



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 \(Hospitalizations/ Home IV\)](#)
- [Advanced Lung Disease](#)

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
 - Employed full time
 - Unemployed Student
 - Disabled
 - Retired
 - Employment unknown
- Smoking
 - Patient smokes cigarettes during the reporting year
 - How often was this patient exposed to secondhand smoke
 - Patient lives in household where someone smokes
- Parent characteristics
 - Patient's mother's height in cm
 - Patient's father's height in cm
 - Father's education
 - Mother's education
- Research involvement
 - Patient participated in interventional (drug) studies
 - Patient participated in observational studies
- Patient pregnancy
 - Patient was pregnant during the reporting year
 - Outcome of patient's pregnancy

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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
 - Eligible for ETI by genotype
 - Eligible for ivacaftor by genotype
 - Lumacaftor/ivacaftor eligible by genotype
 - Tezacaftor/ivacaftor eligible by genotype
- 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

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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 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 was seen by a Respiratory Therapist
 - Patient was 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

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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 2012 reference equations
 - Measure of FEV1
 - CF specific FEV1 percentile for kids
 - Predicted Value for FEV1 using GLI 2012 reference equations
 - Measure of FEF25-75
 - Predicted Value for FEF25-75 using GLI 2012 reference equations
 - Measure of FEV1FVC
 - Predicted Value for FEV1/FEV1 using GLI 2012 reference equations

- Race-neutral GLI FEV1/FVC % predicted
- Race-neutral GLI FEV1 % predicted
- Race-neutral GLI FVC % predicted
- Race-neutral GLI FEV1/FVC % predicted post-transplant
- Race-neutral GLI FEV1 % predicted post-transplant
- Race-neutral GLI FVC % predicted post-transplant

- Pulmonary function tests - summarized by calendar year
 - Any pulmonary data completed
 - # unable to perform pulmonary function test
 - # FVC measurements in the review year
 - Predicted Value for FVC using GLI 2012 reference equations
 - Predicted Value for FEF25-75 using GLI 2012 reference equations
 - Measure of FEV1/FVC
 - Predicted Value for FEV1/FVC using GLI 2012 reference equations
 - # FEV1 measurements in the review year
 - Predicted Value for FEV1 using GLI 2012 reference equations
 - # FEV25-75 measurements in the review year
 - Measure of FEF25-75
 - Post-Transplant Predicted Value for FEF25-75 using GLI 2012 reference equations
 - Post-Transplant Predicted Value for FEV1 using GLI 2012 reference equations
 - Post-Transplant Predicted Value for FEV1/FVC using GLI 2012 reference equations
 - Post-Transplant Predicted Value for FVC using GLI reference equations
 - Race-neutral GLI FEV1/FVC % predicted
 - Race-neutral GLI FEV1 % predicted
 - Race-neutral GLI FVC % predicted
 - Race-neutral GLI FEV1/FVC % predicted post-transplant
 - Race-neutral GLI FEV1 % predicted post-transplant
 - Race-neutral GLI FVC % predicted post-transplant

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

- Lab values - summarized by calendar year
 - Any lab data completed
 - WBC count x1,000/microL
 - Platelet Count x1,000/microL
 - Hemoglobin (grams per deciliter)
 - Serum creatinine level (mg/dL)
 - Alanine Aminotransferase (ALT or SGPT), IU/L
 - Aspartate aminotransferase
 - Total bilirubin, mg/dL
 - GGTP (gamma glutamyl transpeptidase), IU/L
 - Liver enzymes drawn (pre-2010)
 - Random blood glucose (mg/dL)
 - 1 hour blood glucose (mg/dL)
 - Fasting glucose level (mg/dL)
 - OGTT fasting glucose level (mg/dL)
 - 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

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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
 - First year Burkholderia species was reported
 - First year b. cepacia was confirmed at ref. lab
- Mycobacteria - encounter level
 - Was mycobacterial culture done?
 - **Date of myco culture [PHI]**
 - Type of specimen (for mycobacterial culture)
 - Acid-Fast Bacilli Smear
 - Myco culture results
 - Mycobacterial tuberculosis
 - Mycobacterium terrae
 - Mycobacterium other
 - Mycobacterium abscessus/chelonae
 - Mycobacterium avium complex
 - Mycobacterium fortuitum group
 - Mycobacterium gordonae
 - Mycobacterium kansasii
 - Mycobacterium marinum
- Mycobacteria - summarized by calendar year
 - Was mycobacterial culture done?
 - # mycobacterial culture type: sputum
 - # mycobacterial culture type: induced sputum
 - # mycobacterial culture type: bronchoscopy
 - Any Acid-Fast Bacilli smear: Positive
 - Any Acid-Fast Bacilli smear: Negative
 - Mycobacterium terrae
 - Mycobacterium other
 - Mycobacterium abscessus/chelonae
 - Mycobacterium avium complex
 - Mycobacterium fortuitum group
 - Mycobacterium gordonae
 - Mycobacterium kansasii
 - Mycobacterium marinum
 - Any Acid-Fast Bacilli smear: Not done
 - Any mycobacterial result: no growth/sterile
 - Any mycobacterial result: normal flora
 - Any mycobacterial result: microorganisms
 - Nontuberculous mycobacteria - pre 2010
 - Mycobacterial tuberculosis
- Fungus - encounter level
 - Aspergillus Candida Scedosporium species
 - Other bacterial or fungal microorganism species, specify
- Fungus - summarized by calendar year
 - Aspergillus Candida Scedosporium species
 - Other bacterial or fungal microorganism species, specify

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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 with 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 wth 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 / Tezacaftor / 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 / Tezacaftor / Ivacaftor (i.e., Trikafta)
 - First date taking Elexacaftor / Tezacaftor / 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

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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
 - Ascites
 - 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
 - Gallstones
 - Gallstones requiring surgery/procedure
 - Liver disease, cirrhosis
 - Esophageal varices
 - Gastric varices
 - GI bleed related to varices
 - Splenomegaly
 - Hypersplenism
 - Ascites
 - 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 enzymes elevated (for years 2003-2009)
- Other complications - encounter level
 - Patient does not have any complications
 - Arthritis/Arthropathy
 - Bone fracture
 - Osteopenia
 - Osteoporosis
 - Allergic Bronchial Pulmonary Aspergillosis
 - Asthma
 - Hemoptysis, massive
 - Hemoptysis type
 - 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
- Sinus surgery in the calendar year was the first
- Sinus surgery in the calendar year
- 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
 - Urinary incontinence

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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

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IX. Advance Lung Disease (ALD)

- ALD Initiation
 - Date patient flagged for ALD
 - Massive Hemoptysis (>240 ml)
 - Pneumothorax (recurrent and/or persistent)
 - Rapid rate of FEV1 decline
 - Respiratory failure (requiring ICU admission)
 - Other
 - FEV1pp<40% when stable
 - Daytime supplemental oxygen at rest
 - Pulmonary Hypertension
 - Six-minute walk distance <400m
 - Hypercarbia
 - Referred for lung transplantation evaluation
 - Supplemental oxygen requirement with exercise or sleep
 - First year FEV1pp less than 40
- ALD data summarized by calendar year with flags
 - Did patient have a thoracic procedure during the reporting year?
 - Thoracic procedure type: Lung resection
 - Thoracic procedure type: Thoracotomy
 - Thoracic procedure type: Video thoracotomy
 - Thoracic procedure type: Tube thoracotomy
 - Thoracic procedure type: Pleurodesis
 - Pleurodesis side: Unilateral
 - Pleurodesis side: Bilateral
 - Was patient enrolled in pulmonary rehabilitation during the reporting year?
 - Was the patient seen by a palliative care specialist during the reporting year?
 - Documented advance care planning conversation
 - Documentation of an advance directive
 - Referral to palliative care specialist
 - Referral to hospice
 - Referral to outpatient/community palliative care
 - Number Embolizations For Hemoptysis
 - Last embolization date
 - Was patient admitted to ICU this year?
 - Number of ICU admissions
 - Last ICU admission date
 - Was ECHO performed during the reporting year?
 - Date of last echocardiogram
 - Could PA pressure be estimated?
 - Estimated RVSP or PASP, mmHg
 - Tricuspid annular plane systolic excursion, mm
 - Was Right Heart Catheterization (RHC) done
 - Date of last RHC
 - Mean PA pressure from RHC, mmHg
 - Systemic systolic pressure, mmHg

- Systemic diastolic pressure, mmHg
- Heart Rate
- PA systolic pressure, mmHg
- PA diastolic pressure, mmHg
- Pulmonary capillary wedge pressure (PCW), mmHg
- Cardiac Index (L/min/m²)
- Number of six-minute walk tests
- Number of blood gas analyses
- Was transplant discussed with patient
- Was patient referred to a transplant program
- Patient stable
- Substance misuse (alcohol, marijuana, illicit drugs etc.)
- Chronic opioid use
- Smoking tobacco
- Other
- Inadequate medical insurance
- Limited social support
- Adherence issues
- Poor nutritional status
- Microbiological reasons
- Mental health
- Uncontrolled diabetes
- Physical inactivity/deconditioning
- Patient declined to pursue
- Was patient referred to a transplant program
- State of primary transplant program
- Primary transplant program
- Was patient evaluated for transplant at primary transplant program?
- Transplant evaluated outcome: listed
- Transplant evaluated outcome: deferred
- Transplant evaluated outcome: declined
- Transplant evaluated outcome: unknown
- Transplant evaluated outcome: denied
- Date listed for transplant
- Was the patient removed from the transplant waitlist
- Reason remove from waitlist: Condition improved
- Reason remove from waitlist: Patient declined transplant
- Reason remove from waitlist: Too sick to transplant
- Reason remove from waitlist: Died
- Reason remove from waitlist: Other
- Reason deferred: Patient stable
- Reason deferred: Additional testing necessary
- Reason deferred: Poor nutritional status
- Reason deferred: Other
- Reason deferred: Limited social support
- Reason deferred: Financial hardship
- Reason deferred: Substance misuse
- Reason declined: Too sick
- Reason declined: Microbiological reasons
- Reason declined: Limited social support

- Reason declined: Substance misuse
 - Reason declined: Malignancy
 - Reason declined: Poor nutritional status
 - Reason declined: Other, specify
 - Reason declined: Renal disease
 - Reason declined: Liver disease
 - Reason declined: Financial hardship
 - Was pt referred to another program for second opinion?
 - State of second transplant program
 - Second transplant program
 - Outcome if patient evaluated for transplant at second program: Listed
 - Outcome if patient evaluated for transplant at second program: Deferred
 - Outcome if patient evaluated for transplant at second program: Declined
 - Outcome if patient evaluated for transplant at second program: Unknown
 - Outcome if patient evaluated for transplant at second program: Denied
- ALD ICU admission
 - Date of admission to ICU
 - Date of discharge from ICU
 - Primary reason for admission:
 - Primary reason for admission:
 - Primary reason for admission:
 - Primary reason for admission:
 - Primary reason for admission:
 - Primary reason for admission:
 - Primary reason for admission:
 - Primary reason for admission:
 - Was patient intubated?
 - Date of intubation
 - Date of extubation
 - Was a tracheostomy performed?
 - Was patient placed on ECMO/Novalung?
 - Date ECMO/Novalung started
 - Date ECMO/Novalung ended
 - Cannulation strategy used: VV-Single Cannula
 - Cannulation strategy used: Code 2VV-Double Cannula
 - Cannulation strategy used: Veno-Arterial
 - Cannulation strategy used: Veno-Arterial-Venous
 - Cannulation strategy used: Unknown
 - Was ECMO/Novalung used in combination with ventilator?
- ALD Blood Gas
 - Date of blood gas analysis
 - Type of blood gas analysis:
 - Type of blood gas analysis:
 - Type of blood gas analysis:
 - Type of blood gas analysis:
 - Type of blood gas analysis:

- Blood pH
 - PaO₂, mm Hg
 - PaCO₂, mm Hg
- ALD Embolization
 - Embolization date
- ALD Six min walk
 - Date of six-minute walk test
 - Six minute walk distance, m
 - Was supplemental oxygen titrated?

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