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Perspective

The Emperor has No Clothes: The Nonexistence of COPD and the Mismanagement of PALDS

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Abstract

The designation of COPD as "a disease" for public health purposes has led to significant misdiagnosis and treatment of individual patients. This article puts into perspective the myriad of persistent airflow limitation diseases (PALDs) and argues for their specific diagnosis and management in individual patients.

Keywords: *COPD; PALDS*

Introduction

GOLD is short for the Global Initiative for Chronic Obstructive Lung Disease, a collaboration between the National Institutes of Health and the World Health Organization. It is a public health effort.

Like most public health ("population medicine") efforts, they treat populations as homogenous and with a single condition or disease. It is for the "greater good." They ignore subgroup and individual differences to simplify understanding and interventions. The practice of Medicine, on the other hand, treat individuals. Clinicians treat the individual patient's disease or infirmity.

Thus, even though the practice of Medicine and Public Health have similar goals, they have dissimilar approaches. One deals with populations and the other with individuals. The melding of the two approaches by GOLD into one, has greatly diminished the medical knowledge regarding all the diseases with persistent airflow limitation found in individuals. And, without knowledge of the individual's disease, clinicians cannot competently manage them. The following presents the rationale for re-introducing all the Persistent Airflow Limiting Diseases (PALDs) back into clinical practice.

In order to reduce confusion, it is suggested the term "COPD" be retired. Instead PALDs for Persistent Airflow Limiting Diseases be use for public health efforts for the group of diseases with persistent airflow limitation In clinical practice the actual disease such as Diffuse Obstructive Emphysema, Chronic Bronchitis, Asthmatic Bronchitis, Small Airways Disease and the like (See Appendix) should be used as the diagnosis.

Current Clinical Misconceptions

According to The Global Initiative for Chronic Obstructive Lung Disease (GOLD) [1], Chronic Obstructive Pulmonary Disease (COPD), a common preventable and treatable disease, is characterized by persistent airflow limitation that is usually progressive and associated with an enhanced chronic inflammatory response in the airways and the lung to noxious particles or gases. Exacerbations and comorbidities contribute to the overall severity in individual patients.

Spirometry is required to make a clinical diagnosis of COPD; the presence of a post-bronchodilator FEV1/FVC < 0.70 confirms the presence of persistent airflow limitation and thus of COPD.

Clinical Reality

It is curious that COPD bronchodilators advertisements mention the terms "emphysema and chronic bronchitis". If COPD is 'a disease' what are emphysema and chronic bronchitis? Not COPD? COPD sub-types?

Also, ICD-10 diagnosis coding system requires the specification of COPD 'with emphysema' or 'with chronic bronchitis'. Again why? It also includes the diagnosis of 'asthmatic bronchitis'. Are these sub-types of a single disease?

In reality, COPD is NOT a disease but instead a large variety of diverse diseases requiring different treatments. What all these diseases have in common is that they have "persistent airflow limitation" on spirometry. Most do have dyspnea, chronic cough and/or sputum production; yet, all one has to do is look at their pathologies to see they are very different diseases. The diagnosing of these patient as "the disease COPD" causes their mismanagement.

Case Examples

1) A 36 yr black woman was referred because of 3 - 4 exacerbations of "COPD" per year since her early 30's. She denied ever smoking cigarettes; nevertheless, her PCPs continued to accuse her of smoking. She had every 2 - 3 month exacerbation of 'COPD' symptoms and required a month long Prednisone course to get better. She reported dyspnea and cough with her exacerbations but no sputum. Her PMH, SH, Occupational History, FH, Allergy history and rest of Review of Systems were all noncontributory.

Her physical exam revealed a short stocky woman with diffuse decrease breath sounds and no wheezing or rhonchi were present. The skin showed no lesions and the rest of the exam was unremarkable.

The Chest Radiograph revealed large bilateral apical bullae confirmed by CT scan.

Her Complete PFTs revealed: a) severe persistent airflow limitation, b) moderately severe decreased lung volumes, c) low diffusing capacity and d) increased airway resistance.

On laboratory evaluation, her serum angiotensin-converting enzyme (ACE) level was very high and her Lactate Dehydrogenase (LDH) level was only slightly elevated.

She was diagnosed with Bullous Sarcoidosis and treated with every other day Prednisone because of her frequent symptomatic exacerbations. She had no more "COPD exacerbations" In retrospect, her month long course of Prednisone every 2-3 months was intermittently (and unknowingly!) treating her Sarcoidosis symptoms in an unproven manner.

2) A 70 yr retired Executive was referred because of worsening COPD. He had smoked 2 PPD of cigarettes until age 50 before quitting due to medical advice. He had a 70 pack year smoking history. He had no breathing difficulties when he quit smoking and continue to have none until age 68. For the previous 2 years, he had experienced very gradual increase in dyspnea on exertion esp. while golfing. At age 68 he could walk the entire course but by age 70 he could not walk a single hole and required the use of a golf cart. He had no cough or sputum production. He was concerned that he would have to stop golfing. His PMH, SH, Occupational History, FH, Allergy history and rest of Review of Systems were all noncontributory. He was not a woodwind instrument player.

His physical exam revealed a well nourished, well developed man with mild barrel chest and diffuse decrease in breath sounds. No wheezing or rhonchi were heard. The rest of the exam was unremarkable.

His Chest Radiograph revealed hyperinflation. His laboratory evaluation was unremarkable.

His Complete PFTs revealed: a) severe persistent airflow limitation with FEV1 of 45%, b) his TLC was increased at 135% of Predicted c) his diffusing capacity was normal and d) his airway resistance was slightly elevated.

Small Airways Disease was suspected and he was prescribed a month long course of Prednisone along with inhaled bronchodilators. He returned in a month and reported that his dyspnea had resolved and he was again walking 18-holes of golf. The prednisone was tapered and the bronchodilators continued. He was referred back to his Primary Care Physician.

Updated Concepts

1) To call COPD "a disease" based on persistent airflow limitation alone is equivalent to calling Cancer "a disease" based solely on the presence of malignant cells. Or, calling chicken pox and small pox, "Pox Disease" because they both have skin lesions, i.e., "pox".

Clinically, malignant cells only indicates malignancy, i,e., the presence of one of a myriad of malignant neoplasms. It is then up to the clinicians to diagnose the specific type of neoplasm; so, it can be treated specifically. Even the same cell type such as "squamous cell" have markedly different ramifications if it is found on the skin versus the lung. Specificity is paramount to competent clinical management.

Analogously, clinically persistent airflow limitation only indicates the presence of a disease with persistent airflow limitation, i,e., the presence of one of a myriad of PALDs It is then up to the clinicians to diagnose the type of disease; so it can be treated specifically. Specificity is paramount to competent clinical management.

2) Thus, it is wrong to believe that persistent airflow limitation on Spirometry is caused by a single disease. Many diseases cause a persistent airflow limitation on spirometry.

3) Complete Pulmonary Function Tests are needed to differentiate between the varied and diverse PALDs. These include Spirometry, Lung Volumes, Diffusing Capacity, Airway Resistance and Maximum Voluntary Ventilation. Other more exotic testing may occasionally be needed.

Pathophysiology

1) Even though airway inflammation is common in PALDs, it is the varied ramifications of inflammation that crucially differentiate between the common PALDs. Again, one only needs to look at the PALDs pathologies to see the differences.

2) There are 4 major pathophysiologies causing persistent airflow limitation. Most PALDs have only one or two causes predominate and these features are usually disease defining.

a) Chronic Airway Narrowing due to airway edema from inflammation such as in Asthmatic and Chronic Bronchitis, mucosal hypertrophy like in Chronic Bronchitis, endobronchial lesions such as in Sarcoidosis and Bronchiolitis Obliterans and/or parabronchiolar lesions like Lymphangioleiomyomatosis or Tuberous Sclerosis.

b) Dynamic Airway Compression predominately expiratory due to loss of parenchymal structural support of airways such as in Diffuse Obstructive Emphysema or destruction of the airway wall supporting structures such as Bronchiectasis and Cystic Fibrosis.

c) Bronchospasm as seen in Asthmatic Bronchitis and possibly Chronic Bronchitis.

d) Secretions due to airway inflammation as seen in Asthmatic and Chronic Bronchitis, Bronchiectasis and Cystic Fibrosis

3) The most common PALDs that every clinician should know well are the following. The myriad of other PALDs (See Appendix) should be known by the specialists that care for them.

[CAVEAT: Research studies looking at therapies for "COPD" lump all the following diseases into one. Since they don't differentiate between them it is impossible to determine the proportion of each in the study and whether there are any differences in responses to their interventions between them. It is likely that most of the positive responses occur in one or another (but not all!) of the diseases. Reanalysis of these old studies (if possible) and/or new studies distinguishing between these diseases are critically needed].

a) Diffusive Obstructive Emphysema ("Pink Puffer")

Diffuse Obstructive Emphysema is caused almost exclusively (99%) by smoking cigarettes and is a pathologic diagnosis. Since lung biopsies for the diagnosis of emphysema have unwarranted risks, typically complete pulmonary function tests results are substituted for its diagnoses. These tests have a 97% correlation with the pathology.

Emphysema patients tend to be thin/asthenic and even cachectic. They are rarely cyanotic even with exacerbations; thus, the moniker "pink puffer

Airflow limitation is caused by dynamic expiratory airflow limitation. The increased expiratory intrathoracic pressures lead to dynamic expiratory airway collapse; thus, they "can't breathe out." Paradoxically, the harder they try to exhale, the worse the dynamic airway narrowing and the worse the expiratory airflow limitation. In contrast, the decreased inspiratory intrathoracic pressures dynamically open the airways; thus, they can "breathe in" even though hyperinflation does limit its degree. Since they can inhale without undue difficulty they tend to maintain near normal blood gases (PaO_2 and $PaCO_2$) until the disease is far advanced.

Complete PFTs reveal persistent airflow limitation, marked hyperinflation with increased residual volume ("air trapping"), and low diffusing capacity even when corrected to alveolar volume due to lung parenchymal destruction. These results are considered diagnostic.

Treatment is to decrease dynamic expiratory airway collapse through "pursed lip" breathing which is usually spontaneous and the use of CPAP/BiPAP to prop the airways open and decrease work of breathing. The management of dyspnea with opiates and anxiety with benzodiazapines are frequently beneficial. Since there is little to no bronchospasm or secretions and little airway edema, bronchodilators, steroids and antibiotics are of unclear benefits.

b. Chronic Bronchitis ("Blue Bloater")

Chronic Bronchitis is caused by smoking 60% of the time. Its diagnosis is based on a history of several months long episodes of recurrent cough and sputum for 2 or 3 years in a row.

These patients tend to be heavy set or stout. They are frequently cyanotic and have peripheral edema due Cor Pulmonale (pulmonary hypertension leading to right heart failure); thus the moniker "blue bloater".

Airflow limitation is due to fixed airway narrowing due to mucosal edema and hypertrophy and airway secretions. They are breathing "through a straw" and since the inspiratory muscles are weaker than the expiratory muscles, they cannnot do the work of breathing. Thus, they hypoventilate resulting in low blood gases PaO_2 and $PaCO_2$ (" 60/60 club")

Complete PFTs reveals persistent airflow limitation, minimal hyperinflation and air trapping (if any), normal diffusing capacity and high airway resistance.

Treatment is for the airway edema and secretions due to inflammation with corticosteroids and antibiotics. Bronchodilators may be of help esp. inhaled anticholinergics.

c. Asthmatic Bronchitis

Asthma has intermittent airflow limitation, i.e., the airflow limitation seen on spirometry returns to normal. Asthmatic Bronchitis has acute on persistent airflow limitation, i.e., the airflow limitation significantly improves but never returns to normal.

Asthma and Asthmatic Bronchitis are diagnosed by their pronounced and intermittent bronchospasm, i.e., wheezing. Frequently allergies and allergic sinusitis are present.

Airflow limitations is predominately due to bronchospasm, airway edema and secretions.

Complete PFTs reveals reversible airflow limitation in Asthma and partially reversible but with residual persistent airflow limitation in Asthmatic Bronchitis on spirometry, hyperinflation on lung volumes, normal diffusing capacity and high airway resistance.

Treatment is predominately for bronchospasm and airway edema and secretions caused by inflammation. Bronchodilators, steroids and antibiotics play key roles. Short-term subcutaneous epinephrine or terbutaline and medium-term oral beta-agonist and theophylline may have roles with severe bronchospasm exacerbations.

Also important, esp. for patient who have nasal allergies, is the early treatment and management of allergic rhinitis/sinusitis because these frequently lead to lung bronchospastic exacerbations.

Summary

The practice of medicine requires the diagnosis and management of the individual's disease or infirmity. Diagnosing a patient with a disease class such a COPD (or PALDs) does them limited good. The individual's specific type of PALDs needs to be delineated and treated for the maximal good.

Also, all research into PALDs must subgroup patients into specific diseases. Without this subgroup differentiations, clinicians do not know if their patient was in the subgroup that responded to the intervention.

APPENDIX: Miscellaneous PALDs [2]
1) Common: Bronchiectasis, Cystic Fibrosis
2) Uncommon:

a) Agenesis, Hypoplasia and Ectopia of the lungs
b) Variants of Pulmonary Emphysema:
    - Bullous Emphysema
    - Unilateral and Lobar Emphysema
    - Congenital Lobar Emphysema
    - Senile Emphysema
    - Paraseptal Emphysema
    - Compensatory Emphysema
c) Idiopathic Pulmonary Microlithiasis
d) Pulmonary Lymphangiomyomatosis
e) Congential Pulmonary Lymphangiectasis
f) Bronchopulmonary Dysplasia and Pulmonary Dysmaturity Syndrome
g) Congenital Cystic Adenamotoid Malformation

h) Pulmonary Blastomas and Hamartomas

i) Congential Anomalies of the Tracheobronchial Tree

- Agenesis of Trachea or Bronchi
- Constrictions of the Trachea and Bronchi
- Tracheobronchomalacia
- Compression due to Vascular Anomalies
- Tracheopathia Osteoplastica
- Tracheobronchomegtly
- Tracheal Diverticula
- Bronchogenic Cysts
- Tracheoesophageal and Bronchoesophageal Fistula
- Subnumerary and Supernumerary Bronchi, Lobes and Fissures
- Bronchopulmonary Sequestration
- Scimitar Syndrome

j) Miscellaneous Disorders of Airway

- Small Airways Disease
- Bronchiolectasis
- Bronchiolitis Obliterans
- Mucoid Impaction
- Broncholithiasis
- Hemoptysis
- Upper Airways Obstruction (Oropharynx, Larynx or Trachea)

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