# Cystic Fibrosis*

**Clinical Manifestations**

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|---|---|
| **Pulmonary** | Chronic airway disease w/ infection (H flu, S. aureus, P aeruginosa, Burkholderia, Steno, MRSA, atypical), bronchiectasis, gas trapping, hypoxemia, hypercarbia |
| **Sinus** | Sinus infections, nasal polyposis |
| **GI** | Meconium ileus, constipation, distal intestinal obstructive syndrome, deficiencies in A, D, E, K |
| **Endocrine** | CF related diabetes, osteoporosis from vitam D deficiency |
| **MSK** | Hypertrophic osteoarthropathy |
| **Reproduction** | Congenital absence of vas deferens |
| **Renal** | Nephrolithiasis due to chronic metabolic acidosis |
| **Hematologic** | Recurrent venous thrombosis due to chronic inflammatory state |

**Diagnosis**

- **Diagnostic Criteria:** CF in 1+ organ system AND evidence of CFTR dysfunction through either elevated sweat chloride, two disease causing mutations, or abnormal nasal transepithelial potential difference
- **Sweat Test: ≤6 mos:** normal ≤29 mmol/L and abnormal ≥60 mmol/L, **≥6 mos:** normal ≤39 mmol/L and abnormal ≥60 mmol/L
- **Newborn Screen:** Massachuesetts NBS measures immunoreactive trypsinogen (IRT) by radioimmunoassay or enzyme-linked immunoassay
- **CFTR Genetic Analysis**

**Pulmonary Exacerbations**

**Symptoms:** Increased cough, change in sputum color/quantity, decreased appetite, weight, tachypnea

**Chronic Pulmonary Treatment**

- **Agents to increase mucus clearance:** Pulmozyme, albuterol, inhaled hypertonic saline, chest PT
- **Anti-inflammatory therapy:** Azithromycin if P. aeruginosa
- **Persistent Pseudomonas Colonization:** Inhaled tobramycin and aztreonam
- **Vaccines:** pneumococcal, yearly influenza
- **Supplemental O2:** If intermittent or chronic hypoxemia
- **Nutritional support:** pancreatic enzymes, replacement of fat-soluble vitaminas, nutritional counseling
- **CFTR modulators:** Ivacaftor "Kalydeco" (CFTR potentiator for C551D mutation) and Lumacaftor/ Ivacaftor "Orkambi" (CFTR potentiator + corrects the Phe508del mutation and increases amount of functional CFTR at surface)
- **Annual Screening:** OGTT if >12, abdominal US w/ Doppler, audiogram

**Treatment CF Exacerbations**

**Lab monitoring:** Qweek (CBC diff, LFTs, CRP), Qmon/Thurs (BUN/Cr, Abx trough)

| Class | Antibiotic | Dose | Side Effects | Monitoring |
|---|---|---|---|---|
| Amino-glycoside | Tobramycin | IV 10 mg/kg q24 *OR* INH 300 mg BID *OR* Podhaler 4 caps INH BID | Ototoxicity<br>Nephrotoxicity<br>Phototoxicity | Peak/trough w/ 2nd dose, goal peak is 20-40, tough < 1 (IV only) |
| | Amikacin | IV 30 mg/kg q 24 or INH 250mg BID | | Peak<br>AFB= 20-30<br>PSA or Short term dosing =40-60<br>Trough < 2.5 |
| B lactams | Meropenem<br>Imipenem | IV 40 mg/kg q8 (max 2g q8) | Transaminitis<br>GI intolerance | |
| | Ceftaroline (5th generation cephalosporin) | 15 mg/kg/dose IV Q8 (max 600 mg IV Q8hrs) | | |

**Cystic Fibrosis continued on next page →**