


BOOP: YOU'RE THE BOSS



YOU CAN MANAGE IT

**Five Steps To Take
Charge of Your BOOP**



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Diagnosis: A Series of Questions and Tests to Lead You There

After meeting your physician, you will be asked about the beginning of your illness. How did it start? What was the sequence of symptoms? Were any events associated with the onset? You also will be asked the five lung-symptom questions about cough, phlegm, wheezing, shortness of breath, and coughing up blood.

Is there a cough? If so, when did it begin? Does it occur every day? Does it occur at night? Cough means irritation of the airways; there could also be airway inflammation or abnormalities in the lungs. Bronchitis from a viral or bacterial infection is the most common diagnosis because the inflammation is in the moderate- and large-sized bronchial airways. The cough might also be due to inflammation of the small bronchiole airways. Other causes of airway inflammation can include postnasal drip, stomach acid from esophageal reflux that irritates the airways, and tumors.

You will also be asked about phlegm or sputum production. Is there phlegm with the cough? If so, when does it occur – in the morning, throughout the day, or in the evening? How much – a teaspoon or a half

cup? Millions of glands in the airways produce mucus and defensive substances to keep the airways moist and protect against disease. There is ongoing production of mucus at an undetectable level. Inflammation, however, will cause an increase in mucus and phlegm. If an infection is causing the inflammation, the mucus will be yellow and green in color. Antibiotics might be needed.

Is there wheezing, which is a whistling sound during an expiratory breath? If so, does it occur in the morning or at night? Before or after the cough? The airways are surrounded by millions of circular, smooth muscle fibers, giving them shape and preventing them from collapsing. Irritating substances or inflammation will cause these smooth muscles to constrict and narrow the airways, which is referred to as bronchospasm. These narrow airways create a high-pitched sound, or wheezing, when you exhale. In severe situations, wheezing occurs when you inhale. Wheezing usually means asthma, which is inflammation that occurs internally within the airways. Other causes of bronchospasm and wheezing include inflammation from an infection, as in bronchitis or an allergy. In rare instances, a tumor obstructing the airways will cause wheezing.

Is there shortness of breath or breathlessness with certain activities? If so, what is its severity, on a scale of one to four, with grade one being shortness of breath with exertional activities; grade two, while climbing a flight of stairs; grade three, while walking on level ground for less than 100 yards; and grade four, while performing routine activities such as talking or dressing? Shortness of breath might mean inflammation of the lungs. The cause of the inflammation may be an infection, as with pneumonia, or unknown, as in BOOP. It may be also be due to a weakened heart or weakened muscles. With shortness of breath, more than two-thirds of the lung is involved in the abnormal process.

Has there ever been blood in the sputum? This is caused by severe inflammation of the airways or lungs and may be due to infection or tumor. Sometimes a blood clot in the lungs in the form of a pulmonary embolism can cause blood in the sputum.

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Other questions include whether a flu-like illness began the respiratory process and whether there has been chest pain.

Next, you will be asked about smoking, your family's medical history, and your own past respiratory and other medical illnesses as well as past surgeries. For smokers, when did you start? How many packs per day? Did you stop, and if so, when?

Are there any family members with asthma, allergies, recurrent pneumonia, tuberculosis, lung cancer, emphysema, chronic obstructive pulmonary disease, or COPD, sarcoidosis, or pulmonary fibrosis? Have you had any respiratory illnesses in the past, such as asthma, pneumonia, bronchitis, chest colds, sinus infections, or postnasal drip? Are you allergic to any foods, medications, animal dander, or pollen? Do you have hay fever?

There are several nonpulmonary questions having to do with diabetes, high blood pressure, peptic ulcer, gastroesophageal reflux disease, or GERD, disease of the heart, kidney, or liver, rheumatological or immunological disorder, seizures and other neurological condition, and cancer.

What are the names and dosages of the medications that are you taking? When did you begin taking them? Have you had radiation therapy? Answers to questions about medications such as amiodarone for the heart or chemotherapy for cancer, and about past radiation therapy, will help to determine the cause or type of BOOP.

Finally, you will be asked about occupational or environmental exposures. A helpful way to answer the questions is to begin with your place and date of birth; what you did during the summers; if you had any exposures during school or at home; and what your first job involved, including title and description. The questions will continue with a chronological review of your jobs and potential specific exposures, such as to accidental fumes, ongoing fumes or chemical releases, and toxic dusts like asbestos, beryllium, or silica. If you had an exposure, what was the amount? When did it occur? What about your office environment? Have you had exposures to birds such as pigeons? Do you breed birds as

a hobby? Answers to these questions can identify an environmental or occupationally-related BOOP.

Now that you have answered the questions, it's helpful to summarize the course of the illness and to add any information that might aid in making the diagnosis of BOOP.

Next, your vital signs will be obtained, including blood pressure, pulse, temperature, respiratory rate, and oxygen saturation level. The latter test measures the percentage of oxygen contained in the blood hemoglobin. Values over 95% are usually normal, while those of less than 85% are usually abnormal. In moderate-to-severe BOOP, oxygen saturation may be near normal while you're resting, but it can decrease to less than 85% during a walk down the hall or a climb up a flight of stairs.

The doctor will examine your fingernails for clubbing – a curvature of the fingernails resembling a parrot's beak – or for a bluish color, which may mean low oxygen.

Your ears, eyes, and throat will be inspected for any abnormalities. Your neck and the top of your shoulders will be felt to determine if any lymph nodes are enlarged. The back of your chest may be thumped to search for fluid in your lungs.

The doctor will use a stethoscope to listen for any abnormal sounds. With BOOP, crackles will usually be heard. Crackles are sounds that the lungs make due to inflammation surrounding the airways. As the airways open during inspiration, they create a popping noise. There are usually no wheezes or squeaks.

The remainder of the physical examination usually yields normal findings unless there is a rheumatological cause, in which case there would be abnormalities of the joints.

Laboratory studies are often normal. During the flu-like-illness phase, blood studies may show increased white blood cells and inflammation. There may be an increase in the sedimentation rate, or C-reactive protein. These tests have been available for many years and are nonspecific

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measures of inflammatory reactions. There are no specific blood tests for BOOP.

Radiographic studies are helpful in the diagnosis. The chest x-ray shows patchy infiltrates in both lungs. Cavities in the patchy shadows sometimes occur but are rare. Fluid around the lungs is also rare.

The high-resolution chest CT scan shows the densities as ground-glass opacities. Ground glass is a term used by radiologists to describe a hazy, homogenous shadow in the lungs. Ground-glass opacities are important findings because they reflect inflammation instead of scarring. Inflammation can be treated successfully with anti-inflammatory medicine, corticosteroids, or prednisone. These findings are common in BOOP because it is inflammation of the lungs. Ground-glass opacities in the shape of triangles, with their bases on the surface of the chest wall and their apex tips toward the center of the chest, are almost always diagnostic of BOOP.

Single round nodules or multiple nodules represent a special form of the disease, called focal nodular BOOP. Sometimes the patchy nodules are fleeting, occurring in one section of the lung for one or two weeks and then in another section. Rarely, there may be tiny nodules in both lungs. For these unusual appearances, a lung biopsy is often needed to establish the diagnosis.

The diagnostic process also consists of three pulmonary function tests, or PFTs. The forced vital capacity test, or FVC, measures the amount of air in the lungs. The one-second forced expired volume test, or FEV₁, measures the amount of air expired in a second. The diffusing capacity test measures the ability of the lungs to exchange oxygen from air into blood circulating in the lungs.

These tests are designated as percent predicted, which means that your results are compared with those of hundreds of people who do not have BOOP. Values above 80% predicted are normal. A mild decrease is from 60% to 80% predicted and a moderate decrease, from 50% to 79%

predicted for vital capacity and from 40% to 79% for diffusing capacity. Values below 40% predicted indicate severe lung impairment.

For BOOP, the vital capacity is usually mildly decreased. The diffusing capacity is moderately decreased because inflammation in the spherical alveoli gas-exchange units has blocked and slowed the exchange of oxygen from air to blood. The moderate- and large-sized airways referred to as bronchi are normal in BOOP; therefore, the FEV₁ test is normal and there is no airflow obstruction. There may be airflow obstruction, however, in smokers. With mild decreases in the vital or diffusing capacity, there may not be any symptoms or there could be mild shortness of breath. With moderate or severe decreases, shortness of breath may occur during exertional activities.

Sometimes the diagnosis of BOOP can be established on the basis of the above information, and a management plan can be developed for either a brief monitoring period without treatment or a course of prednisone therapy. But because of the many lung diseases that can mimic BOOP and the different types and durations of treatment, a sample of lung tissue is usually needed to confirm the diagnosis. Tissue can be obtained through a bronchoscopy, which involves the insertion of a tube through the nose into the lungs and the performing of transbronchial biopsies. The procedure does not result in a scar and sometimes yields the diagnosis; however, the samples are often too small to rule out other possible diagnoses. In addition, there are the risks of bleeding and collapse of the lung, and the latter may require insertion of a tube into the chest. The low diagnostic yield must be weighed against the risk.

Tissue also can be obtained from a video-assisted thoracoscopy, or VAT, procedure. This is a commonly used method because it provides an answer in almost all situations and the risks associated with anesthesia and surgery are better controlled. This biopsy procedure is performed in a hospital setting. A small tube in the chest is needed for 12 to 24 hours afterward.

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Several years ago, I gave a career-day seminar at a high school on the discovery of BOOP. A few days later, one of the students, Steve, heard his mother talking about their friend who was in the intensive care unit with pneumonia that hadn't responded to antibiotics and was rapidly deteriorating. The student interrupted and exclaimed, "It's probably BOOP!"

His mother was totally dumbfounded but astute enough to talk to the doctors. Their friend was transferred to another facility, diagnosed with BOOP and successfully treated, and is doing well to this day.

Understand the diagnostic process – you never know where you might find helpful information.

The diagnosis is established by symptoms, lung crackles, low vital capacity and diffusing capacity, and a chest x-ray showing patchy infiltrates. As the diagnostic process continues, it's important to understand each step and to ask questions about the benefits of testing, especially the probability of establishing the diagnosis with any given procedure. At the same time, ask about the risks of each diagnostic procedure. When you have these answers, you'll be able to weigh the benefits against the risks and to make a decision that is best for your situation.

Let's follow Quinn Stone during his BOOP-management journey. He was celebrating his 46th birthday and had no difficulty blowing out the five candles that his wife, Jean, had added to the "Happy Birthday, Dad!" cake that she and their children had made.

During the next several days, Quinn developed aches and pains similar to the flu, but he ignored them because he was working on a deadline at the office. The flu-like illness persisted, and he developed a cough. He thought it was part of the flu and continued working.

"I have a Friday deadline," he told himself as he tried to ignore the symptoms.

"You look like you're out of breath," Jean said to her husband as he entered the house after shoveling snow.

"Of course, I've been shoveling a ton of snow," Quinn remarked casually. But during the day, he noticed shortness of breath while climbing the stairs, and he still had that nagging cough.

"Maybe I should go to the doctor," he said to Jean.

"That's hard for you to admit, but you seem to be worse, so it sounds like a good idea to me," Jean said, hoping for a positive response.

"What brings you here today?" asked Tara Spencer, Quinn's family doctor, as they sat in her suburban office.

"I have the flu and a lingering cough," he said.

"I hear crackles in your lungs," the doctor told Quinn as she leaned over his right shoulder to listen to his lungs.

"What does that mean?"

"You probably have pneumonia from the flu," Dr. Spencer said. "I think you should have a chest x-ray."

The radiograph confirmed the diagnosis, as it showed patchy infiltrates in both lungs. "Let's treat this with an antibiotic, and I'll see you next week," the doctor said.

Quinn returned to work. He wondered how he had developed pneumonia, but he knew that it could happen after the flu. After several days, however, he was puzzled by the ongoing symptoms, as he expected to be recovering.

His suspicions were confirmed when his follow-up visit to the doctor showed no change in the crackles. The chest x-ray was the same, except it now showed a new small ground-glass infiltrate in the middle of the right lung.

Dr. Spencer was also puzzled. "Let's try one of the new antibiotics," she said.

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Quinn returned home, somewhat skeptical about his prognosis, and began to think more frequently about his symptoms. After two more days without improvement, he called Dr. Spencer with his suspicion that the new antibiotic was not working.

“Let’s send you to a lung specialist,” Dr. Spencer answered, believing that Quinn’s illness was probably not typical community-acquired pneumonia.

Quinn’s friend at the office had told him that Dr. Erica Sinclair was a very special person. She was a specialist and had helped a friend cure a rare form of cancer.

“So you have BOOP?” Dr. Sinclair casually asked Quinn as they sat in her New York City office.

“What?” Quinn said, surprised at her question. “What’s she talking about?” he asked himself. He had never heard of BOOP. It was a funny name. Rapid-fire questions flew through his mind. “Is it an evil disease? What’s going to happen to me? How does she know I have BOOP?”

“I was treated for pneumonia,” Quinn told the doctor. “What’s BOOP?”

“We’ll get to that soon enough,” Dr. Sinclair said. “When did you first develop symptoms?”

“About six weeks ago,” Quinn explained. “I had a flu-like illness for a few days, then a cough, and recently I became short of breath while climbing stairs.”

“You had no wheezing, no coughing up of blood, and you never smoked,” she said rhetorically.

“That’s right, and I never had any breathing problems,” Quinn said. “I have no allergies or medical illnesses, and I don’t take any medications. I’ve never been exposed to fumes or toxic chemicals.”

“I hear crackles on both sides of your lungs,” Dr. Sinclair told him as she listened to his lungs with her stethoscope. “Let’s look at your CT scans because your shortness of breath probably means there is inflammation in the lungs.” Together, they looked at the computer screen displaying the scans.

“This is BOOP,” Dr. Sinclair said as she showed Quinn the patchy infiltrates with the air-filled airways. “These triangular-shaped, ground-glass shadows almost clinch the diagnosis.”

The doctor sent Quinn to the building’s laboratory, where a full set of pulmonary function tests was obtained. The vital capacity test was performed by having him take a huge deep breath and blow it out as fast and as far as he could while the respiratory technologist encouraged him with loud and determined commands. The vital capacity was slightly more than three liters, which was 68% predicted. Normal values are above 80% predicted, so this signaled mild impairment.

As expected, there was no obstruction of the airflow as seen in asthma or emphysema, especially since he had been easily able to blow out the candles on his birthday cake. This was confirmed by the one-second forced expired volume test, or FEV₁. The value was slightly decreased because the vital capacity was decreased. The FEV₁ to forced vital capacity ratio, or FEV₁/FVC, which is the most sensitive test to show airflow obstruction, was slightly above normal, and it didn’t show obstruction of airflow as seen in asthma or emphysema.

He also had the diffusing capacity test, which is more complex, but useful for finding out how well oxygen is being exchanged between the lungs and the bloodstream. Again, Quinn took a deep breath as he held a tube in his mouth, held it for ten seconds, and blew out as fast as he could. The computerized machine captured the last of the expired air from deep in his lungs, and analyzed it to determine the diffusing capacity. This result shows the ability of the lungs to exchange oxygen from the air to the blood.

Normally, there is one thin cell between the lungs and the blood that allows instant oxygen exchange and a normal diffusing capacity. BOOP is inflammation of the spherical alveoli in the lungs, so these structures are filled with inflammation that does not allow instant oxygen exchange and the diffusing capacity is decreased. There are millions of these spherical structures, and more than one-third of them must be involved for the

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diffusing capacity to be low. If more than two-thirds are involved, shortness of breath will develop, and if more than four-fifths are involved, the oxygen will be so low that admission to the intensive care unit, or ICU, and mechanical ventilation may be required.

Quinn's test showed 14.5 milliliters per minute, which was 54% of the predicted level, or moderate lung dysfunction. The diffusion was sufficiently decreased to cause shortness of breath because the muscle requirement for oxygen was greater than the availability of oxygen in the blood after it had passed through the lungs.

"What's next, Dr. Sinclair?" Quinn asked anxiously as he waited for some good news and the relief of his symptoms.

Quinn had gone through the diagnostic process. He had understood the testing and the reason for all of it. We'll find out about his treatment in the next chapter.

BOOP: You're the Boss

This book tells you how to manage your BOOP. You'll learn the five vital steps to take charge.

- Learn everything you can about BOOP.
- Understand how the diagnosis is established.
- Know your treatment options.
- Monitor your BOOP.
- Create a healing environment.

You can manage your BOOP better than anyone else can. You just need to know how.



The author, **Gary R. Epler, M.D.**, has written the critically acclaimed personalized health book, *You're the Boss: Manage Your Disease*. He believes personalized health allows people to manage their health and disease. Dr. Epler is world-renowned for describing the lung disorder bronchiolitis obliterans organizing pneumonia (BOOP), which spurred international research and study. He discovered a parasite in South America, chronicled the nutritional needs of North African children, and managed the tuberculosis refugee program in Southeast Asia. He is a frequent guest on radio and television. Active in his community, Dr. Epler has coached soccer, hockey, basketball, and baseball. He lives in the Boston area with his wife Joan and his two sons.

Visit Epler Health at www.eplerhealth.com for personalized health information and learn how people can manage their health and disease. Learn as much as you can. Become your own boss and take charge of your BOOP.

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