Interstitial lung disease: end of life care

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In conversation with

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This video has been developed and funded by Boehringer Ingelheim
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Slide 1: Interstitial lung disease: end of life care

Slide 2: By the end of this module you should be able to:
- Outline the impact of ILD on life expectancy in patients with a diagnosis of ILD
- Identify appropriate palliative care for symptom control and when to consider:
  - Oxygen therapy
  - Opiates
  - Benzodiazepines
  - Describe which patients may be suitable for a lung transplant

Dr Steve Holmes (SH): So, can we talk a little bit about prognosis? What is the future for our patients with interstitial lung disease?

Slide 3: IPF has the worst prognosis of ILDs with a survival rate of 20–30% over 5 years\(^1\)–\(^3\)

Professor Joanna Porter (JP): So unfortunately, the prognosis for patients with the most severe forms of interstitial lung disease is not great. And before treatment was brought in then average life expectancy was 2–3 years for patients with IPF.\(^1\) But one has to be mindful that these diseases, they are a very heterogeneous group of patients and some patients will deteriorate more quickly than that and others will go on for many years. And actually, this is really what’s made drug development so difficult is that clinical trials have had to take into account the heterogeneity of the population, so has needed very large groups of patients to show significant differences. So, I always say to patients all we can do is we can give you an estimate of what an average patient with your illness would... life expectancy – but that doesn’t necessarily mean that applies to you.

SH: And I guess in the palliative care zone, that fits in with very much this unpredictable or you can’t quite say when the end will be, and it fits in with the non-malignant conditions where making a precise estimate is much more complex: linked into the disease severity, the aggression of the disease and how things progress?

JP: Absolutely. So, there are scoring systems. There’s something called the GAP score for IPF which takes into account gender, age and lung function, and this gives us a slight prediction as to whether they’re in a very poor prognostic group, or less so.\(^4\)

Slide 4: The overall goals of treatment in ILD are to improve quality of life, maintain activity levels, control symptoms and prolong survival\(^5\)
JP: And we’re working all the time at UCL, and I think other people are, to find better prognostic markers so that we can pick out the patients that are going to do badly and refer them early for transplantation if appropriate. And we’re then able to...introduce palliative care at a time that’s appropriate. But I think that one thing I would say is that every time that we meet patients, we should be mindful that we should be addressing potential palliative care needs and we will often refer them for symptom control even when we still feel they have a reasonable length of life ahead of them.

SH: And that’s probably very important for us to remember as well, isn’t it? Is that even if we’re treating, if we can improve symptoms as well as change the prognosis, we’re making a difference to that person’s life. So, the fundamental improving symptoms, be that their psychological difficulties with what’s going on, sometimes their dysfunctional breathing technique, sometimes their anxieties about what the future will hold. If we can work with those and allay some of those, we can make a difference.

SH: You mentioned transplant in passing, what are the key groups of people where you might consider a transplant?

JP: So, I think we... although there’s no specific age cut off for patients for transplant, I think we have to be mindful that older patients definitely don’t do as well as younger patients. So, for most patients, what we would look at is the risk–benefit of undergoing a lung transplant. And as I say to patients: a lung transplant is not like having your batteries changed, but you exchange one chronic disease for another, and you’ll be on life-long medication and probably with progressive lung deterioration even in the transplanted lung. So, we’re looking at patients who actually have a reasonable quality of life, but we think the disease will catch up with them at some point and probably within the window of 40–60 years of age and with really very few other comorbidities. So, even a whiff of ischemic heart disease or coronary artery disease they wouldn’t be accepted onto a transplant list because of the acceleration of that disease once the transplant has been carried out.

SH: Are there any key tips you have for looking after those patients who are moving into a much more symptom or palliative-care-led zone that are different perhaps then people with COPD or with other non-malignant conditions?

JP: I think probably, what’s different from COPD is just the rapid rate of progression towards the end. So, I know COPD patients who can really go on for many, many years with terrible lung function and very poor quality of life, but actually they do carry on like that for years. In IPF it’s usually... that’s usually not the case. In our experience patients often die of infection or ischemic heart disease. Their pulmonary fibrosis is caused... has been a key player in that. So progressive respiratory failure, and we try to keep these patients at home or at least where they choose that they want to die.

JP: These discussions are very difficult, talking to patients about where they want to die and how they want us to plan for their death can be very difficult, and in my experience it’s almost better being brought up really early on actually after the diagnosis, so that they still feel that’s a long way off and we can talk about it slightly more freely. I think when patients really are nearing the end, the conversation then can become a bit more difficult unless they know you very well. I think GPs play a crucial role in all this because you know the patients. You’ve often known the patients for years and you’ve often known the families for years and...
SH: I guess one of the common things we see with symptom control, you mentioned hypoxia earlier as an issue with interstitial lung disease, what about oxygen in that palliative care phase?

JP: So many patients, I think, are terrified of not being in hospital because they’re terrified of the shortness of breath and the pain and the fear that they may experience. So, most of our patients will be on some sort of long-term oxygen therapy actually as they approach the end, and I see no reason to limit that actually at the end. And very few of them are in type 2 respiratory failure anyway, so we can give high-flow oxygen, and we can use opiates to take away the feelings of breathlessness, also to help with the cough, and benzodiazepines, again we can use those to take away that awful feeling of air hunger that these patients suffer from.

SH: And those are often quite well-controlled on low doses, aren’t they, of the opiates...

JP: Absolutely right. It’s often just a...

SH: … rather than escalating for pain?

JP: … just a low dose that just makes the patients feel so much better. Even when they still have a reasonable length of life ahead of them, we will often use opiates to control the symptoms.

SH: Good. So what I’ve picked up from this section is that, although we’re getting an improved prognosis for a lot of the interstitial lung diseases and especially with interstitial pulmonary fibrosis, we’re at a zone where still a considerable number of these people may have more aggressive disease that we have to manage in a palliative way.

The palliation is linked in very much to the similar groups we deal with that are non-malignant and require good symptom control and excellent communication. And again, the more we can talk across boundaries to make that happen and the more we can share that experience and the support for the patient and their family, the better.
References:


