Advances in the Treatment of Interstitial Lung Disease

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Henry Lasky

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Introduction

Andrew Schorr:
Managing what's called interstitial lung disease, which has many causes, many subtypes, it can be quite complex. So when you're faced with this diagnosis you really need a specialized team on your side. Coming up you'll hear from the director of the interstitial lung disease program at UCSF Medical Center and his patient, Henry, as they discuss the latest in diagnosing and treating ILD. It's all next on Patient Power.

Hello and welcome to Patient Power sponsored by UCSF Medical Center. I'm Andrew Schorr. Well, we're going to learn today about really a constellation of illnesses that broadly are called interstitial lung disease, and it can be quite complex, have all sorts of causes, some that are known and some that are not, and unfortunately what it can lead to is scarring, sometimes inflammation of tissue in the lung, but the bottom line is you're not doing well with the transfer of oxygen to the blood and you can imagine what that leads to. Fatigue, shortness of breath, makes your heart work a lot harder. It can be quite serious. Now, there are a variety of treatments for it and there’s a lot of research going on.

We're going to hear about all that, but first I'd like you to meet someone who was diagnosed with it in 2002, and that is Henry Lasky. We could also call him Judge Lasky because for many years he was a federal administrative law judge in San Francisco and all around the western states. But in 2002, Henry, tell us what was going on with you that led to this diagnosis. How were you feeling?

Henry’s Story

Henry:
First of all, very happy to be here.

Andrew Schorr:
Thank you.

Henry:
And I was having a problem with shortness of breath, for example particularly on stairs, and there had been almost a couple of years had gone by where I
complained about it to my primary care physician whose response was that I have to get used to the idea that I’m getting older and it's all part of getting older and I can’t run up and down stairs the way I used to as a younger man.

**Andrew Schorr:**
Right. And we should say you're 75 now, so we'll go back a few years. But you had been a runner. Used to run, I know.

**Henry:**
Yeah, I jogged. I wouldn't say run. I prefer jogging.

**Andrew Schorr:**
The dogs would take you out too, and then you were a tennis player, an avid tennis player.

**Henry:**
And I played tennis a couple times a week with a group, which I enjoyed.

**Andrew Schorr:**
This feeling of shortness of breath to you felt, though, more than just getting older.

**Henry:**
Yes. It felt more than just being a little winded. I actually would walk up--if I walked up the stairs of my house I felt like I was wiped out.

**Andrew Schorr:**
All right.

**Henry:**
It was that debilitating.

**Andrew Schorr:**
So you asked for a referral to a pulmonologist. What happens then?

**Henry:**
Ultimately, ultimately I did, because I think you have to be a very strong patient advocate.

**Andrew Schorr:**
We call this Patient Power. I agree with that.

**Henry:**
I had to push my primary care doctor to give me a referral to a pulmonologist, and he did.

**Andrew Schorr:**
And ultimately you were referred to UCSF.
Henry:
The local pulmonologist referred me to UCSF to see Dr. Golden specifically.

Andrew Schorr:
Who is a pulmonologist in this special area that deals with interstitial lung disease at UCSF. They have a center for it. So ultimately they take a biopsy of your lungs. They go down with a little scope, right?

Henry:
Well, I don't know how they do it.

Andrew Schorr:
Oh, okay.

Henry:
This was a surgical procedure. I was in the hospital overnight.

Andrew Schorr:
All right. We'll find out more about that because we're going to have the head of this group in just a second. So the biopsy comes back and they tell you you have a form of interstitial lung disease. What is your form called?

Henry:
It's called respiratory bronchiolitis.

Andrew Schorr:
And this must have been a shock or you probably never heard of it.

Henry:
Well, in one respect it was. Dr. Golden had said to me the good news was I didn't have idiopathic pulmonary fibrosis, but I did have respiratory bronchiolitis, which he felt was somewhat of good news compared to what it could have been.

Andrew Schorr:
So you went on different treatments, first prednisone at high dose, which is tough.

Henry:
Yes, it is.

Andrew Schorr:
And then later an immunosuppressant, a drug called CellCept.

Henry:
That's correct
Andrew Schorr:
And that worked for quite a while, and while you did have some hospitalization and complications I know we should say that now you live a pretty good retired life in Petaluma. You use oxygen but you're a busy guy?

Henry:
Yeah, I'm still very busy. I only use oxygen on exertion, for example like when I'm working out in the gym or when I'm walking my dogs.

Andrew Schorr:
How many dogs do you have, Henry?

Henry:
I have four.

Andrew Schorr:
So they walk you.

Henry:
They walk me. That's correct.

Andrew Schorr:
And you're involved in a support group that the UCSF program has, and I understand you meet once a month?

Henry:
That's correct.

Andrew Schorr:
Has that been helpful?

Henry:
Very much so. Very much so because it has given me a great deal of perspective, and, you know, there's a number of people in this particular group who are much younger and have far more serious disease symptoms than I do.

Andrew Schorr:
Yeah, you can be an inspiration to them, though, just showing what someone can do.

Let's meet the head of the program, and that is Dr. Harold Collard who is a pulmonologist. He's director of the interstitial lung disease program at UCSF Medical Center.

First of all, Dr. Collard, must make you feel good that here's a man with this diagnosis, and I know there are many forms, and he's living a pretty good life.
Dr. Collard:
Yeah, it sure does. Henry is a great example of a success story.

Diagnosing Interstitial Lung Disease

Andrew Schorr:
Now, I described the disease or all these subtypes as having the common feature of either scarring of the lung tissue or maybe inflammation, but the bottom line being limiting oxygen. Did I say it right? And how many different types do we have anyway?

Dr. Collard:
I think that's right, Andrew. There are many types of this. I think that's one of the big challenges in interstitial lung disease. There are some major categories but really dozens of different conditions that can cause scarring or inflammation in the lungs, so one of the big challenges is trying to sort through that because both prognosis - what happens over time, and treatment depend on an accurate diagnosis.

Andrew Schorr:
Now, Henry said he went to his doctor and his doctor had what probably most doctors would say to a man, you know, of retirement age and maybe you'd think could be slowing down a little, well, if you're short of breath that's going to happen to all of us. Or there could be much more common diagnoses. Somebody could have asthma, some allergy that's popped up, or even COPD maybe if they'd been a smoker. So it would seem like it takes a while to get to an accurate diagnosis.

Dr. Collard:
Yeah, it does take a while. We've looked at this a little bit in survey form. It's usually over a year from onset of symptoms to time of diagnosis, and, you know, some of that is just unfortunately the nature of relatively rare conditions, but I think some of it is also a need for increased awareness of the condition and diseases.

Andrew Schorr:
Well, we're working on that today. So if someone comes to your program then what sort of exams will you do to see if in fact it is one of these conditions that leads to this interstitial lung disease and then have a plan that's right for that type?

Dr. Collard:
There really are, I think, three key things that can be done really in certainly any pulmonologist's office to really get at this diagnosis. The first is a careful physician exam, and although there are no specific findings to specific causes of interstitial lung disease, noises on the respiratory exam called crackles, really kind of a crackling sound in the lungs, is very common and should make people think of this diagnosis.
The other two tests which are really pretty standard measures in pulmonary medicine are, number one, something called a pulmonary function test which is a test of your breathing mechanics, and then the second one which is a certain form of CT scan or computed tomography scan which is called a high resolution CT scan.

Andrew Schorr:
Now, Henry mentioned that he had an overnight procedure where you went down into his lungs to get some tissue to see exactly what type he had and then I guess leading to a decision on what would be appropriate treatment. Tell us about that, about actually getting tissue to see what you’re dealing with.

Dr. Collard:
About half the cases, maybe a little less than half of cases, the CT scan doesn't give you enough information. Sometimes you can make the diagnosis of the specific condition causing the interstitial lung disease from the CT scan and from the history, but when you can't do that a surgical lung biopsy is what's generally needed, and what Henry had was a procedure called a VATS biopsy. It's a minimally invasive surgical approach, and it involves taking a couple of small samples of lung tissue out to look at under a microscope, and the pattern of abnormality on that biopsy can help us make the diagnosis.

Andrew Schorr:
So he talked about a certain type and he was told by Dr. Golden, one of your colleagues, well, it wasn't this other type idiopathic pulmonary fibrosis. First of all, what does that word mean, idiopathic?

Dr. Collard:
It means we don't know the cause, so it's of unknown etiology. So one point I like to make about it is that we know a lot about the condition, we just don't know why it happens. But this specific form, idiopathic pulmonary fibrosis, is one of the causes of interstitial lung disease, and it's we think a somewhat unique cause in that it has a different biology to it than many of the conditions.

Andrew Schorr:
So step one is which one—is it in fact an interstitial lung disease. Step two is which type is it. So then tell us about your array of treatments today and then maybe where it's headed too.

Treatment Types

Dr. Collard:
So there are two main classes of therapies, and you hit on them earlier in the introduction. One is therapies aimed at inflammation, and those are ones that Henry has been on. Prednisone is kind of the old standard and we still use it a lot and it's very effective in certain people. And then there are other nonsteroidal, we call them immunomodulator therapy so they suppress to some extent the immune system which reduces inflammation.
There are three that we most commonly use in interstitial lung disease. One is called Imuran or azathioprine, which is the generic name. And the one that Henry was on is called CellCept, or mycophenolate is the generic name. And both of those have been very effective in certain forms of interstitial lung disease that have an inflammatory component to it. So that's one kind of group of medications we use.

The other group is really aimed at treating fibrosis, so not the inflammation but the scar that forms as a result of inflammation. And this field is much newer and we have fewer proven agents. This is where a lot of research is happening and looking at therapies that are targeting the scar formation itself. And particularly in the disease idiopathic pulmonary fibrosis which has been mentioned, which we abbreviate IPF, this type of therapy again aimed at fibrosis is critical because inflammation seems to play a much smaller role in that form of interstitial lung disease than in others like respiratory bronchiolitis that Henry has.

So I think in terms of treatments we have there really are those two camps, but there is a very active both basic and clinical research program both nationally and internationally, but we're very active here at UCSF as well, and I think that is a cause for a lot of hope and excitement in this field. Because really in the last 20 years we've learned a tremendous amount about the biology of these diseases, and now the challenge is to translate that from the lab into patients, and that's where we're at now and we're making progress but there's still a ways to go.

Andrew Schorr:
Let's talk about your program now. Henry, you said you participate in a support group.

Henry:
Yes.

Andrew Schorr:
And that's helpful. So I imagine there's staff that's involved in helping facilitate the support group, so that's part of the program.

Henry:
Yes. Sally McLaughlin, that Dr. Collard mentioned, is the facilitator of the support group.

Andrew Schorr:
And she's a nurse, Dr. Collard?

Dr. Collard:
That's right. She's our clinic nurse.
Andrew Schorr:
Okay. So tell us about your group. Dr. Golden, we heard about, yourself, pulmonologists, there may be others. What other specialties come into play at an academic medical center such as yours to support people with this illness?

Dr. Collard:
That's a good question and it's an example of why we exist actually because our real mission is to help patients and their providers to understand the diagnosis and management of these diseases because it is complex and multidisciplinary. So the key disciplines really are pulmonary medicine, radiology and pathology or looking at lung tissue. So we have specialists in each of those fields that are part of the program and that meet weekly to go over cases and talk about research. We also have rheumatologists because a lot of rheumatologic conditions like rheumatoid arthritis, can be associated with interstitial lung disease. Dermatologists are often involved. Cardiologists because of the link sometimes with a condition called pulmonary hypertension, which is a kind of a cardiac, cardiovascular correlate or complication of these diseases.

So often it requires really getting together a number of subspecialists and ideally sitting in the same room and talking about the case because it's a complicated enough picture and an individualized enough disease that everybody is a little different, and understanding all of the nuances can be very difficult if you don't have that kind of a team approach.

Andrew Schorr:
I believe that. Henry, how about you? You feel you came to the right place and your care is in the best hands now?

Henry:
Oh, absolutely. Absolutely. I've feel very, very fortunate actually to be a patient of this group of physicians because I just have full confidence in them. They're very, very capable.

The Importance of Early Diagnosis

Andrew Schorr:
Dr. Collard, so lung tissue is being damaged, and it would seem to me that if somebody could get to a diagnosis earlier you might be able to help them more. Am I right about that? And how do we do that?

Dr. Collard:
Yeah, you're right. And I think actually in listening to Henry's story, Henry is a perfect example of where early diagnosis makes a difference. I think--I've thought a lot about how to do this. It's difficult because interstitial lung disease can creep up on you, and you can have significant impairment of your lung function and not necessarily notice it a lot because you have a lot of extra lung function built in. So sometimes you won't have symptoms until things have moved along.
But what often is the case, as Henry described it, is you have symptoms and it's just a slow process of getting to the right answer, and I think there are a couple things that are important. Probably the most important is patient advocacy and patients taking control and responsibility for their condition, and that's a lot to ask but it's also really essential to getting good care. And Henry is nice to credit our program, but I think a lot of his success is because of his own approach and being proactive about things. I think that's a critical thing and why I'm really glad to be part of this show, Andrew, because I think things like this help people to realize they can do that.

The other key component of it is in the community making sure that primary care providers, pulmonologists and the other fields I mentioned, radiologists, pathologists, people who may see these conditions come up unexpectedly perhaps, a preoperative CT scan for example, having them understand the importance of an early diagnosis and how that impacts management I think is critical. So if we can make progress in those two areas I think that will get us a long way towards early recognition.

Hope for the Future

Andrew Schorr:
One last question for you and then we'll give the last word to Henry. Dr. Collard, are you hopeful? I know there are many different subtypes and different situations are different, but what would you say to people listening as far as giving them hope that they can hopefully have a good quality and hopefully a longer life?

Dr. Collard:
Oh, I think there's tremendous hope in this field. And as I mentioned earlier we've made so much progress in understanding the biology of disease and that is the essential first step in moving forward. For many of these conditions we have effective therapy, we just need to get to patients early enough because once scarring is established and set that's where we are limited in our ability to reverse things. So for many patients we're already at the point where getting to things early and getting on appropriate therapy makes a big difference.

For others who have conditions that we still struggle with treating, I think the hope really should be based on our very sound understanding of the biology and a knowledge that that's how medicine progresses, and the more involved people can be in their disease and even if interested in participating in clinic research studies that help move these findings into patient care, that's the way to move things forward. And I think we'll see a lot of progress in the next several years.

Andrew Schorr:
Well, Dr. Harold Collard, director of the interstitial lung disease program at UCSF Medical Center, we wish you and your team well, and thank you for all you do.
And, Henry Lasky, the last word to you. Are you hopeful about your continued busy life? You take some oxygen, but it sounds like you are still living a full life despite the diagnosis.

**Henry:**
Oh, yes. I just keep moving. Let's put it this way, so long as I'm stable I try to do everything that I can to maintain a normal lifestyle. I'm a very involved person. I enjoy the activities that I do, and I just will continue to do so. I'm also very fatalistic about everything, and I'm in good hands, as they say, and I'm doing everything I can do. I can't be concerned about things that are out of my control.

**Andrew Schorr:**
Good for you. Well, sounds like you and the dogs and all your activities there, good for you in Petaluma. Judge Henry Lasky, thank you for being with us and we wish you all the best.

**Henry:**
Thank you so much. It's a pleasure.

**Andrew Schorr:**
This is what we do on our Patient Power programs. Thanks to UCSF Medical Center for helping make them possible and connecting you with leading experts from UCSF Medical Center and really inspiring people like Henry Lasky. I'm Andrew Schorr. Thanks for joining us. Remember, knowledge can be the best medicine of all.

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