

Pulmonary Fibrosis – Progression and End of Life

DISCLAIMER: This document contains information about end of life. This is a subject area that some do not wish to have knowledge of and/or would upset them. Please DO NOT read this document unless this is information you wish to know.

Through my mom's journey with PF, we knew very little. We didn't have access to much support, so we were very much alone and very uneducated. All we knew was that PF was terminal... there was no treatment... and she was going to die from this disease.

We had been told that she would likely succumb to pneumonia if she made it to the very end and how breathing would be very difficult. Mom said she would rather go earlier and quickly from something else rather than "choke to death" in the end. The idea of not being able to breathe is always a daunting one. For anyone who has ever been breathless, they know how frightening it is. **I can tell you that we were somewhat misled by how things can go in the end stages of PF.**

Shortly before mom passed away, I joined my first online support group. This group became a lifesaver when it came to learning about this disease and how to handle things – especially at the end. I am going to share what I have learned from them, as well as so many others over the years – caregivers, medical professionals, and most importantly, patients. I am also going to share information that is helpful to both patients and their loved ones.

DISCLAIMER: Please note that I am not a medical professional. I was simply the caregiver for my mom for 6 years and since then have been a PF Advocate for Patients and Families and the moderator and owner of the Breathe Support Network of Groups. What I am about to tell you is from personal experience and years of learning by talking with others.

First of all, the thing to know about end of life with PF is that patients do NOT have to suffer needlessly. I know that mom and many patients (and their loved ones) are frightened at the thought of "choking" or "suffocating" to death. There are things that can be done to mitigate that for patients.

Before getting to the end stages, it is important for patients and families to talk about last wishes. Where does the patient want to pass on? Do they wish to be at home or in a hospital or hospice facility? Most seem to want to be at home, surrounded by their loved ones. While this is comforting for them, it may seem daunting for the loved ones who are then tasked with handling end of life care and all that goes with it.

There are resources to help. One resource is the support groups. But professional resources are also available. There are two levels of help. The first is Palliative Care, followed by Hospice Care as the end draws more near.

Palliative Care is an in-between. It allows patients to continue with their regular medical care by their doctors, while still offering some in home care by trained medical professionals. A nurse will come into the home and check on them and help them with their care. Many don't even know that this level of care exists so don't take advantage of it.

Hospice Care is the norm for end of life care. However, the old way of doing this is out. It used to be that Hospice couldn't be brought in until a patient had been told they have 6 months or less to live. With PF, it is impossible, in most cases, to know when that is because PF cannot be adequately "staged" (like cancer) and because it is an elusive disease and a patient can go from being stable to passing on – with no warning. PF brings with it what is referred to as "acute exacerbations". This means the disease progresses for no apparent reason. Patients can even go from being early on in the disease – not on oxygen at all or even full time – to being at end of life overnight. Suffice it to say, PF is frequently a moving target.

Having Hospice on board earlier rather than later is a huge help to patients and their families. Patients with PF have been known to be on Hospice for years. They are typically re-evaluated every 6 months to see if Hospice is still applicable. This level of care can really enhance a patient's quality of life and also help the caregiver/s. Hospice care also does NOT mean a patient has to give up all life saving treatment.

Typically patients will be asked to sign a DNR with Hospice care on board. However, if at any time the patient changes their mind and they want to seek medical attention outside of Hospice, they can simply be removed from Hospice care, treated, and then re-enroll in the Hospice program. Additionally, Hospice does NOT mean "no medical treatment". There are many things patients are still treated for while under Hospice. For instance, when a PF patient gets a respiratory infection, they are treated as any other patient – with antibiotics and other medications – to cure the infection. There are many illnesses a PF patient can live through and continue to enjoy a longer life expectancy – with quality. Patients are also allowed treatment for chronic conditions (high cholesterol, thyroid disease, diabetes, etc.) while on Hospice care.

One area where Hospice is invaluable is with coughing and breathing problems.

Many with PF experience a debilitating cough. The cough can start early in the disease or come on later. Some are lucky and never experience this. The cough can be so severe that patients will turn blue, vomit, break ribs, and even lose consciousness. The cough can be severe enough that patients will not go out and enjoy life because they are afraid of what others who don't know them will think. The general population associates cough with being contagious with an illness. This is unfortunate.

There are some over the counter remedies and also natural ways to treat cough. However, with the PF cough, these don't often work for very long or don't work at all. Hospice can help because Hospice nurses have access to prescription narcotic cough medications. Narcotics are the best treatment for cough. Many doctors, especially now with an "opioid crisis" do not want

to prescribe narcotics to patients themselves because they are all “controlled substances”. They are worried about misuse and their own medical licenses. They are, however, willing to let Hospice nurses handle this for them.

Many know that medications like Morphine relax breathing and actually decrease respirations. So why use something like this on a PF patient? The reason is because it truly does relax the breathing effort. The lungs don't fight as hard for each breath and overall, breathing is easier. The meds are not given in a lethal dose. They are given in a therapeutic dose. They are also essential to quality PF care – especially closer to the end of life.

As the lungs become more fibrotic, less air can be taken in. Patients feel more short of breath as time goes on. Not being able to breathe is an awful scary feeling. Morphine and other narcotics are a humane way of treating this problem.

Another medication used is Ativan (or another anti-anxiety drug). Anti-anxiety meds help the brain to not react to breathing problems quite as much. Essentially... the human brain doesn't freak out as bad when breathing is difficult. This makes breathing problems easier for patients to handle.

In the very end, these medications become essential to lessen suffering for PF patients. Withholding these medications will almost surely mean PF patients suffer needlessly.

The other reason Hospice care is essential in the end of life is that Hospice nurses are very adept at letting the family know just how close the end is. With PF, it may be difficult to know when the disease is going to win, but once the body starts into the end of life process, a good nurse can predict more closely when this will happen. Hospice can also offer more in home care for the patient and the family. In the end of life with PF, patients require around the clock 24x7 care. I can tell you that, as much as you may wish, you can't care for your loved one 24x7. You need to care for yourself as well. You need sleep. You need to attend to your own needs or risk causing yourself your own medical problems. One of the worst things is when our loved one passes on and we find we now need a caregiver ourselves. The absolute worst is when we ignore our own needs so much that we pass on before our loved one with PF.

So what things can start to happen as someone with PF progresses and nears the end stages?

As PF progresses, patients who make it to the very end will find that they are likely fighting multiple respiratory infections. As the disease progresses, the lungs can't fight off infection. Many patients who truly go the end of PF may succumb to something like pneumonia because no amount of antibiotics can rid the lungs of the infection. This is why it is so important to treat respiratory infections early on and aggressively. It is also why avoiding exposure is so important.

Other co-morbidities can also appear for PF patients due to the lungs becoming weaker. Heart problems can (and usually do) crop up. Many can be treated to give the heart help. Pulmonary Hypertension is another common side effect of PF. PH is high blood pressure of the lungs. When secondary to PF, it doesn't typically respond well to treatment, though a doctor should still attempt treatment with PH meds. PH is another incurable disease that causes shortness of breath and heart problems.

As the fibrosis spreads, breathing will become much more difficult and coughing will increase for many. Controlling cough is important and use of supplemental oxygen – no matter the liter flow needed – to keep oxygen saturation from falling below 90% is crucial. This process can go on for weeks, months, and even years for PF patients.

Eating becomes more difficult as the body uses oxygen to both chew and digest food. Eating more frequent smaller meals – and foods that require little to no chewing – can be very helpful. Soft foods, shakes, and smoothies will make eating easier. Many patients will get to where they eat very little. At this stage, it is important to help them get what they “want” and not necessarily what we think they “need”. If all they want are milkshakes and hot dogs, it is better than not eating at all.

The natural course for PF is that patients will have too much carbon dioxide (Co2) in their blood. Co2 retention is much like carbon monoxide poisoning. The symptoms are the same. While living with PF, the supplemental oxygen helps keep the Co2 at a lower level. However, as the disease progresses and the lungs become more and more fibrotic, the Co2 will increase. Some patients are put on BiPap to help with this. BiPap uses a mask with a tight seal and pressurized oxygen to help push the Co2 out of the body. Unfortunately, as things progress, patients may “sleep” for long periods of time. It's almost like a comatose state. Patients have been known to sleep for days at this point, then wake and be fine for a few days, then sleep some more. Patients have even been known to go through this over and over again before they finally succumb to PF. *(NOTE: The only way to know for sure if Co2 retention is happening is with a blood draw from the artery in the wrist called an “arterial blood gas”.)*

Finally in the very end, the body starts to shut down. Kidney function is a tell tale sign that end of life is nearing. When the patient's kidney's stop functioning, they typically only have days left.

Because PF patients “sleep” a lot during the end, this actually helps with suffering. They suffer less. This does not however mean that they are unaware of what is going on around them. Much like we have heard patients in a coma or sedated for surgery can “hear”, someone with PF who is in this “sleeping” state may also be able to hear. It is important to keep conversations around them positive. It is also important to talk to them and comfort them. Be with them. Touch them. Human touch is an amazing comfort. Simply stroking their arm, holding their hand, or stroking their hair can be of comfort. Telling them encouraging positive things can help make their crossing over easier.

Because patients may be able to hear while in this state, hard as it may be, it is important to **not** say things that might make them fight to live. The will to live is very strong and some will fight because they are afraid of how their loved ones are going to feel when they are gone.

It is human nature to want to tell our loved ones “I am going to miss you so much” or “please don’t leave me” or “you can fight this”. The reality is that part of life is death and one must ask themselves, “How much do I want my loved one to suffer so that I am not alone”? I have been through this with my mom and I can tell you it is terribly difficult to do this.

You might be wondering “How can I medicate my loved one when they are sleeping and they are at home”? That is a great question and there is a way. Morphine and many other medications come in a drop form and/or in a small dissolvable pill that goes under the tongue. These are very helpful at easing suffering in this situation. Those of you caring for your loved one with PF will know if they are suffering. You will be able to detect it in how much they are struggling to breathe. You will most likely know when to administer these medications to help them out.

It is important to know that the end of life process can take time or it can be short. Having multiple people around through this is helpful. Family, close friends, and Hospice (or being in a hospital setting). Doing what is necessary to lessen suffering is essential.

It is important for both PF patients and their loved ones to keep open lines of communication and be on the same page.

I know for many, all of this can be overwhelming and terrifying. When I was taking care of my mom, I wondered how in the world I was going to handle this. When the time came, I did handle it. I don’t know how, but I did. You all will too.

I hope this information has been helpful and somehow helps ease your mind about what might be coming. Please note that how PF progresses, what a patient goes through, and what end of life might entail can be different for everyone. There are no guarantees surrounding how it may go for you or your loved one. Being open, honest, and prepared can help everyone involved.

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