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Review Article: Care of the Hospitalized Patient with Cystic Fibrosis: A Summary of Current Practice Guidelines; Recommendations for the Hospitalist, Part 2

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In a short review of our last discussion, I will remind the reader that cystic fibrosis is a complex genetic disease affecting many organs, but it is often lung disease that brings the patient to our attention. The natural history of lung disease begins with the production of abnormal mucus (a result of the gene mutations associated with this disease). We refer to a vicious cycle of disease, including early and persistent infection of the airway, concomitant inflammatory response, and, over time, progressive airway obstruction. These processes ultimately result in respiratory failure. As disease worsens there is an increased likelihood of respiratory complications. In addition, patients with cystic fibrosis often present with multiple manifestations of disease upon presentation to the hospital, complicating assessment and management. This second installment of recommendations for the hospitalist will briefly address these complications to assist in the management of these patients.

The cystic fibrosis care team often acts as the gate keeper for hospital admissions given the specialized ability to recognize a pulmonary exacerbation. However, patients may present to the emergency department with symptoms requiring urgent attention, such as massive hemoptysis or pneumothorax. The following recommendations are summarized from guidelines provided by the Cystic Fibrosis Pulmonary Therapies Committee. The CF Foundation pulmonary therapies committee consists of a multi-disciplinary group including representative physicians, nurses,

respiratory therapists, physical therapists, pharmacists, CF families and CF Foundation staff. High volume cystic fibrosis centers participated in a questionnaire, a literature search was performed, and an expert panel reviewed all of the data collected. This information was brought to the committee for review with eventual creation of these guidelines following group consensus.

Hemoptysis:

Hemoptysis is a common symptom in patients with CF; therefore it is important for care providers to inquire about bleeding in the outpatient setting. The literature indicates that about 9% of patients report hemoptysis, which the author estimates to be the average at this center as well. Often there is scant-to-moderate bleeding, described as streaks or spots in the sputum. However, on occasion the patient will describe a large volume of blood. It is known that massive, life-threatening bleeding can occur. The literature indicates 4% of all patients with cystic fibrosis will suffer a massive hemoptysis during his or her lifetime. Frequently patients have difficulty in defining the volume of blood. The author is able to recall only one patient with an accurate measurement. While driving his car, he reported an episode of hemoptysis where he coughed up blood into his coffee cup and filled it to approximately 8 ounces. However, patients often expectorate into the sink or the toilet making volume estimation difficult. The patient may overestimate volume, particularly if it is his or her first episode of hemoptysis. Based on volume of expectorate, other symptoms, and circumstances surrounding the hemoptysis, we elect either to monitor or treat. We have asked our patients to contact us with (1) The first episode of hemoptysis, and (2) Expectored blood greater than a teaspoon with or without changes in pulmonary symptoms.

We also would recommend treatment if patient has other pulmonary symptoms with ongoing small-volume hemoptysis (greater than 5 ml, but less than 50 ml). Treatment may consist of oral antibiotics at home but may also require an admission to hospital. We prefer to observe and evaluate patients with hemoptysis of 50-250 mL, even if they do not have other pulmonary symptoms.

The patient with massive hemoptysis should always be admitted to the hospital. This can be life-threatening.

Admission recommendations include:

1. Discontinue any NSAIDs;
2. Hold nebulized hypertonic saline;
3. Administer vitamin K to correct possible deficiency (recall that CF patients often experience malabsorption of fat soluble vitamins);
4. Treatment for pulmonary bacterial colonization in patients with changes in sputum, increased cough, or other symptoms suggestive of a pulmonary exacerbation. Recall Pseudomonal infection is treated with two anti-pseudomonal medications (with a consult to the cystic fibrosis team);
5. Continue airway clearance including chest physiotherapy with vest, bronchodilators, Pulmozyme, and/or inhaled antibiotics. We recognize that mucus

plugging and the resulting inflammation of the airway is likely the etiology of the bleeding; therefore we will need to address this. However, we may hold IPV and if bleeding continues or increases, consider holding Pulmozyme first and then other modalities for short time until bleeding subsides;

6. In the case of massive hemoptysis, we like to alert the bronchoscopy lab, Intensive Care Unit, and Interventional Radiology in case of acute decompensation. In addition, therapies that should be withheld include BiPAP, airway clearance therapies, all other inhaled medications, and NSAIDs. Vitamin K should still be administered. Pulmonary function testing should be held until significant bleeding has subsided.

Pneumothorax:

According to the literature, the average and annual incidence of pneumothorax is 0.64% or one in 167 patients per year. Along with hemoptysis, pneumothorax occurs more commonly in older patients with advanced disease. The author can recall only one pneumothorax in the past eight years of practice, occurring in a patient just before the time of her transplant who suffered pneumothorax while traveling to Denver by airplane.

1. Patients with a large pneumothorax should be admitted to the hospital, but those with small pneumothoraces who are clinically stable may be closely observed in an outpatient setting.
2. The patient with large pneumothorax should have a chest tube placed.
3. The patient with first pneumothorax should not go undergo pleurodesis. However patients with recurrent large pneumothoraces should undergo pleurodesis to prevent recurrence. The preferred method is surgical pleurodesis.
4. Since pneumothoraces typically occur in patients with advanced obstructive airways disease, one could argue for treatment with antibiotics: however, there is no consensus on this issue.
5. BiPAP should be withheld from patients with pneumothorax as long as the pneumothorax is present.
6. The patient with pneumothorax should not fly in a plane, not lift weights, and should not perform spirometry for 2 weeks after the pneumothorax has resolved.
7. Mechanical airway clearance methods such as Intrapulmonary Percussive Ventilation and Positive Expiratory Pressure (PEP) should be held in large pneumothorax, but there is no consensus on withholding vest or CPT.
8. Inhaled airway treatments should NOT be held unless cough is more severe with them.

GI spectrum of Constipation-Obstipation-DIOS (Distal Intestinal Obstructive Syndrome):

Complete or incomplete intestinal obstruction occurs when there is accumulation of viscous fecal material combined with sticky mucoid intestinal content, which adheres to the intestinal wall of the terminal ileum and proximal colon. DIOS is another common complication in cystic fibrosis. A significant mass of material may be interwoven and connected to the crypts and villi of the distal intestine and may prove difficult to remove. In many cases, this is a chronic

condition that can be aggravated intermittently. It may occur with a wide range of severity. Some incomplete obstruction cases may go unnoticed by the CF care team when patients assume they had a transient “GI bug” that resolved without medical intervention. Again, these complications seem to occur more often in adults than children, more frequently in pancreatic-insufficient patients (although occurs in pancreatic sufficient patients as well), and in post-transplant patients.

In constipation, the bowel symptoms and imaging indicate a long-standing condition. Fecal material is distributed throughout the colon. In DIOS, there is a right lower quadrant mass that may be palpable and is also seen on a plain film of the abdomen. It may present acutely or sub-acutely with intermittent abdominal pain associated with abdominal distention.

The phenomenon is again the result of the abnormal gene product CFTR (Cystic fibrosis Transmembrane Conductance Regulator). This is the same chloride channel found in lung epithelium, where chloride secretion and sodium absorption are the driving force for fluid secretion into the lumen. In the lung, a non-functional chloride channel causes dehydration of the mucus and subsequent plugging of small airways. In the intestine, abnormalities in the mucus also predisposes to obstruction. In addition, other factors influence the viscosity of luminal contents and gut motility. Inflammatory processes are theorized to contribute to neuromuscular dysfunction. Intestinal wall thickening has been seen involving the muscularis mucosa. Hypertrophy may be the consequence of dysmotility as well as the effect of viscid intestinal contents.

Risk factors for the development of GI problems include dehydration precipitated by illness (such as a pulmonary exacerbation), dehydration associated with hot weather, and CF-related diabetes which may alter hydration status.

Treatment for this GI spectrum includes oral rehydration combined with stool products for mild, incomplete obstruction or constipation. Osmotic laxatives, which draw more fluid into the intestinal lumen, are the most effective and best tolerated. Gut lavage with balanced electrolyte solutions and intravenous fluids are rarely required. Care should be taken to avoid rapid fluid shifts. We attempt to avoid dehydration and optimize enzyme dosing to decrease the risk of recurrence. Some patients seem to do better with prophylactic osmotic laxative therapy.

CF-Related Diabetes (ESRD):

CFRD is the most common comorbidity in cystic fibrosis and occurs in 40-50% of adults. The disease has features of both type I and type II diabetes mellitus; however it is a clinically distinct phenomenon. The primary cause is insulin insufficiency due to damage to the pancreas. However, fluctuating levels of insulin resistance also occur and are related to chronic inflammation with acute intermittent illnesses. Diagnosis is generally not made during acute hospitalization for pulmonary exacerbation or other illness but rather during a time of stable health.

Because the main pathology in CFRD is insulin insufficiency, the treatment is insulin. There is clear evidence that patients with insulin therapy who achieve good control of blood sugars also

demonstrate improvements in weight, protein anabolism, pulmonary function, and survival. There is no single proven insulin regimen, and each case requires individualized attention. CFRD patients still produce some endogenous insulin and except during acute illness, insulin needs are often low such as the case with type I diabetes in the so-called “honeymoon period.” It is during an acute illness or with oral steroid therapy that insulin requirements may steeply rise. Once the illness resolves, it may take 4-6 weeks for insulin requirements gradually to return to baseline.

The goals of glucose management are the same as ADA recommendations for all patients with diabetes. Hemoglobin A1c values less than 7% are the usual target. However, hemoglobin A1C is often low in cystic fibrosis patients secondary to an increased rate of turnover in red blood cells.

As you may recall from our first article, meeting nutrition requirements is essential in the management of this disease. Adequate caloric intake to maintain target BMI is critical to health and survival in cystic fibrosis patients. Therefore those patients with CFRD should NOT decrease calorie intake. These patients require a very high calorie diet that is usually 120-150% of the recommended daily intake for age. This is because there is increased resting energy expenditure with increased work of breathing and there is increased loss of calories through malabsorption.

CF evidence-based guidelines also recommend exercise for its benefit to overall health, cardiovascular fitness, and pulmonary clearance. Virtually all cystic fibrosis patients, even those with severe pulmonary disease, are capable of participating in strength training and aerobic exercise.

Cystic Fibrosis “Pearls “

- Cystic fibrosis is a progressive genetic disease, marked by episodes of pulmonary exacerbation
- Treatments are based on pathophysiology
- Airway clearance, nutrition, and antibiotics are key elements to manage in treatment
- A patient’s “usual” or home oral antibiotics are NOT stopped upon presentation to hospital for intravenous antibiotics
- Examples of treatments to promote airway clearance include bronchodilators, mucolytic agents, mucus hydration agents, inhaled antibiotics and mechanical clearance (vest, IPV, accapella, coronette, aerobika etc)
- Give CF patients vitamin K for hemoptysis
- Think about airway effects when considering treatments for patients with both hemoptysis and pneumothorax

- Antibiotics should be considered whenever there is a change in pulmonary symptoms; Pseudomonas is treated with two agents with different mechanisms of action
- Most cystic fibrosis patients have some degree of constipation and bowel management is always with osmotic agents
- Do not put CFRD patients on a calorie restricted diet
- Do not put CF patients on a salt-restricted diet

Resources:

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ASK A SPECIALIST: ASK A PATHOLOGIST

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