

## How Rare Is Your Lung Disease?

By John R. Goodman BS RRT

How rare does a lung disease have to be so that it is actually called a “rare” lung disease? Well, let’s look at the lung diseases that most everyone has heard of first. Probably the most common lung disease today is the group of diseases we call Chronic Obstructive Pulmonary Disease or COPD. According to the most recent numbers released by the American Lung Association there are about 13.1 million Americans over the age of 18 with COPD. Broken down a little further there are some 9.9 million patients with a diagnosis of Chronic Bronchitis, and around 4.4 million patients with a diagnosis of Emphysema. The math doesn’t add up exactly because of the million or so patients who have both Chronic Bronchitis and Emphysema. These two numbers are always moving targets for statisticians. According to the latest Census Bureau estimate, the population of the United States is at 311,591,930....but 25% of that number is under the age of 18. That leaves a total of about 233,000,000 adults over the age of 18. So if we use round numbers we can see that about 13 million patients out of a total of 233,000,000 gives us a figure of about 5.5%. This is just about 40 COPD patients per 100,000 population. Don’t think anyone would argue that COPD is *not* a rare disease. But much is known about COPD as it has inched its way up the charts in both the number of deaths per year (over 125,000 and now ranked third), and the costs to our economy (49.9 billion dollars annually).



Emphysema Patient



Hyperinflated lungs of Emphysema

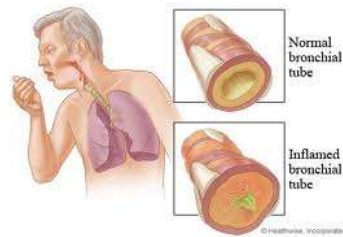
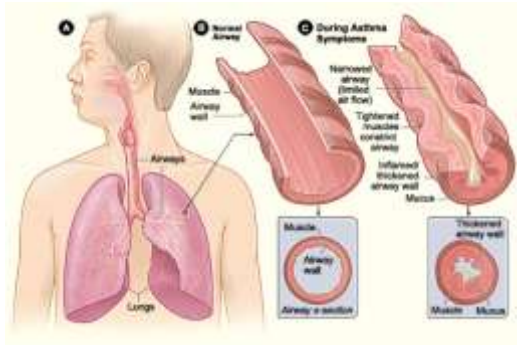


Diagram Chronic Bronchitis



Pt. with Chronic Bronchitis

Let’s take a look at some other well known diseases. How about Asthma? According to the CDC about 8.2% of adults over the age of 18 have been diagnosed with Asthma. Another 9.4 % of children under the age of 18 also have Asthma. So that means we have a total of a little over 26,000,000 Americans with Asthma. That number just about doubles the total number of patients with COPD. In fact, it wasn’t all that long ago that Asthma was lumped in to the diagnosis of



Asthma is a disease of the airways  
It is episodic and reversible



Asthma isn't funny  
at any age



Young kids learn  
inhaler technique early

COPD along with Chronic Bronchitis and Emphysema. Fortunately for both disease groups, Asthma was separated from the other two diseases because Asthma is episodic and reversible unlike Chronic Bronchitis and especially Emphysema. Asthmatic patients may represent 8-10% of the US population. So again Asthma indeed is *not* a rare respiratory disease.

Another disease that has been classified as obstructive disease in the past is Cystic Fibrosis (CF). The Cystic Fibrosis Association reports that there are somewhere around 30,000 patients in the United States with CF. Obviously this is a very, very small percentage of our total population (children and adults) roughly .00009%. Interestingly, over 10,000,000 Americans are carriers of the defective CF gene and are totally without symptoms. Around 1,000 new cases of CF are identified each year. There were about 4,000,000 babies born in the United States in 2012. CF shows up in about 1 in every 3,700 live births. CF *is* very definitely a rare disease by any criteria.



CF now diagnosed  
very early



chest vibration therapy  
to loosen secretions



inhaler therapy very  
important therapy



CF does not discriminate  
between boys and girls

To move away from the obstructive side of lung disease for a minute, let's see where patients with Interstitial Pulmonary Fibrosis (IPF) show up in our statistics. Is IPF a rare disease? Well, it is estimated that there are approximately 200,000 Americans affected by IPF. This works out to something like 3-6 cases of IPF per 100,000 population, or 0.0009% of the adult population. There are over 100 different causes or pathways that may lead to the development of IPF, and 40,000 IPF patients die every year. Although the numbers don't begin to approach those of the COPD group, Interstitial Pulmonary Fibrosis does not offer its patients the many options that are available for most COPD patients. This *is* a rare and very different disease that is finally getting the national attention it so richly deserves.

Another disease that is finally getting some attention primarily through the blogosphere and social networking is a terrible genetic disease called Lymphangiomyomatosis (LAM) It is pronounced lim-fan-gee-o-ly-o-my-o-ma-to-sis, and is normally abbreviated as LAM disease. Although first described by Von Stossel way back in 1937, it wasn't until 1995 that the LAM foundation was established to study and support research into this disease. LAM almost exclusively affects young women of child-bearing age. LAM is a systemic disease that results in cystic destruction of the lung. These cysts commonly rupture causing a pneumothorax (collapsed lungs). In fact, 6 out of 7 LAM patients can expect this to happen to them multiple times. For reasons still unknown, there is an abnormal growth of smooth muscle cells that invade healthy lung tissue, including the airways, blood vessels, and lymph vessels. Although technically this isn't cancer, the cells do grow uncontrollably. Ultimately there is destruction of normal lung tissue and the ability to transfer oxygen into the blood is significantly impaired. There is no known cure for LAM, but there have been some recent successful medical treatments under study. The average age at the time of diagnosis is 34, with 70% of those women diagnosed between the ages of 20 and 40. Unbelievably, there have even been a few men (less than 10) diagnosed with LAM. At some point virtually all LAM patients will require supplemental oxygen, and most will have to consider lung transplantation as LAM is universally relentless, progressive and fatal.

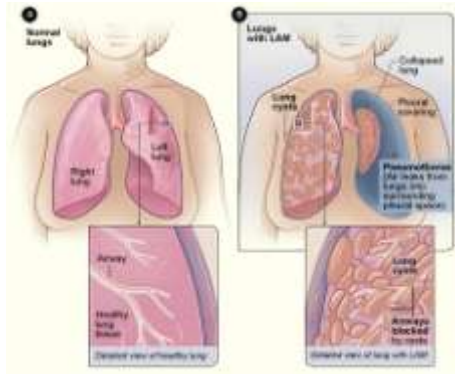
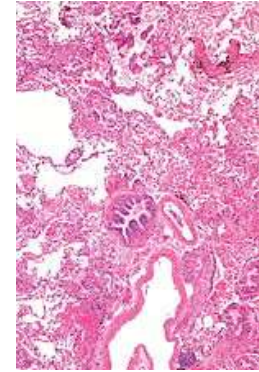


Diagram of LAM disease



LAM chest x-ray showing damage



Nowhere for oxygen to go

So how rare is LAM disease? The latest estimate on the number of LAM patients in the United States is right at about 675 patients. There are probably hundreds more but part of the problem in getting accurate numbers is due to the fact that the typical LAM patient spends 3-5 years seeking a diagnosis *before* finally getting to the right institution or physician to actually get the LAM diagnosis. This means that a LAM patient represents just 0.000003% of the adult population. LAM patients with this incredibly rare lung disease affectionately call themselves “Lammies.”

Well if you think there couldn't possible be any lung disease rarer than LAM disease...well you'd be wrong. A “new” lung disease has (pardon the pun) popped on to the respiratory scene over just the past 10 years or so. It is called Popcorn Lung and it is no joke. It is more medically classified as Bronchiolitis Obliterans (BO), or sometimes Constrictive Bronchiolitis. This is a rare obstructive lung disease where scar tissue gradually obstructs the bronchioles of the lung. Anatomically we are talking about airways around 1mm in diameter and smaller.

BO is not a disease to fool around with. In advanced stages it becomes irreversible and may only be treated by lung transplantation. It is pretty clear at this point that the chemical *diacetyl* is the agent that causes the problem. It is used in a number of industries but especially in the buttered popcorn industry. Diacetyl is the chemical that gives popcorn the natural buttery smell and taste that so many people love.

The first time diacetyl came under scrutiny was back in 1985, but it wasn't until the year 2000 that a significant outbreak at the popcorn factory in Jasper Missouri was identified. Eight workers at the Glister-Mary Lee plant developed Bronchiolitis Obliterans. This led to some super detective work by a number of

agencies that determined that those workers who spent more time in the areas where the diacetyl was sprayed on to the popcorn were 2.6 times more likely to report cough and shortness of breath than other non-exposed workers. Over the next several years enough research proved that exposure to diacetyl could indeed cause BO in patients with sufficient exposure.

Consumers have been repeatedly reassured by the Flavor and Extract Manufacturers Association that there is virtually no risk of developing Popcorn Lung from eating buttered popcorn. And this would seem to be the case as the average per capita consumption of buttered popcorn in the United States is currently 73 quarts per year! Even more if you subtract all the babies who can't chew food yet and those who just don't like cleaning popcorn bits from their teeth.

And yet.....on September 20, 2012 a Colorado jury awarded Wayne Watson 7.2 million dollars in a lawsuit filed against both Glister-Mary Lee Corp. and Kroger foods. His claim was that he first noticed breathing problems during his church choir practice that he just couldn't hold some notes like he used to. Some might consider Mr. Watson as uncommonly devoted to microwave popcorn. He ate two bags a day for 10 years and said he regularly sniffed up the puff of delicious odor of a newly opened bag of popcorn. His argument was there should have been warning labels on the popcorn bags, much like we already have on cigarette packages. The ruling is of course under appeal. It is estimated that there are about 200 people in the United States with actually diagnosed Popcorn lung. Now *that* is pretty rare. On the order of 0.0000008% of the adult population.



Popcorn lung isn't funny!



Pt. with Popcorn Lung



May be 7.2 million \$ ahead

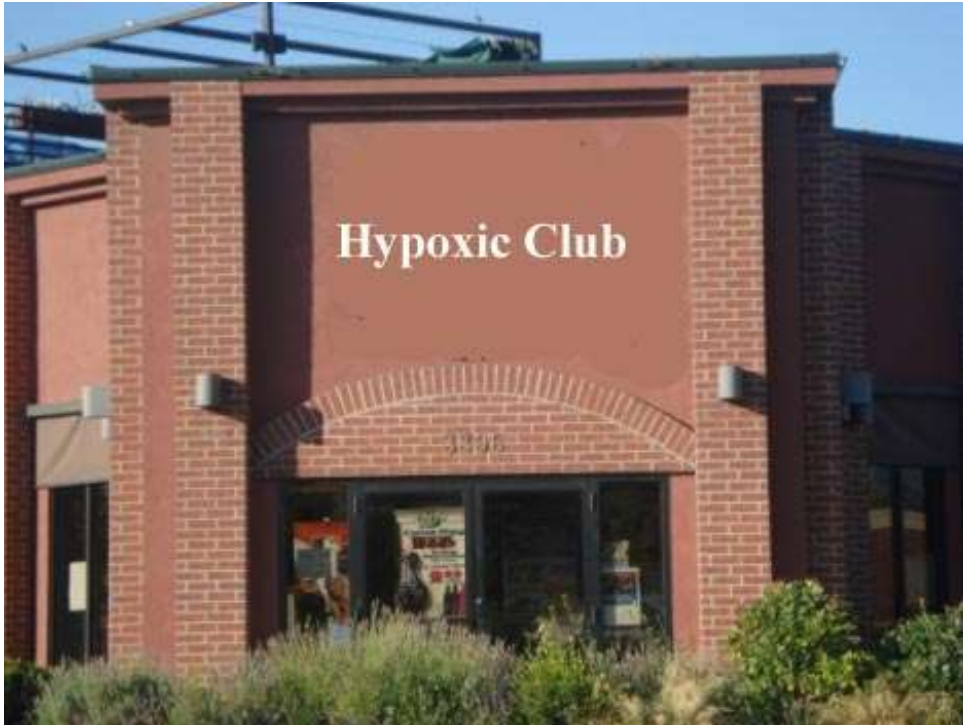


How far do we take it?

Is there one thing that may be found in common with each of these very different diseases? Is there one drug common in the treatment of each of these diseases? Of course there is, and that drug is oxygen. As each of these diseases progresses nearly every one of these patients will require supplemental oxygen (excluding the vast majority of asthmatic patients). They are all (unwilling) members of the "Hypoxic Club." No one really wants to be in the Hypoxic club, because once you are a member, it is usually a lifetime membership. The Hypoxic Club is not



selective. It doesn't really care who you are, what you do, or what your plans are. The only rule is your blood oxygen level on room air is less than normal for where you live. So no matter how common or rare your lung disease is, all members strictly adhere to the club motto, which is "Let your oximeter be your guide."



Welcome to the Hypoxic Club...Remember to titrate when you migrate