

DOCTOR FILES

When coughing is too distasteful

A lifetime of suppression leads to infection -- and a very unladylike treatment for Lady Windermere Syndrome.

By Francis V. Adams, Special to The L.A. Times
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Coughing was a no-no for a proper lady. (Columbia Pictures)

I saw another Lady Windermere the other day. Over the years I have seen several patients who could have borne this name. The character originated in Oscar Wilde's play "Lady Windermere's Fan." She was a fastidious woman who at one moment refuses to shake hands with a visitor ("My hands are all wet with the roses"). Wilde's character would become a symbol of the Victorian era, an age when women wouldn't do anything they thought vulgar, such as spitting.

My first Lady Windermere was Agatha. I met her not long after I finished my training in pulmonary disease and opened my private practice. I glimpsed her in the waiting room as I picked up her chart. A few minutes later, as she sat in my office, I inquired as to what had brought her, and she went on to describe an unrelenting cough she'd had for nearly 10 years. During the interview, she coughed fitfully but would not expectorate.

Agatha was 63, very thin, almost skeletal, with high cheek bones, thin lips and a straight nose. Her forehead was made more prominent by her graying hair, which was pulled straight back. She held a tiny lace handkerchief in her left hand and covered her mouth as she coughed.

I proceeded to take Agatha's medical history. She had been in good health except for her chest problems, which she described as frequent colds that always settled in her chest. Agatha had been hospitalized twice for pneumonia. She was an actress and had done stage and film, mostly small parts as she described them. Agatha's physical examination did not add much to what I had already observed. Her lung sounds were a bit quieter than normal but I did not detect any congestion. I proceeded to take an X-ray, which showed that the air passages in the middle sections were thicker than normal.

We sat in my office and I told her that it would help if she could bring up some sputum for the lab to analyze for infection. I also explained that additional X-rays would be helpful.

Agatha said that she was used to having X-rays but doubted if she would be able to produce a sample of her sputum. Just then she coughed again, and I noted that she seemed to be trying to suppress her effort.

I told her that I had a few tricks for getting people to cough up. I took Agatha into another room and introduced her to a nebulizer, a simple machine that creates an aerosol mist by forcing air through a solution. I placed saline into the machine, attached some tubing, turned on the power and saw a steam-like vapor emitted.

I asked her to breathe the mist for 10 minutes and placed a sputum cup on the counter next to the nebulizer. When I returned to the room, I saw that the cup was still empty. I placed more saline into the machine and asked her to try again. After the second treatment, Agatha coughed and produced a tiny bit of yellow sputum. The sputum sample was sent off to the lab and her X-rays

scheduled. I explained that I hoped to have the results of both tests in a few days.

Agatha's X-rays showed that there was evidence of old and new infection in the middle sections of both lungs. The bronchial tubes in both areas had been damaged causing them to dilate and become congested with mucous, a condition known as bronchiectasis. Bronchiectasis is usually produced by an untreated lung infection. In many of my elderly patients, the infection had occurred in childhood when antibiotics were not yet available.

The culture of the tiny piece of expectorated sputum yielded an organism known as mycobacterium avium intracellulare. This is a ubiquitous germ that lives in nature and can be found in the soil or water. At this point in my practice, I had seen this infection only in immune-compromised individuals but knew it could occur in anyone with damaged lungs. Agatha had no history of childhood infection so I wondered if the over-fastidiousness that kept her from clearing secretions had in fact promoted the development of her condition. I proceeded to outline a course of treatment that would include three antibiotics over a period of one-and-a-half years. I also placed her on an expectorant and arranged for a physical therapist to cup and clap her chest twice a week, hoping to help clear her air passages. Despite these efforts, my patient's cough did not produce sputum.

During the long course of Agatha's treatment, I saw two more women who bore not only a physical resemblance to her but also the identical illness. Irene was 68, a teacher with a widow's peak, and Constance was 60, a librarian. Both had similar X-ray changes as Agatha, and their sputum, which I obtained with great difficulty, also yielded the same organism. All three women were cooperative, intelligent, and easy to work with, but I became increasingly frustrated by their failure to clear their lungs despite the many maneuvers that I put them through.

After my third case, I consulted my colleagues and the medical literature and found that I was not alone. Other doctors were seeing similar patients. In 1992, 15 years after I first met Agatha, two radiologists published a report of "The Lady Windermere Syndrome." They had observed six women with the same characteristics as my patients. The authors noted that the middle portions of the lungs extend outward toward the front of the chest and require vigorous coughing for clearance of secretions. They concluded that mycobacterial infection had occurred in these overly fastidious women due to voluntary suppression of cough.

In the last several years, greater numbers of cases of mycobacterial infection have been reported. Unfortunately the treatment is not always successful and may be difficult to tolerate due to adverse effects of the antibiotics.

Agatha's infection was not cured by years of treatment but did improve. I continue to see a few women each year with the same striking features and pride myself on making the correct diagnosis simply from observing their appearance and hearing their cough before they are seated in my office.

A few of these delicate women have found me through Internet searches so that after introducing myself to one of these ladies recently, she replied: "And you may call me Lady Windermere."

Dr. Francis V. Adams is a pulmonologist in New York City and the author of "The Asthma Sourcebook" and "Healing Through Empathy."