

# What do patients value and what needs to be considered?



**Care of IPF patients is more than just drugs...**

Meena Kalluri M.D.  
Multidisciplinary ILD Clinic  
University of Alberta

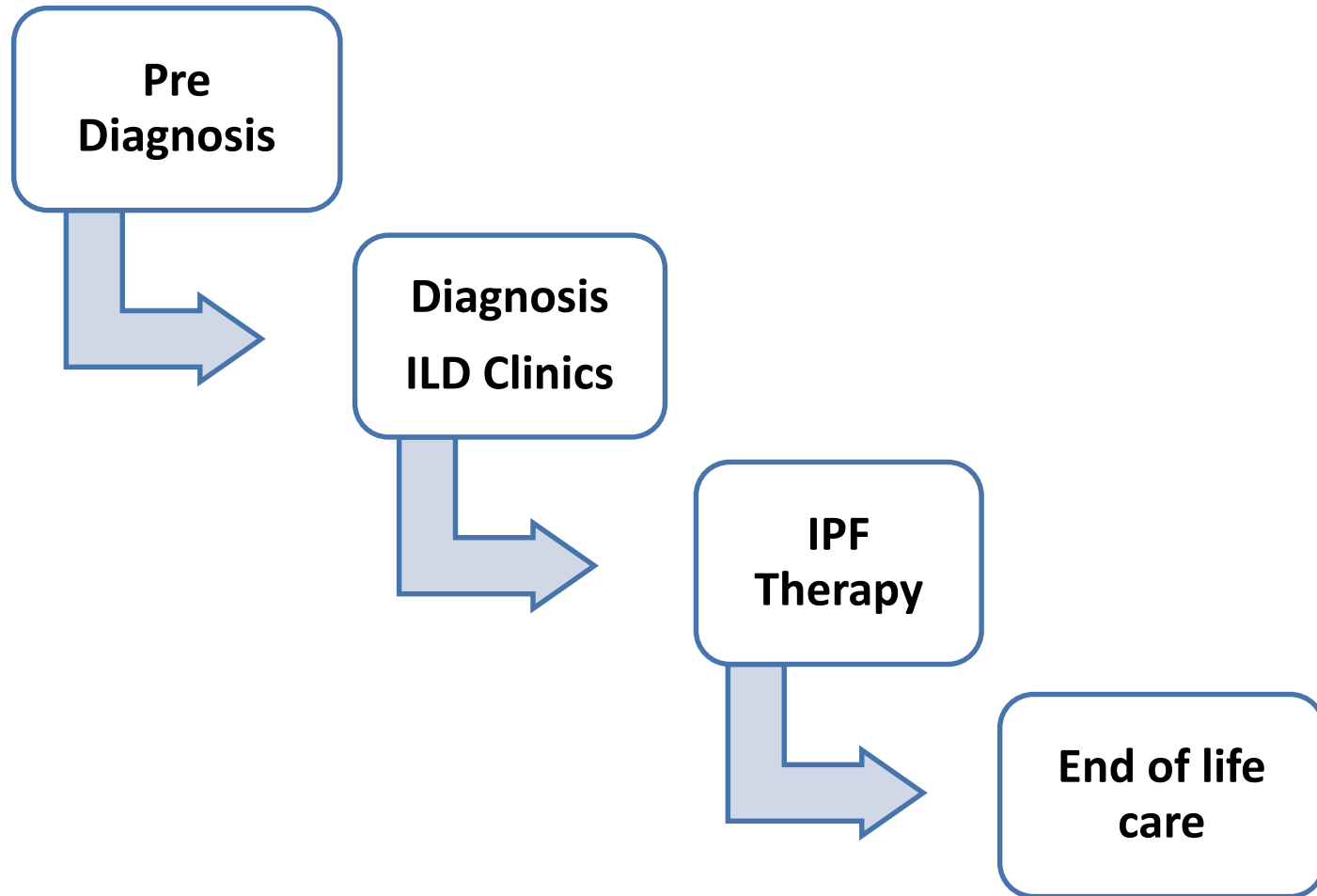


“You matter because you are you and you matter to the last moment of your life. We will do all we can to help you, not only to die peacefully but to live until you die.”

Saunders

# IPF Journey

Patient and caregiver experience



# What do IPF patients value most?

- Pre-diagnosis phase

- Delayed diagnosis

- *“It started about 5 years ago, but I didn’t realize that something was terribly wrong. It just came gradually. I was diagnosed with something “fibrotic” and was told that I probably had untreated asthma.” (Patient 01)*

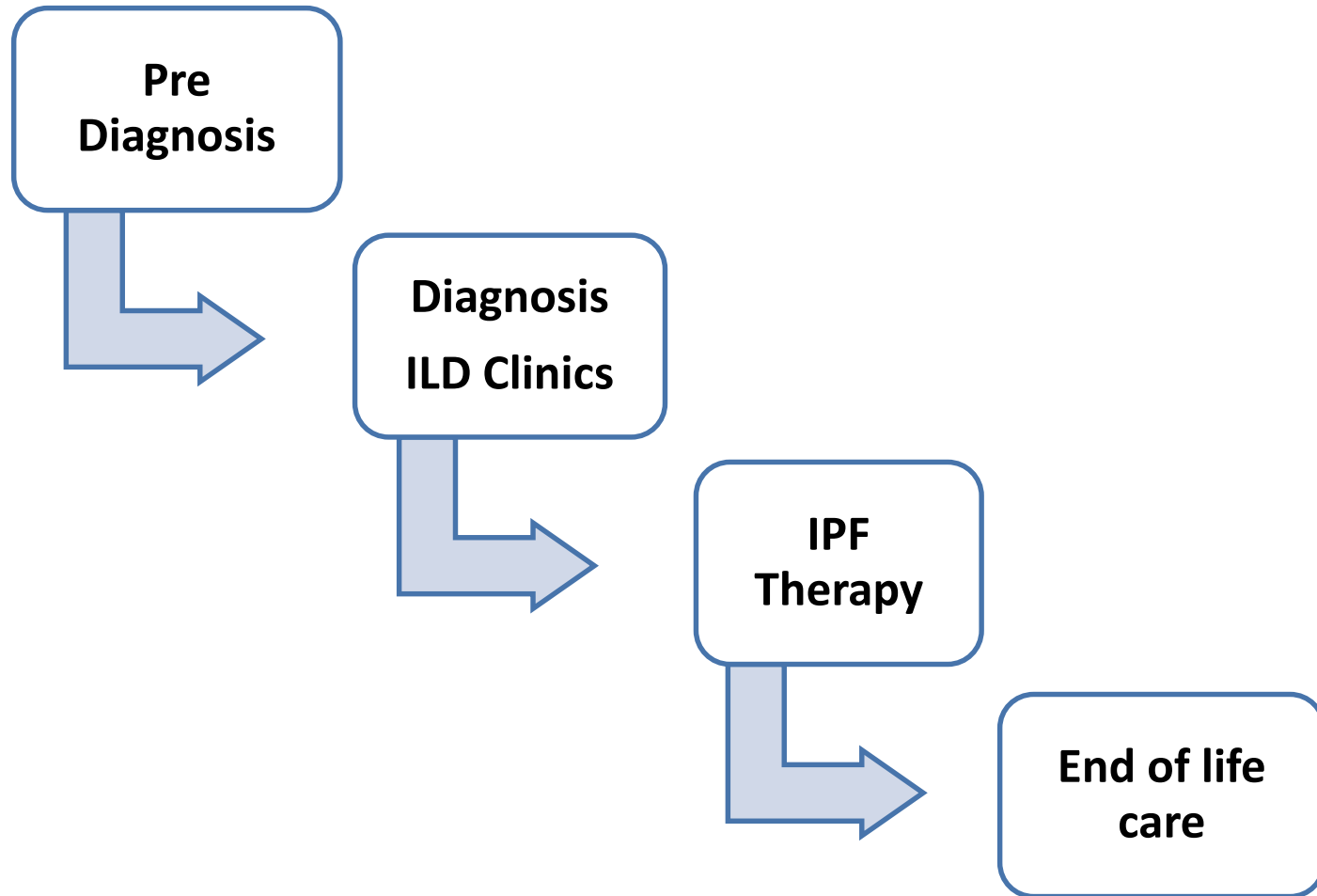
- Lack of sensitivity

- “I think the best thing would be if the hospital were a bit more aware of gathering the family and having a good talk about this.” (Caregiver 01)
    - “In the beginning I felt terrible. When will I die and how? It has been awful, and I have been sad ... I felt I was on the scaffold.” (Patient)

- Delayed referral to a specialist

# IPF Journey

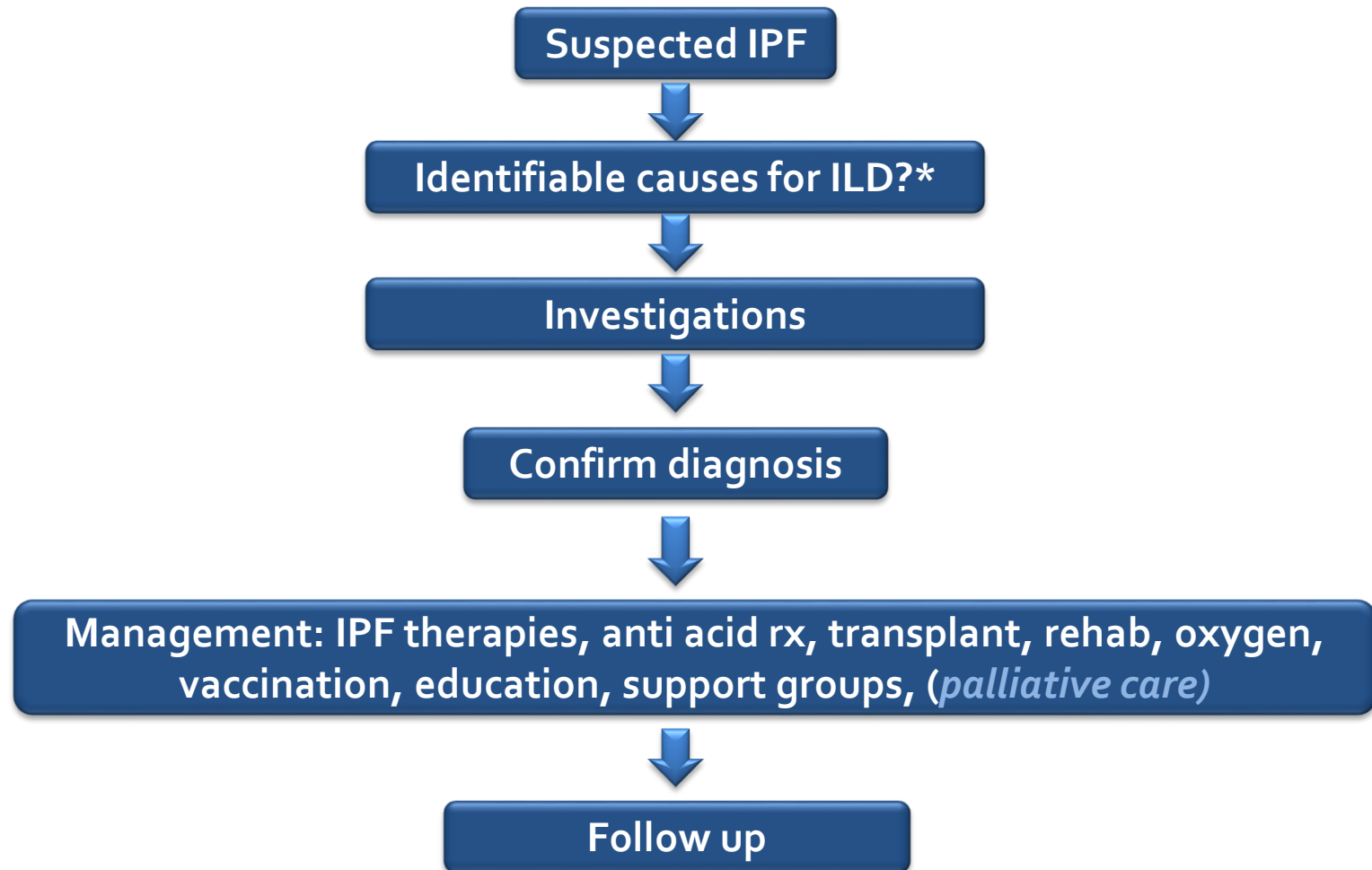
Patient and caregiver experience



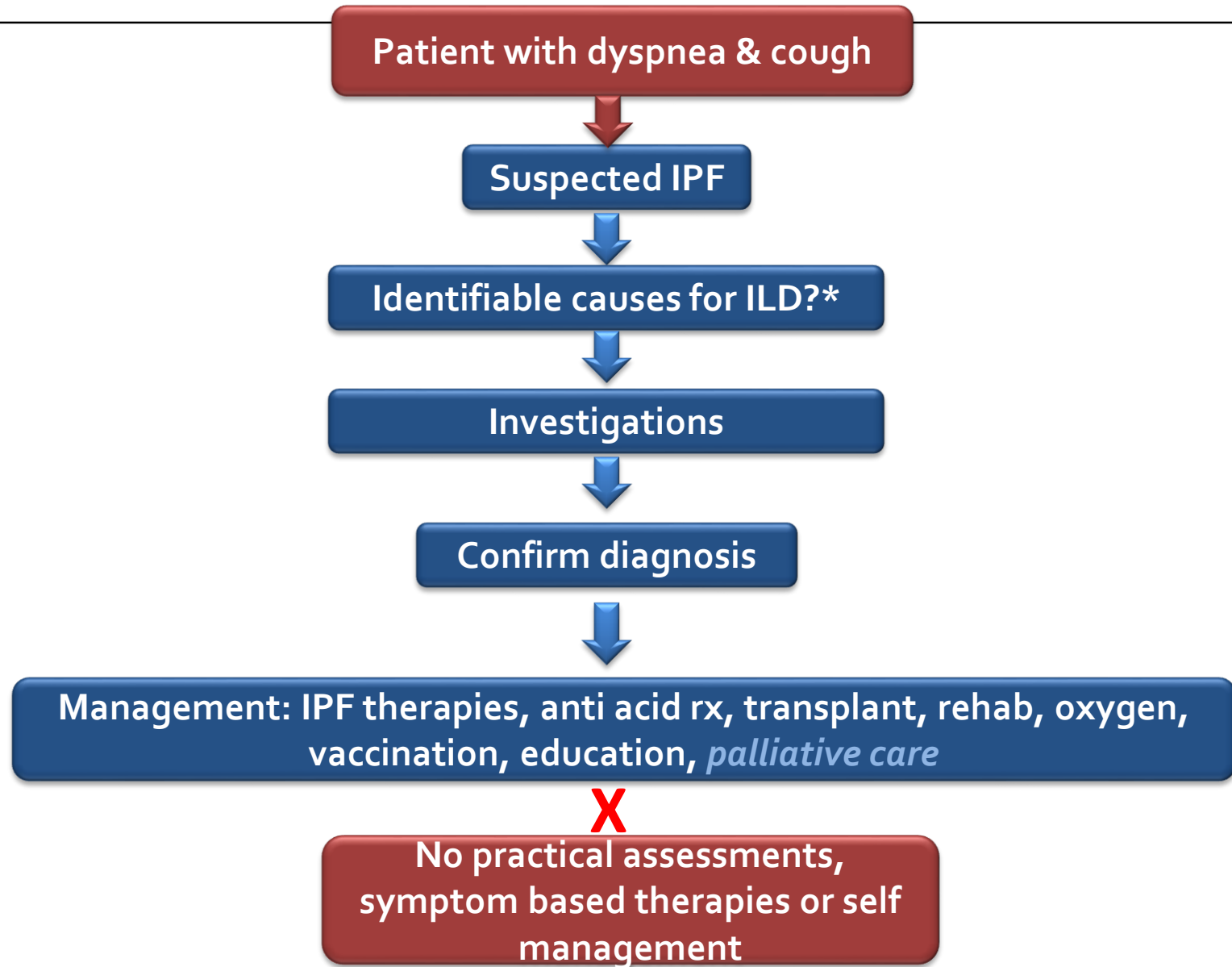
# What do IPF patients value most?

- **Diagnosis and ILD Clinics**
  - Numerous qualitative studies
  - Patient and caregiver observations
    - Very grateful to have a reliable source of information but..... the focus of the consultation is “Disease oriented”
    - The clinic experience was completely disconnected from their disease experience
    - Want clinics to incorporate pragmatic needs assessment that captures the impact of the disease on their lives

# Diagnosis of IPF



# Diagnosis of IPF





# Symptom burden in IPF

Qualitative study of patients, caregivers and health professionals

that breathlessness was the overwhelming symptom. Many participants stated how their breathlessness had taken on a life of its own and was consuming them. This is illustrated

I'm breathless, always breathless. (Ruth, in her 50s, Black Caribbean IPF patient)

Coughing bothers me incredibly in the morning and also in the evening, and generally all day [simulating a cough], if I do anything strenuous I have a slight continuous cough ... and the phlegm tastes bad. (Patient 01)

when it's really really bad, I'd make a trade with the devil (...) because I'm so (...) flat and exhausted and [I] think well I'd rather not go on. (Peter, in his 60s with advanced IPF)

she'll panic because although she tries not to but em she would panic because it's not nice not being able to breathe you know ... (Anthony, husband to Betty)

I can't go anywhere [...] ... I don't don't [really] have a life I'm sitting indoors everyday ... I used to be meet friends and have coffee and it [would] give you a bit of life back ... (Mary, in her 70s with advanced IPF)

Um I mean I have never seen quite so much phlegm (laughs nervously) and he was literally choking on it, he was deeply blue and and there was a sense of sort of hopelessness that nobody could actually do anything about it and I thought there probably were few few worse ways to die than that when I saw it in that instant, sort of haunts me a bit today. (Palliative Care Consultant)

The patients get used to the breathlessness, their doctors and nurses get used to the breathlessness, and (...) the penny doesn't drop that maybe they need to have um (1) to use drugs for the symptomatic relief of breathlessness. (GP).

## The active identification and management of chronic refractory breathlessness is a human right

Currow DC, *et al. Thorax* 2014;**69**:393–394. doi:10.1136/thoraxjnl-2013-204701

score of  $\geq 3$  chronically. Despite the prevalence, severity and chronicity of this symptom and an evidence base of affordable and safe interventions, chronic refractory breathlessness remains grossly undertreated. Many

patients and clinicians accept the presence of the chronic refractory breathlessness as an inevitable part of an illness, with no thought of treating the symptom despite an evidence base for its safe treatment. Consensus

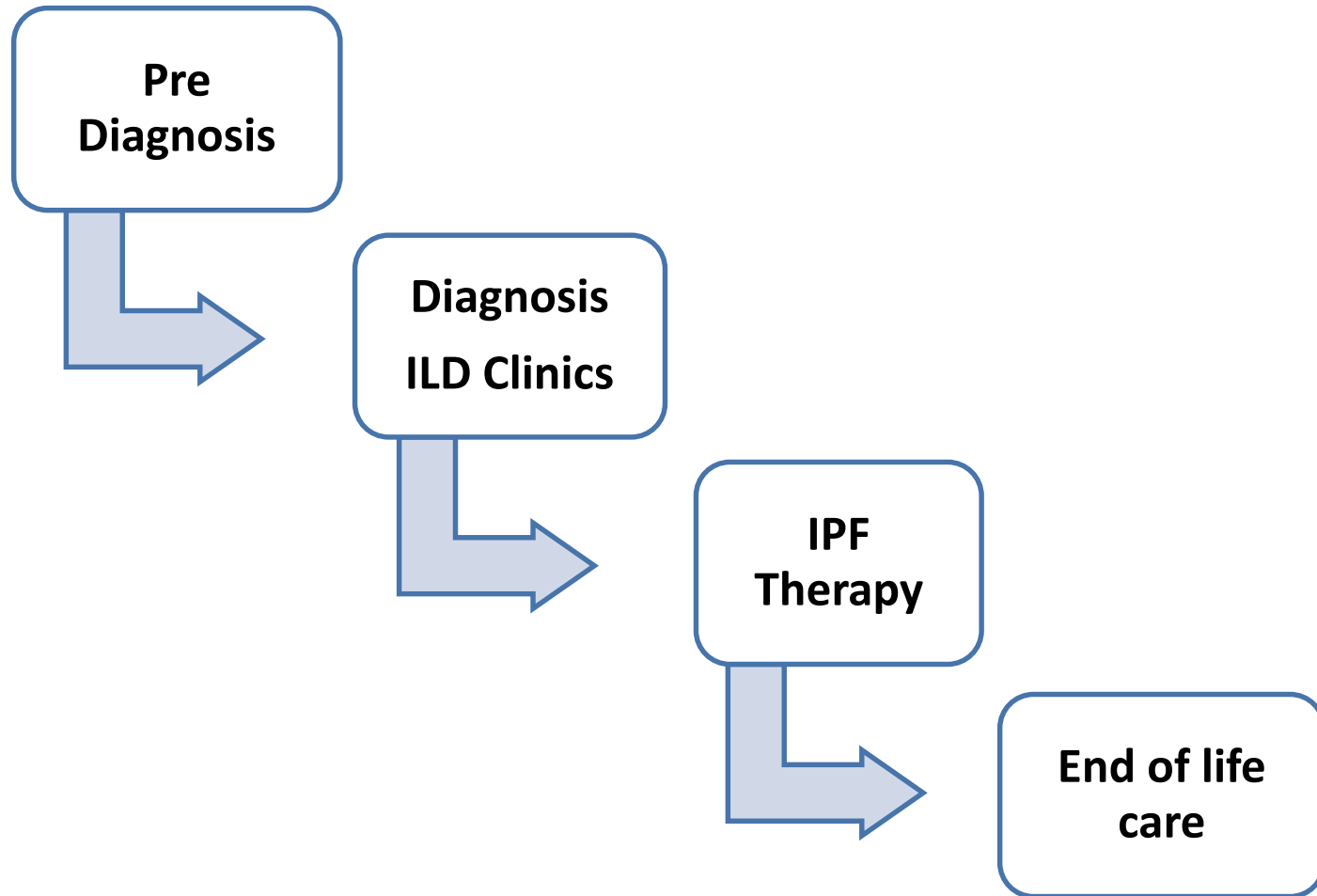
now endorse such a clinical course. Failure to enquire about, assess and properly treat chronic refractory breathlessness with opioids as outlined in specialist clinical guidelines is now an unacceptable level of care ethically and is, arguably, a breach of people's human rights. Adequate pain control through access to pain

## Specialist Palliative Care is More Than Drugs: A Retrospective Study of ILD Patients

- Review of medical records of 45 patients
- 93% (42/45) had breathlessness in the last year of life
- 63% no palliative care consultation
- Treatments
  - Non pharmacological treatments rarely used
  - 22/45 received opioids
  - 8/45 received benzodiazepines
  - All had documented benefit
- Advance care planning
  - 87% not discussed
  - 17% (8/45) had preferred place of care documented
  - 13% (6/45) had preferred place of death documented
  - 75% (34/45) died in a hospital

# IPF Journey

Patient and caregiver experience



# IPF Care issues

- Equal and fair access to IPF specific therapies
  - Criteria: stop rules
  - Coverage
- Easy access to supplementary oxygen
  - Terminal condition- waive burdensome rules and criteria
- Symptom based therapies
- Pulmonary rehabilitation
  - ILD specific content

# Be Honest and Help Me Prepare for the Future:

## What people with ILD want from education in Pulm rehab

Strong themes emerged:

- ILD specific content is not being delivered

*“Some of it(the education) was really good, some of it was common to all of us, but there wasn’t anything that was specific to ILD.”*

- Need for systematic advance care planning in clinics/ rehab programs
  - the importance of knowing what the future might bring and the need for honesty from clinicians
  - wanted information about end-of-life planning

*“Oh, just to listen to the patient and their concerns about the future.” (P4, IPF)*

*“Well I haven’t got a very long future so ah, I suppose I’d like a bit of honesty from them . . . I’ve been sort of kept in the dark a little.” (P1, IPF)*

*“One other thing I am interested in is when you reach the end stage and you’re struggling to breathe and all these things, what can be done about it to reduce my anxiety level? No one has talked to me about that.” (P7, IPF)*

# CaNoPy Study

- Implications for clinical practice
  - Outside clinics
    - Early capture of changes in care needs between visits
    - Care needs of those living alone
  - Triggers for focused, early interventions
    - Initiation of Oxygen therapy
    - Changes in breathlessness and cough
    - Early changes in physical and social changes

# What do IPF patients value most?

## Invisible suffering in IPF: Unmet needs

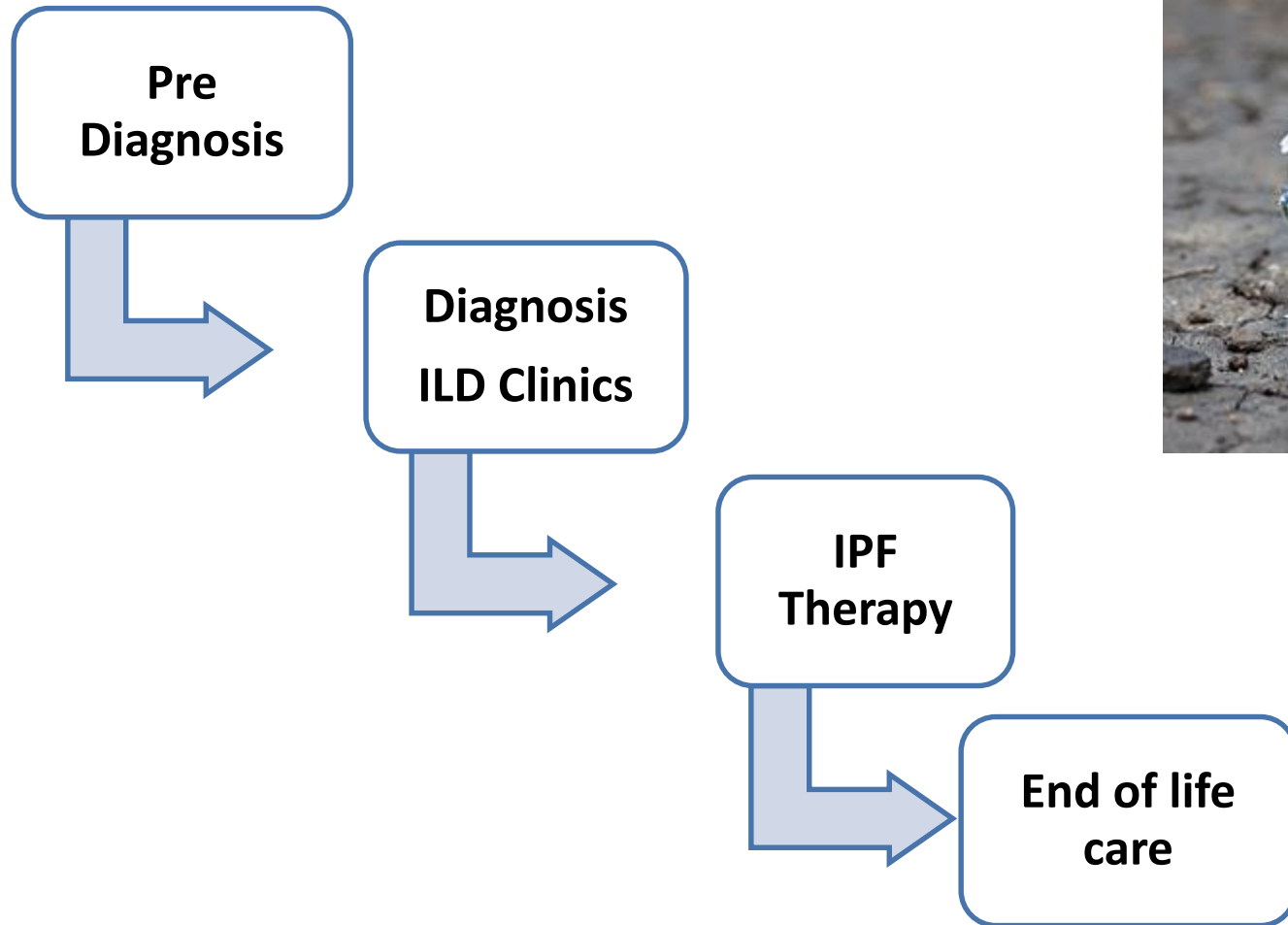
- IPF patients have a poor quality of life<sup>1-4</sup>
  - High prevalence of symptom burden in IPF<sup>1-4</sup>
    - Across disease spectrum
    - Particularly at end of life
  - Palliative care needs are largely unmet<sup>5</sup>
- Effective care must include strategies beyond just drugs!

1. Raghu G, et al. Lancet Respir Med 2014;2:566–72.
2. King TE Jr, et al. N Engl J Med 2014;370: 2083–92.
3. Richeldi L, et al. N Engl J Med 2014;370:2071–82.
4. Nathan SD, et al. Chest 2011;140: 221-229.
5. Ahmadi Z, et al. Thorax 2016;0:1–7.



# IPF Journey

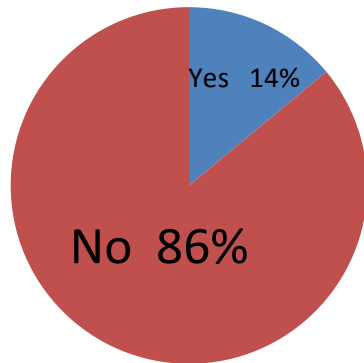
Patient and caregiver experience



# Palliative Care and Location of Death in Decedents With Idiopathic Pulmonary Fibrosis

*Kathleen O. Lindell, PhD, RN; Zhan Liang, MSN, RN; Leslie A. Hoffman, PhD, RN; Margaret Q. Rosenzweig, PhD, FNP-BC, AOCNP; Melissa I. Saul, MS; Joseph M. Pilewski, MD; Kevin F. Gibson, MD; and Naftali Kaminski, MD*

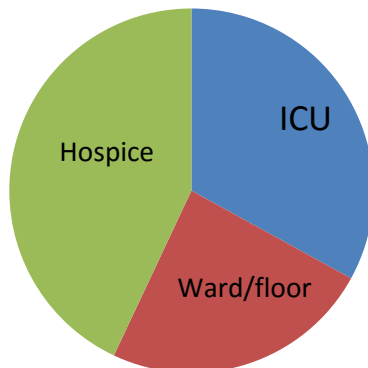
## IPF Symptom based care



## Timing of the palliative consult

**71% Last month**  
**18% Last 6 months**  
**11% Last 12 months**

## IPF Deaths



## Location of death

**No patients died at home- preferred place of death**  
**40% died in ICU**  
**17% wards**

# What do IPF patients value most?

## Invisible suffering in IPF: Unmet needs

- IPF patients have a poor quality of death<sup>1-5</sup>
  - Lack of good symptom control
  - Lack of advance care planning : preparation, moment of death etc
  - Lack of whole person care
  - Lack of adequate family engagement
  - Preponderance of hospital/ICU deaths

1. Raghu G, et al. Lancet Respir Med 2014;2:566–72.

2. King TE Jr, et al. N Engl J Med 2014;370: 2083–92.

3. Richeldi L, et al. N Engl J Med 2014;370:2071–82.

4. Nathan SD, et al. Chest 2011;140: 221-229.

5. Ahmadi Z, et al. Thorax 2016;0:1–7.

# The care needs of patients with idiopathic pulmonary fibrosis and their carers (CaNoPy): results of a qualitative study

- Findings regarding caregivers:
  - Expressed feelings of not being prioritized
  - Feeling unsure how to help
  - Expressed fears about coping with domestic situation in the future
  - Need to advance care planning
  - Better acknowledgement of their role and needs
  - Clinician's need to change attitude re caregivers as passive observers to active participants
  - Expressed ambiguity in how they were perceived with negative impact on both partners

# What IPF patients wants and what are the needs to be considered?

## Pre Diagnosis

- Early diagnosis
- Early referral to ILD centers

## Diagnosis ILD Clinics

- Patient centric vs disease centric focus
- Multidisciplinary teams
- Dedicated ILD nurse

## IPF care

- Fair and equitable access to therapies
- Focus on symptom based therapy
- Education, empowerment through self efficacy
- Care giver engagement
- Early detection of changes in between clinic visits

## End of life care

- Early and Integrated palliative care
- Advance care planning
- Symptom relief, family engagement
- Facilitate home/hospice deaths

# What IPF patients wants and what are the needs to be considered?

