Care Episode Site ID
  - Total Nights for care episode
  - # nights in hospital
  - # nights on home IV
  - # nights with reason pulmonary complication
  - # home IV nights with reason pulmonary exacerbation
  - # hospitalization nights with reason pulmonary exacerbation
  - # home IV nights with reason pulmonary complication
  - # hospitalization nights with reason pulmonary complication
  - # home IV nights with reason GI complications
  - # hospitalization nights with reason GI complications
  - # home IV nights with reason Transplant related
  - # hospitalization nights with reason Transplant related
  - # home IV nights with reason sinus infection
  - # hospitalization nights with reason sinus infection
  - # home IV nights with reason non-Transplant surgery
  - # hospitalization nights with reason non-Transplant surgery
  - # home IV nights with reason other
  - # hospitalization nights with reason other
  - # home IV nights with reason unknown
  - # hospitalization nights with reason unknown
  - # hospitalization nights with reason NTM
  - # home IV nights with reason NTM

- Care episodes - summarized by calendar year
  - Any hospitalization in the year
  - # hospitalizations (including partial) in year
  - # nights in hospital
  - Any home IV in the year
  - # nights on home IV
  - Any episode data
  - Total # care episodes in the year
  - Total # pulmonary exacerbations treated by IV antibiotics
  - # nights in hospital for pulmonary exacerbations
  - # nights on home IV for pulmonary exacerbations
  - Total # pulmonary exacerbation treated by IV antibiotics (incl unknown)
  - # nights hospital for pulmonary exacerbation (incl unknown)
  - # nights home IV for pulmonary exacerbation (incl unknown)
  - # nights with reason pulmonary complication
  - # nights with reason GI complications
- # nights with reason transplant related
- # nights with reason sinus infection
- # nights with reason non-transplant surgery
- # nights with reason other
- # nights with reason unknown
- # nights with reason NTM