



Larry Hoffman

Alpha-1 Antitrypsin Deficiency

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When I was born with jaundice, they did not know how to treat it properly. Five others in my pediatric unit died, but somehow I pulled through. I now know that other alpha-1 antitrypsin patients were born with liver issues as well. All that was 62 years ago, and since then, I have been affected by breathlessness in many ways.

In my younger days, I was last in running races, wrestling, and football. I was told it was because I was out of shape or too chubby. I would be constantly out of breath whenever I had to do something physical. I developed a way to hide these breathless moments by yawning.

I married my wife 38 years ago, and we had a son and daughter. During these years I had two careers. I spent 17 years driving a semi-trailer, delivering and picking up construction materials, including coal tar pitch, fiberglass, and hot asphalt. I was also exposed to constant diesel fumes. Then I was self employed as a remodeling contractor, which exposed me to lead paints, old plaster dust, roofing materials, drywall, and fiberglass. I wore protection over the years, which was to some benefit, but I was still exposed to many hazards, and I even smoked cigarettes for a period of time.

That exposure likely contributed to my shortness of breath and diminished lung capacity. Some days I found myself gasping for air. Sometimes, I felt like I was breathing through a cocktail straw. Had I known about Alpha 1 early on, I would have made some lifestyle changes.

Once I hit 50, things got worse. Daily activities began to get harder to accomplish with winter bronchitis bouts. I was always running out of energy and getting short of breath. In the winter of 2009, I was on a hunting trip in Nebraska and ended up so

ALPHA-1 ANTITRYPSIN DEFICIENCY

Alpha-1 antitrypsin deficiency is an inherited form of emphysema. Patients with the disease do not have enough of a protein called alpha-1 antitrypsin (AAT) in their blood. This protein is made in the liver, and it protects the lungs so they can work normally. Without enough AAT, the lungs can become damaged by emphysema. It also can cause liver damage. There is no cure for Alpha-1 antitrypsin deficiency.

Testing is recommended for those who have:

- Obstructive lung disease.
- Family history of alpha-1.
- Early-onset emphysema (younger than 45 years old).
- Emphysema without an obvious risk factor.
- Emphysema that is worse at the bottom of the lungs.
- Chronic asthma (in adolescents and adults).
- Recurrent pneumonia or bronchitis.
- Unexplained liver disease.

Learn more: ATS Patient Education Series. "What is Alpha-1 Antitrypsin Deficiency?" New York, NY. 2006. patients.thoracic.org

sick and short of breath that I had to be assisted in walking. A respiratory specialist diagnosed me with alpha-1 at 58 years old. I had the normal “why me” thoughts. But I look back at that now as a blessing since I finally knew what was causing these symptoms.

Winters have become harder, as I’m becoming more affected by the cold air. I must keep my face covered outside when the temperature is low. I also have to bring up phlegm in the morning along with a deafening cough which is not a favorite household sound. I can pretty much walk comfortably on flat ground. But climbing stairs, hills, and quick starts are hard. I’m on disability, and I felt like I didn’t deserve the help in the beginning. But I can no longer do a lot of the remodeling tasks.

At certain times, I need my wife to help me do things I never would have asked years ago. At first, this gave me thoughts of being less, but I know that she is the best caregiver for me. My family is also aware of the extra help I need, and we work together. I am learning to accept my limits.

Larry Hoffman was a patient speaker at the ATS 2013 International Conference in Philadelphia.