Living with pulmonary fibrosis

Pulmonary fibrosis, which means scarring of lung tissue, is a serious lung disease, making it more and more difficult for the lungs to deliver oxygen to the blood. Patients become short of breath, experience fatigue and have much less energy than they used to. Pulmonary fibrosis can be caused by inhaling various noxious substances, using certain medications and by radiation therapy. Certain familial forms of pulmonary fibrosis have been described, but in many cases the cause remains unclear. It is usually associated with a poor prognosis.

BY: JENNY PENDERS

My mother also had pulmonary fibrosis. She died when she was 50 and I was 23. For many years I had no physical symptoms at all. I had no serious problems until the age of 28, but then I developed back pain. A chest X-ray revealed an abnormality and the doctor decided to operate to see what was the matter. It turned out to be a benign congenital problem, which was removed. At the check-up after the operation, however, my lung capacity proved to be considerably smaller than had been thought. X-rays were made, and I had to come in for regular check-ups. I had lung capacity tests and another X-ray every six months. Life went on and I felt healthy. I had two beautiful children and a busy job.

I slowly started to deteriorate, so slowly that at first I hardly noticed. It became ever more difficult for me to run, I couldn't cycle as fast as I used to, I got out of breath going up the stairs. But you learn to cope with your limitations and life keeps you occupied. I had a lung biopsy, but that didn't show any abnormalities. Years later, after I had suffered a pulmonary embolism, I came into contact with Professor Marjolein Drent, and the tissue was examined again. By then, diagnostic techniques had been further refined, and the diagnosis of pulmonary fibrosis was made. I finally knew where I was at.

At first you don't really realise what it means, and there wasn't much information available either. I tried medication, prednisone and imuran, but they didn't work. Around the same time I got a new job, and I didn't know what information I should give to my new employer; I knew so little myself and the course of the disease was unpredictable. My physical condition gradually deteriorated, I needed special measures like oxygen, a disabled parking permit and a stairlift at home.

It was then that I met Marlies Lahaut at a physiotherapy session for patients like us. She also had pulmonary fibrosis and we hit it off at once. From then on more and more things became clear, we recognised each other's stories. We then decided to set up a patients' association, the

After living with pulmonary fibrosis for many years, Jenny Penders (1960-2014†) had a lung transplant on 25th March 2003, enabling her to resume a normal life. It allowed her to see her two children, her son Joris and her daughter Sara, grow up to become "beautiful people" as she put it. She was one of the founders of the patients' association Belangenvereniging Longfibrosepatiënten (www.longfibrose.nl) and took part in the production of a DVD entitled "Leven met longfibrose" (Living with pulmonary fibrosis) (see also www.ildcare.nl). She worked for the Trajekt public welfare foundation in the city of Maastricht, where she provided information and support to parents and caregivers of patients. She had two sisters, both of whom also died of pulmonary fibrosis, as did her mother.

Longfibrose Belangenvereniging Nederland, which also gave me a chance to get more information (www.ildcare.nl).

My physical condition kept deteriorating, and eventually a lung transplant proved to be the only remaining option. I was screened for such a transplant in Louvain (Belgium), and they placed me on a waiting list at the end of May 2002. My organs had gradually become affected, including my heart and liver, and I had stomach problems.

In the meantime, my eldest sister was also diagnosed with pulmonary fibrosis and her condition rapidly declined. She died in December 2002, aged 43 years, from pulmonary fibrosis and cancer of the lungs.

Soon after that I suffered a pneumothorax and was rushed to hospital. When I was discharged two weeks later, having had 3 pneumothoraxes and an endoscopy, I was a complete invalid. From then on I had permanent home care. I kept hoping I would get a new pair of lungs soon, before it was too late. In a way, you're just waiting for someone to die so you can live on. A bizarre idea.

My youngest sister had her lung tissue examined in January 2003, and she was also diagnosed with pulmonary fibrosis. It's obvious that the disease is hereditary in our family.

In those days I was on disablement benefit, I couldn't work anymore, couldn't do anything really. I would lie in a bed in the living room downstairs, and in the evening I would use a rollator to shuffle to the stairs, where I used a stairlift to get upstairs. As I found it hard to give up my work, I did so gradually. Fortunately my colleagues showed great understanding; the reactions I got were far more sympathetic than I'd expected, and people really empathised with me.

All physical activities, from brushing my teeth to bending over, combing my hair, getting dressed, talking to people, everything took incredible amounts of energy and air. It's often difficult to explain this to healthy people who breathe automatically and never have to think about it. The few times I went outside, I would sit in a wheelchair with an oxygen bottle. That takes some getting used to, being in a wheelchair or having an oxygen tube up your nose.

People sometimes react as if you're from another planet, and it wasn't always easy to deal with that. Especially at first you have to explain a lot, as your problem is not visible from the outside, and that takes a lot of energy and sometimes I felt hurt. In that sense it was almost a relief when it did become visible to others that I was ill; that greatly increased people's understanding.

I first lived by the day, then by the hour, by the minute and finally by each breath I took. It was on 25 March 2003 that I got the phone call that a donor was available. It was in the nick of time; I wouldn't have lasted much longer. I had a successful double lung transplant, and from that time on I managed to regain everything I had lost before. I've been incredibly lucky, as so far (knock on wood!) I haven't had any signs of the new lungs being rejected by my body, and I'm leading a "normal" life with my kids, who are now (2010 ed) 17 and 12 years old.

My youngest sister died in January 2009. She had euthanasia as she had been completely worn down by the terrible wasting disease that is pulmonary fibrosis. She was only 44 years old. Our family was hit terribly hard by pulmonary fibrosis, and I thank my lucky stars each day that I'm still around. I've just turned 50 and we had a big celebration, the motto being: "Life's there to be celebrated." What having pulmonary fibrosis has meant for me is that I've come to look at life in a very different way; I live more intensely and try to enjoy things more, and I can usually manage that, although I sometimes catch myself fretting over unimportant things (aren't we humans a curious lot!). I am more alert to my body's signals now and I live for today rather than always looking ahead to tomorrow. The pulmonary fibrosis has made me have to give up many things, but it has also caused me to live more consciously. As a result, I've changed many aspects of my personal situation, which I was afraid to do for a long time.

In the period leading up to the transplant, I made arrangements for my funeral, drew up a will and discussed my life with a good friend to achieve "closure". That gave me a huge sense of relief. I would advise everyone to do the same: take stock of your life, get rid of what's unimportant and go back to what's essential, it's very liberating and comforting!

The patients' association has provided me with opportunities to talk about my disease and cope with it, and I hope it will continue to do so for many others in the future. I think if you remain positive and concentrate on the things you can do, rather than on what you can't, and if you go on being physically active as long as you can, it will have a favourable effect on the disease, and certainly on your mind.

In its first year, the pulmonary fibrosis patients' association had 20 members. Now, 15 years later, it has about 250.

It still pursues its aims of raising awareness of the disease, setting up peer support groups and stimulating and funding research. The association has an advisory council of experts and closely collaborates with other patients' associations. It has a website providing information, and the association also publishes its own newsletter, entitled "Leven met longfibrose" (On living with pulmonary fibrosis).

IN MEMORIAM Jenny Penders

Being a doctor is a wonderful profession. You have the privilege of counselling people who urgently need your help. You meet people in all stages of life and with all kinds of conditions. I first met Jenny in 1995. Jenny was a special person, who did not let the message get her down and who grasped every opportunity that was offered to her. Her life was never dull; she experienced wonderful ups but also went through terrible downs of course. As she deteriorated, she entered the transplantation programme; she had no other choice, as this was literally her only chance for survival. While waiting for a donor, she took stock of her life, as she realised that a donor might come too late. She was elated that a donor became available for her in time. She never gave up hope, and in 2003, when she had deteriorated to such a degree that she did not have long to live, she finally got the phone call she had been waiting for: a donor lung had become available. In April 2003, she went to Louvain full of hope, where she received two new lungs and, as she put it, a new chance, a new life. She could not have lasted much longer without the transplant, but fortunately she got it! It was such a joy to meet her as she was walking in the Maastricht city centre less than 4 weeks after the operation, with her daughter Sara. Without oxygen, breathing freely! I had goose bumps!

She lived for another 11 years thanks to the lung transplant, which is also how she saw it. She would not ask why her life was so short, but said she was grateful for the extra chance she had been given, a chance that her mother and her two sisters never got. She was very much aware of this. She set an example for many and her openness enabled her to help many people. Nevertheless, things were not always easy for Jenny, and she paid a price even for this new lease on life: having to use lots of medication with side-effects and having to live with the risk of the transplant being rejected. But she remained positive and organised a big party to celebrate life. She was not just one of my patients, but became a very dear friend. I learned very much from her, and I was able to help others better because of her. She was unique, and I will never forget her: her courage, her energy, her drive, her spirit and power, warmth and love, but also her ability to put things into perspective.

Prof. Marjolein Drent, pulmonologist