Guide for Adult CF Care

Background

Epidemiology of CF

Cystic fibrosis is the most common life shortening recessively inherited disease in Caucasians. Although most common in Caucasians, occurring at a rate of 1 in 3300, it also occurs in Hispanics (1 in 9500), Native Americans (1 in 1:11,200) African-Americans (1 in 15,300), and in Asian-Americans (1 in 32,000). When the disease was first described in 1938, most children died in early childhood. However, as improved therapies have become available, survival has increased substantially such that in 2012, the median survival for patients with CF had improved to over 37 years old. Thus, nearly half of the 30,000 patients with CF in the U.S are over 18 years of age.

UNM School of Medicine currently cares for approximately 130 patients with cystic fibrosis, adults and pediatrics combined; in 2011 there were 71 adult patients being care for at our CF center. (18 years and above)

Pathophysiology of CF

CF is caused by mutations in the Cystic Fibrosis Transmembrane Conductance Regulator (CFTR) gene, which encodes a protein of the same name. CFTR belongs to a family of proteins involved in ion transport. There are now approximately 1900 known CF causing mutations; some forms of the mutation seem to confer increased disease severity. CFTR acts as a chloride transport channel, and also is known to have effects on other ion channels, such as the epithelial sodium channel, ENaC. Defective transport of sodium and chloride across the respiratory epithelial cell membrane leads to dehydration of the periciliary liquid and mucous layers overlying the cells, resulting in defective mucociliary and cough clearance. Furthermore, the airways become infected with pathogenic bacteria because of thick viscous secretions; this sets off an inflammatory cascade that over time causes significant bronchiectasis and airway inflammation. As the body attempts to eradicate the infection, neutrophils
accumulate. The neutrophilic damage is two-fold: initially neutrophils release elastase and reactive oxygen species that result in cell damage, and later these neutrophils become apoptotic and release their contents resulting in thickening of the mucus, and further impairing mucociliary clearance. With persistent infection and periodic exacerbations of the chronic infection, progressive lung disease develops. Thus, in spite of the remarkable increase in survival in CF, the overwhelming majority of patients still die from respiratory failure. Pulmonary complications such as massive hemoptysis and pneumothorax occur in patients with end-stage disease.

Although the lungs are the primary organ affected in most CF patients, CF is multisystem disease. The same thick secretions that occur in the lung also occur in the gut. This abnormality can result in pancreatic insufficiency, with malabsorption of calories and fat-soluble vitamins (A, D, E, and K), propensity towards constipation and distal intestinal obstructive syndrome, cholestasis, and sometimes chronic pancreatitis. Thirty-four percent of adult patients also have cystic fibrosis-related diabetes. Other complications in CF adults include osteopenia and osteoporosis and arthritis, as well as chronic liver disease which may be so severe a patient will require liver transplant.

Pulmonary Exacerbations

The overwhelming majority of the care of patients with CF is in the outpatient setting, (64.9% of patients at UNM were NOT treated with IV antibiotics in 2011) however, patients who fail outpatient management of pulmonary exacerbations, or are too sick for outpatient management, are admitted to manage their “exacerbations.” Although there are a few studies in select populations demonstrating equal efficacy between home and in-patient care for CF exacerbations, multiple studies have demonstrated that in-patient management of CF flares results in superior outcomes compared to home IV therapy. Review of the literature by the CFF consensus committee led them to conclude that there is currently insufficient evidence to recommend that home care is equal to in-patient care. This is especially true for NM, where lack of home health care services often precludes adequately being able to treat patients at home. Prior to the standard admission of CF patients for severe CF
flares/failure of outpatient management per the CFF care guidelines, the median FEV₁ of adult CF patients at UNM was 52% (national average 65%). Currently, the median FEV₁ of adult CF patients is above the national average at 63%. Following standards of care does work, and everyone plays a role in helping to make it happen!

REFERENCES
<table>
<thead>
<tr>
<th>Treatment</th>
<th>Recommendation</th>
<th>Certainty of Net Benefit</th>
<th>Estimate of Net Benefit</th>
<th>Recommendation</th>
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<tbody>
<tr>
<td>Ivacaftor*</td>
<td>For individuals with CF, 6 years of age and older, with at least one G551D CFTR mutation, the Pulmonary Clinical Practice Guidelines Committee strongly recommends the chronic use of ivacaftor to improve lung function and quality of life and reduce exacerbations.</td>
<td>High</td>
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<td>Inhaled aztreonam—mild disease†</td>
<td>For individuals with CF, 6 years of age and older, with moderate to severe lung disease and Pseudomonas aeruginosa persistently present in cultures of the airways, the CF Foundation strongly recommends the chronic use of inhaled aztreonam to improve lung function and quality of life.</td>
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<td>Chronic use of ibuprofen (age &lt; 18 yr)</td>
<td>For individuals with CF, between 6 and 17 years of age, with an FEV₁ ≥ 60% predicted, the CF Foundation recommends the chronic use of oral ibuprofen, at a peak plasma concentration of 50–100 μg/ml, to slow the loss of lung function.</td>
<td>Moderate</td>
<td>Moderate</td>
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<td>Chronic use of ibuprofen (age ≥ 18 yr)</td>
<td>For individuals with CF, 18 years of age and older, the CF Foundation concludes that the evidence is insufficient to recommend for or against the chronic use of oral ibuprofen to slow the loss of lung function or reduce exacerbations.</td>
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<td>Azithromycin without P. aeruginosa</td>
<td>For individuals with CF, 6 years of age and older, without P. aeruginosa persistently present in cultures of the airways, the CF Foundation recommends the chronic use of azithromycin should be considered to reduce exacerbations.</td>
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<td>Chronic inhaled β₂-adrenergic receptor agonists</td>
<td>For individuals with CF, 6 years of age and older, the CF Foundation concludes that the evidence is insufficient to recommend for or against chronic use of inhaled β₂-adrenergic receptor agonists to improve lung function and quality of life or reduce exacerbations.</td>
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<td>Oral antistaphylococcal antibiotics, chronic use</td>
<td>For individuals with CF, 6 years of age and older, with Staphylococcus aureus persistently present in cultures of the airways, the CF Foundation concludes that the evidence is insufficient to recommend for or against the chronic use of oral antistaphylococcal antibiotics, chronic use...</td>
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<tr>
<td>Inhaled tobramycin—moderate to severe disease*</td>
<td>For individuals with CF, 6 years of age and older, with moderate to severe lung disease and <em>Pseudomonas aeruginosa</em> persistently present in cultures of the airways, the CF Foundation strongly recommends the chronic use of inhaled tobramycin to improve lung function and quality of life, and reduce exacerbations.</td>
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<td>Inhaled tobramycin—mild disease*</td>
<td>For individuals with CF, 6 years of age and older, with mild lung disease and <em>P. aeruginosa</em> persistently present in cultures of the airways, the CF Foundation recommends the chronic use of inhaled tobramycin to reduce exacerbations.</td>
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<td>Dornase alfa—moderate to severe disease*</td>
<td>For individuals with CF, 6 years of age and older, with moderate to severe lung disease, the CF Foundation strongly recommends the chronic use of dornase alfa to improve lung function, improve the quality of life, and reduce exacerbations.</td>
<td>High</td>
<td>Substantial</td>
<td>A</td>
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<tr>
<td>Dornase alfa—mild disease*</td>
<td>For individuals with CF, 6 years of age and older, with asymptomatic or mild lung disease, the CF Foundation recommends the chronic use of dornase alfa to improve lung function and reduce exacerbations.</td>
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<td>Inhaled hypertonic saline</td>
<td>For individuals with CF, 6 years of age and older, the CF Foundation recommends the chronic use of inhaled hypertonic saline to improve lung function and quality of life and reduce exacerbations.</td>
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<tr>
<td>Azithromycin with <em>P. aeruginosa</em></td>
<td>For individuals with CF, 6 years of age and older, with <em>P. aeruginosa</em> persistently present in cultures of the airways, the CF Foundation recommends the chronic use of azithromycin to improve lung function and reduce exacerbations.</td>
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<td>Oral antistaphylococcal antibiotics, prophylactic use</td>
<td>For individuals with CF, the CF Foundation recommends against the prophylactic use of oral antistaphylococcal antibiotics to improve lung function and quality of life or reduce exacerbations.</td>
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<td>Inhaled corticosteroids</td>
<td>For individuals with CF, 6 years of age and older, without asthma or allergic bronchopulmonary aspergillosis, the CF Foundation recommends against the routine use of inhaled corticosteroids to improve lung function or quality of life and reduce pulmonary exacerbations.</td>
<td>High</td>
<td>Zero</td>
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<tr>
<td>Oral corticosteroids</td>
<td>For individuals with CF, 6 years of age and older, without asthma or allergic bronchopulmonary aspergillosis, the CF Foundation recommends against the chronic use of oral corticosteroids to improve lung function, quality of life or reduce exacerbations.</td>
<td>High</td>
<td>Negative</td>
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<tr>
<td>Other inhaled antibiotics</td>
<td>For individuals with CF, 6 years of age and older, with <em>P. aeruginosa</em> persistently present in cultures of the airways, the CF Foundation concludes that the evidence is insufficient to recommend for or against the chronic use of other inhaled antibiotics (i.e., ceftriaxone, colistin, gentamicin) to improve lung function and quality of life or reduce exacerbations.</td>
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<tr>
<td>Oral antipseudomonal antibiotics</td>
<td>For individuals with CF, 6 years of age and older, with <em>P. aeruginosa</em> persistently present in cultures of the airways, the CF Foundation concludes that the evidence is insufficient to recommend for or against the routine use of chronic oral antipseudomonal antibiotics to improve lung function and quality of life or reduce exacerbations.</td>
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<tr>
<td>Leukotriene modifiers</td>
<td>For individuals with CF, 6 years of age and older, the CF Foundation concludes that the evidence is insufficient to recommend for or against the routine chronic use of leukotriene modifiers to improve lung function and quality of life or reduce exacerbations.</td>
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<tr>
<td>Inhaled or oral N-acetylcysteine, or inhaled glutathione</td>
<td>For individuals with CF, 6 years of age and older, the CF Foundation concludes that the evidence is insufficient to recommend for or against the chronic use of inhaled or oral N-acetylcysteine or inhaled glutathione to improve lung function and quality of life or reduce exacerbations.</td>
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Quick Guide To Daily Rounds for care of patients with CF (see table for detailed recommendations for inpatient stays)

Patients will be admitted to the Silver Medicine Team. Daily rounds on mutual CF patients will be done together; ie the medicine team and pulmonary team will round together as much as possible. This is to ensure optimal communication and patient care. The pulmonary fellow is responsible for contacting the medicine intern who is on Silver medicine to coordinate times to round each day.

Karen Fiato, CF NP rounds independently on the CF inpatients two days a week, generally on Tuesdays and Wednesdays. Karen is not here on Mondays. Acute issues should go to the pulmonary consult team.

Fridays are CF team rounds at 12:30. The entire CF team will round together on CF patients. On team round days, the consult notes on patients should be sent to Theresa Heynekamp by the pulmonary fellow. Medicine intern and attending should plan on attending these rounds.

When a patient is admitted, the medicine team does the History and Physical. CF team will generally (but not always) do a same day clinic note if possible. When a patient is ready for discharge, the medicine team will do the discharge summary.

**RESPIRATORY**

- Patients should be receiving:
  - Airway clearance therapy QID with Albuterol
  - Azithromycin is used in patients as an anti-inflammatory given three days a week, in patients with pseudomonas, though it is now recommended that patients who aren’t colonized with pseudomonas also receive this. *(Patients MUST be AFB negative however)*
  - We typically hold inhaled home antibiotics, given that they are receiving IV abx during their stay
  - Pulmozyme and Hypertonic saline BID unless contraindication exists; NOT all patients use hypertonic saline.
  - Hypertonic saline should not be initiated in a Hypersal-naïve patient prior to doing bronchoprovocation testing.
  - Physical therapy daily (check to make sure consult received). If patients do not want to go downstairs they can have inroom ‘bands’
  - PFTs once per week (will be done by Bill Demaray). *(Does NOT need to be ordered, CF team does this.*

- Wean O₂ to keep sats ≥ 92% (this often requires you doing it on rounds)
- Consider oral steroids for patients with significant RAD component, though we usually do not need to do this.

- **Hold Airway clearance therapy for frank hemoptysis, but not streaks. Hypertonic saline should be held for hemoptysis > 5ml.**

- For questions/concerns about ACT or pulmonary therapies, call Bill Demaray, CF RRT (925-7408, 480-4716)
FEN/GI

• Patients should be receiving high protein, high fat diet + snacks/supplements. If they have CF related diabetes, they should get same diet—they should NOT get ADA diets.
• CF patients are prone to DIOS, especially if on narcotics. Make sure BMs are at least QD and patients should have stool softeners written for.
• F/u vitamin levels done on admission
• Weights at least twice per week
• Ensure that tube feeds are tolerated

Patients with CFRD are seen by Linda Reineke(CF RD), who leaves a note with her insulin recommendations.
• For diet, tube feed regimens, CFRD questions, or vitamin level questions, call Linda Reineke, CF RD/CDE (951-3010)

ENDOCRINE

• Assess adequacy of blood glucose control on current regimen.
  - Patients often require increased insulin when experiencing a flare (or may progress from glucose intolerance to frank CFRD during flare)
  - Check with patient and on MAR to determine if recorded BG are pre/post prandial
• Patients should NOT be told to limit carb/calorie intake or be put on an ADA diet.

• For assistance with changes in insulin regimen, call Linda Reineke, CF RD/CDE. Please do not change insulin without discussing with CF team.

ID

• Antibiotics should always be IV (including cipro) and max dose because of rapid metabolism/poor absorption in CF
  (note that chronic PO azithro, used as anti-inflammatory/anti-biofilm in CF, should be continued)
• Always use two anti-Pseudomonals (Aminoglycoside + β-lactam) when treating P. aeruginosa
• For Tobra dosing, check last pharmacy note (standard dosing is 3.3mg/kg/ and should be dosed q24 hours (pharmacy does this) but some patients have required adjustment to q12 because of clearance)
  Peak/troughs with 4th dose
  For Q 12 dosing, goal peak/trough: 10-14, <1.5; for q 8 dosing, 8-10, <2;
  **For assistance with pharmacokinetics, please talk to 5W pharmacist!!!

• Do NOT change antibiotic regimen based on susceptibilities alone; also don’t change antibiotics without discussing with CF team.
• If patient shows no symptomatic/pft improvement after 1 week, or if new fevers/increased O2 requirement, than we may rethink antibiotic regimen.

• Temp spike >38.5 warrants blood cultures, particularly if patient has B. cepacia or M. abscessus

Infection Control
• All CF patients should be on CONTACT precautions (gowns, gloves) **Masks should only be worn if there is clinical concern for viral influenzæae or there are other indications per hospital policy to be on droplet precautions.**

• Patients with *B. cepacia* are generally housed on different ward if other CF patients are in-house; in addition, patients with Mycobacterium abscessus should also be housed on different floor if possible.

**CF patients, even those on isolation, can leave their rooms to go for walks, go to PT, etc with the appropriate infection control wear (mask for everyone).**

**Psychosocial**
• CF social worker, Trina Zajllzer, LISW, sees patients at least two to three times a week to help with school work/work excuses, and general psychosocial support. Please touch base with her a few times a week to assess further needs/interventions.

**Discharge planning**
• Some patients are candidates for completing IV antibiotics at home. If patient is candidate for completing antibiotics at home (verify that patient is candidate with the pulmonary team,) please coordinate with Karen Fio/o/s/gen med/5W discharge planner at least 2-3 days prior to planned discharge.

  The CF Team does NOT have their own discharge planner; please keep this in mind when planning discharge needs for your patients.
  • Karen Fiao will arrange follow-up apt (at end of home IV course, or within 1 month of hospital discharge if entire course completed in-house)

**General**
Weekly labs: CBC, Chem 7, LFTs, aminoglycoside levels (these are generally ordered by pharmacy)
  We do **NOT** need daily labs unless there is an acute issue

Patients should generally NOT be put on heparin/lovenox for DVT prophylaxis. (if they have pancreatic insufficiency, they are vitamin k deficient.). Also most CF patients are quite ambulatory and walk around daily. However, there are exceptions to this in certain patients but we will inform primary team.

**IMPORTANT PHONE NUMBERS**

<table>
<thead>
<tr>
<th>Name</th>
<th>Role</th>
<th>Pager</th>
<th>Phone number</th>
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<tbody>
<tr>
<td>Bill Demaray</td>
<td>CF RT</td>
<td>951-4297</td>
<td>925-7408, 480-4716</td>
</tr>
<tr>
<td>Trina Zajller</td>
<td>CF SW</td>
<td>951-3643</td>
<td>272-3806</td>
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<tr>
<td>Linda Reineke</td>
<td>CF Dietitian/Diabetes Educator</td>
<td>951-3010</td>
<td>272-0172</td>
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<tr>
<td><strong>PHYSICIAN/CONSULTATION INVOLVEMENT</strong></td>
<td><strong>ADMISSION</strong></td>
<td><strong>ACUTE PHASE (Days 2-6)</strong>*</td>
<td><strong>RESOLUTION PHASE (Days 7-12)</strong>*</td>
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| · Inform CF Center MD if patient is not being admitted from clinic/home  
· Assess need for consultation with GI, ENT, Surgery, Endo, etc  
· Order PICC placement of no port | · Routine surveillance  
· Consult pharmacist for pharmacokinetics  
· Reassess need for consultations | · Routine surveillance  
· Consult pharmacist for pharmacokinetics  
· Reassess need for consultations | · Review and discuss discharge plans with patient including medication, ACT, and nutritional interventions  
· Write Rx for new meds/refills  
· Summarize response to treatment/assess and discuss patient’s current disease status |

| **NURSING** | · Infection control measures  
· Admission assessment  
· Vitals q shift  
· Bowel function QD  
· Resp assessment q shift  
· Check IV site q shift  
· Encourage ambulation as tolerated | · Assess adherence to medications  
· Vitals q shift  
· Bowel function QD  
· Resp assessment q shift  
· Check IV site q shift  
· Encourage ambulation as tolerated | · Encourage supervised exercise  
· Contact appropriate resources needed for discharge | · Contact appropriate resources needed for discharge  
· Teach appropriate skills for home care (if applicable)  
· Ensure that medications are ordered for home use  
· D/c IV at end of treatment course |

| **EDUCATION** | · Orient patient to unit and review infection control measures | · Proper medication use  
· Instruct in or review ACT  
· Dietary counseling (high-calorie diet and proper enzyme use)  
· Observe and instruct family members in all of the above as appropriate  
· Review infection control measures | · Proper medication use  
· Instruct in or review ACT  
· Dietary counseling (high-calorie diet and proper enzyme use)  
· Review infection control measures | · Instruct patient in proper use of maintenance medications and performance of ACT  
· Discuss maintenance diet and use of enzymes |

| **NUTRITION** | · Weight  
· Height  
· Order high cal/high protein diet with snacks and supplements  
· RD eval of intake, enzyme dosing, vitamin replacement, glucose intolerance | · Recheck weight  
· Evaluate adequacy of intake | · Recheck weight  
· Reevaluate adequacy of intake  
· Intensify nutrition support if weight is not back to baseline | · Weigh patient prior to discharge and record in computer |

| **PSYCHOSOCIAL** | · Assessment of psychosocial needs by CF SW  
· Consult psych if indicated | · Ongoing CF SW involvement  
· Plan for and implement continuity of education  
· Provide letter for employer prn  
· Assess appropriateness for passes | · Ongoing CF SW involvement  
· Implement plan for education needs  
· Assess need for family meeting  
· Assess appropriateness for passes | · Assure continuity of SW/psych in outpatient setting  
· Make plans for patient’s return to work/school |
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<td>CBC</td>
<td>F/u oximetry if admit SaO₂ decreased from baseline</td>
<td>F/u oximetry if admit SaO₂ decreased from baseline</td>
<td>F/u oximetry if admit SaO₂ appears patient will need home new rx for home O₂</td>
<td>Extended oximetry/ABG if appears patient will need home new rx for home O₂</td>
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<td>PFTS (+lung volumes/bronchodilator response)</td>
<td>Repeat PFTS (+lung volumes/bronchodilator response)</td>
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<td>PFTS if not done on admit</td>
<td>Aminoglycoside levels with 3rd/4th dose</td>
<td>Aminoglycoside levels/other drug levels if not adequate on initial testing</td>
<td>Aminoglycoside levels/other drug levels if not adequate on initial testing</td>
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<td>IV antibiotics based on most recent culture</td>
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<td>IV antibiotics based on most recent culture</td>
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<thead>
<tr>
<th>DISCHARGE PLANNING</th>
<th></th>
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</thead>
<tbody>
<tr>
<td>If patient candidate for home IV therapy, assess home support/agency availability/home equipment</td>
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<td>If patient candidate for home IV therapy, assess home support/agency availability/home equipment</td>
<td>If patient candidate for home IV therapy, teach skills necessary/contact home care co on discharge</td>
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<td></td>
<td>Provide CF clinic f/u date</td>
<td>Provide CF clinic f/u date</td>
<td>Provide consultant clinic f/u date</td>
<td>Provide consultant clinic f/u date</td>
</tr>
</tbody>
</table>

*Stay may be extended if patient is worsening/not improving.
Adapted from CFF Guidelines: “Treatment of Pulmonary Exacerbations”
Reading List

GENERAL


GENETICS


RESPIRATORY


LUNG TRANSPLANT


GASTROENTEROLOGY


ENDOCRINOLOGY AND METABOLISM


MISCELLANEOUS


