

Even a Physician With Pulmonary Hypertension Can Be Misdiagnosed

By Lynn Brown, MD, Campaign Chair

Diagnosing pulmonary hypertension (PH) is often so tricky that even a patient practicing pulmonology can experience delayed diagnosis. That's what happened to Bonnie Hudak, MD, a new member of the Sometimes it's PH early diagnosis campaign's Education Committee.

Dr Hudak is a pediatric pulmonologist at Nemours Children's Clinic in Jacksonville, Florida, where she often treats asthma and cystic fibrosis. Yet her path to diagnosis parallels that of many other PH patients, particularly middle-age women.

Dr Hudak had long been treated for scleroderma and Reynaud's disease. Her rheumatologist knew of the association between PH and scleroderma. Dr Hudak maintained a healthy weight, exercising regularly while practicing medicine and raising children. In her 40s, exercising became more difficult, but with her busy life she says she paid this little attention. Then while hiking in 2004, Dr Hudak discovered that at altitude she could not walk uphill.

In Jacksonville she underwent an echocardiogram, an EKG, and a chest x-ray. Her doctor called the results "maybe slightly abnormal." He was reassured and attributed her symptoms to perimenopause and deconditioning. He reported that the cardiologist had considered her echocardiogram normal. "They were happy with normal, and I was, too," Dr Hudak said.

Still, Saturday morning tennis games left her tired all weekend. Once, at a neighborhood party, she was chatting with a cardiologist friend. He told her firmly, "Anyone with scleroderma and shortness of breath with exercise has PH unless proven otherwise." Two weeks later she was diagnosed by right heart catheterization and referred to a PH specialty

Dr Hudak's experience at Mayo Clinic in Jacksonville under the care of Charles Burger, MD, highlights the importance of referral to specialty centers, a key element of the Sometimes it's PH campaign. In a single day she received comprehensive testing, including a more detailed echocardiogram, which successfully measured tricuspid regurgitation velocity. Those administering these tests pursued results doggedly.

Dr Burger also admitted Dr Hudak to the hospital for a right heart catheterization that included a vasodilator challenge. Without that thorough procedure and all of the necessary testing, Dr Hudak's vasoreactive type of PH would not have been discovered. Dr Hudak has remained on nifedipine as her sole PH treatment and has improved from Class III to Class I. She has also participated in a clinical trial.

In her practice Dr Hudak now looks for a few more zebras among the horses. She also looks more carefully at the data used to interpret studies. She would advise other physicians to be more vigilant with a patient who has an underlying condition associated with PH and to work up minimal symptoms that may be due to PH. She also suggests further evaluation if existing results don't make sense in the clinical setting.

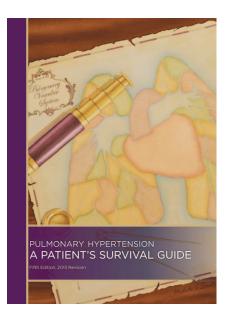
Dr Hudak's experience illustrates that both patients and professionals must be more active in questioning the data and the decisions that drive diagnosis. Her unique insights will be an asset as PHA works to enhance primary and specialty care professionals' ability to diagnose and treat PH promptly and correctly.

To find out more about Sometimes it's PH, visit www.SometimesItsPH.org.

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- Bruce Brundage, MD, former Chair, PHA Scientific Leadership Council

The fifth edition, 2013 revision of Pulmonary Hypertension: A Patient's Survival Guide is now available. Eight of the 17 chapters, plus the glossary and appendices, were updated for this latest version of the 300-page book, covering survival outcomes, insurance coverage, new resources, as well as conventional, drug, and surgical treatments. For the first time, PHA is offering this valuable resource as both a paperback and an e-book.

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