Prise en charge globale du patient dans la Fibrose interstitielle diffuse

Comprehensive care for patients with pulmonary fibrosis

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- All fees were paid to my institution

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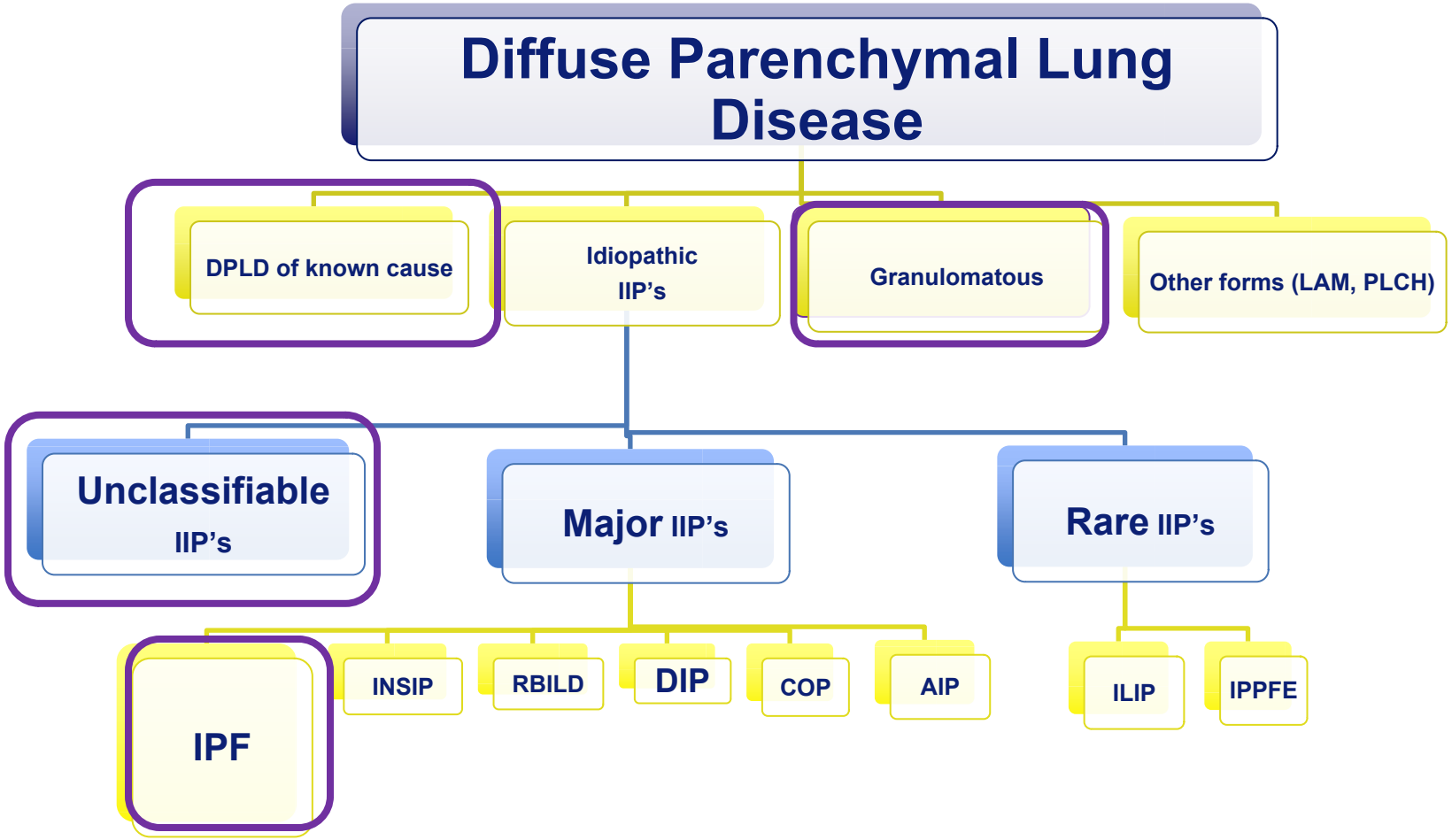
- **The impact of disease:** patient and partner's needs
- **Therapeutic needs and palliative care go hand in hand**
- **Holistic approach to ILD care:** ABCDE of ILD care

Many Interstitial Lung Diseases show fibrosis



ATS/ERS Update Classification of diffuse parynchymal lung diseases 2013

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IPF IS A CHRONIC PROGRESSIVE AND DEADLY DISEASE

Increasing symptoms

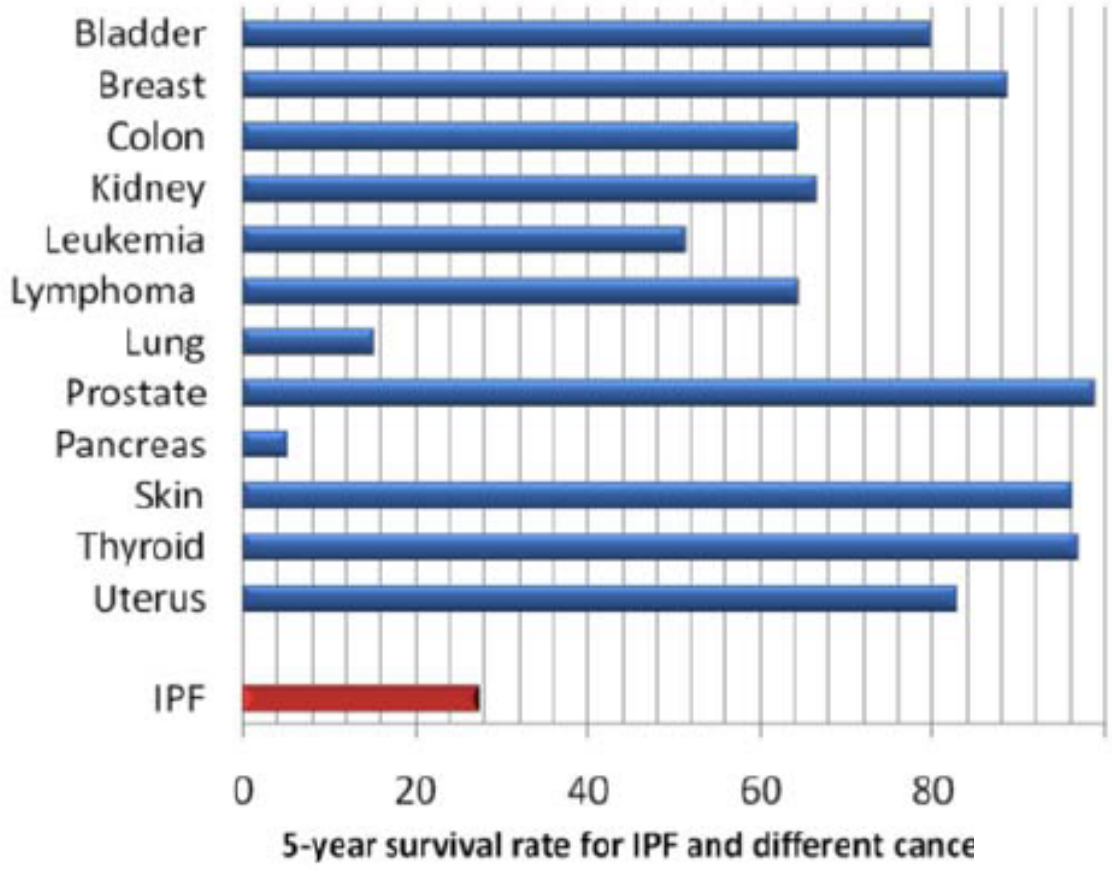
- Shortness of breath
- Fatigue
- Cough

The Voice of the Patient

A series of reports from the U.S. Food and Drug Administration's (FDA's) Patient-Focused Drug Development Initiative

Idiopathic Pulmonary Fibrosis

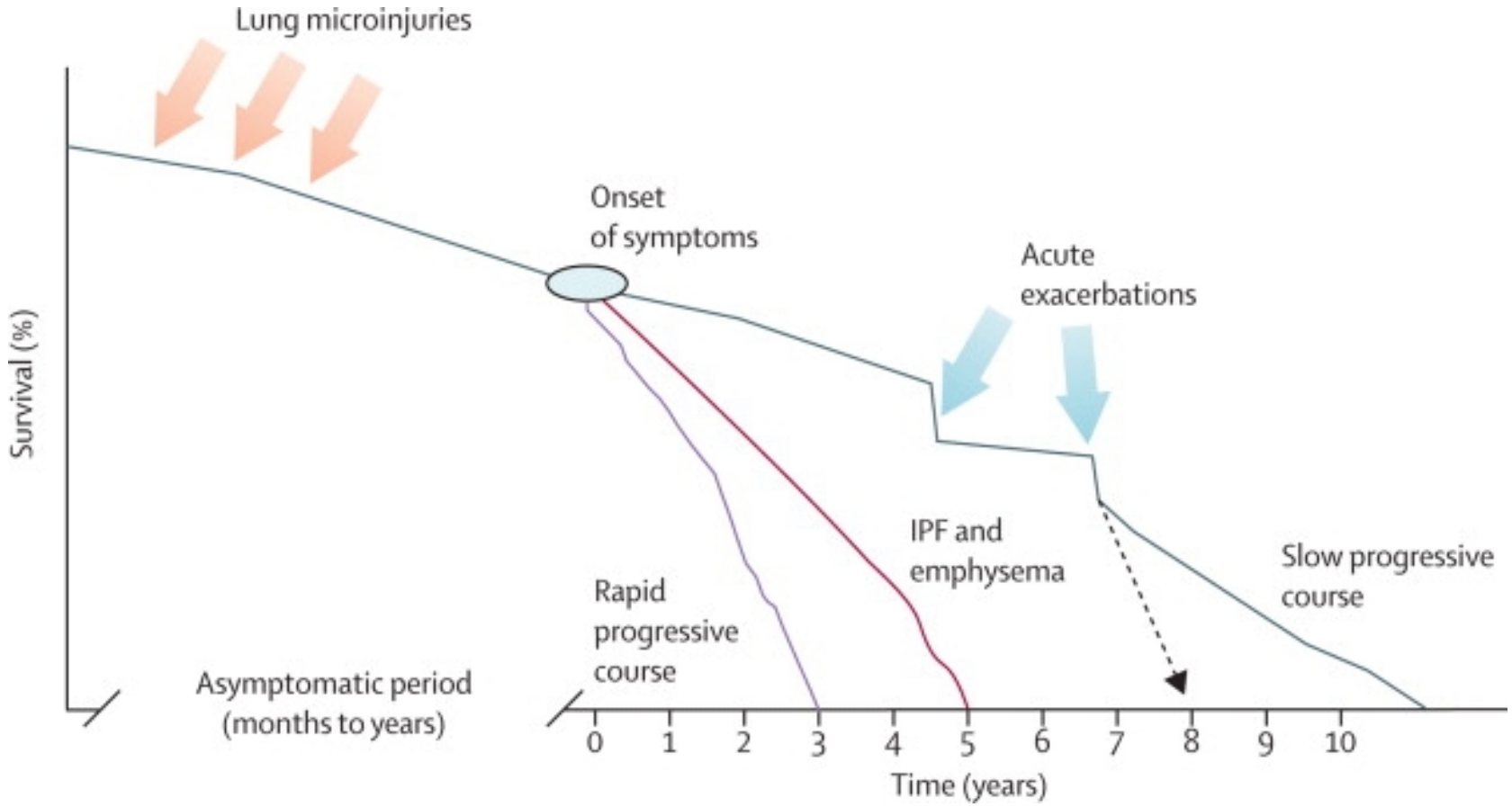
More lethal than many cancers



Vancheri ERJ 2011

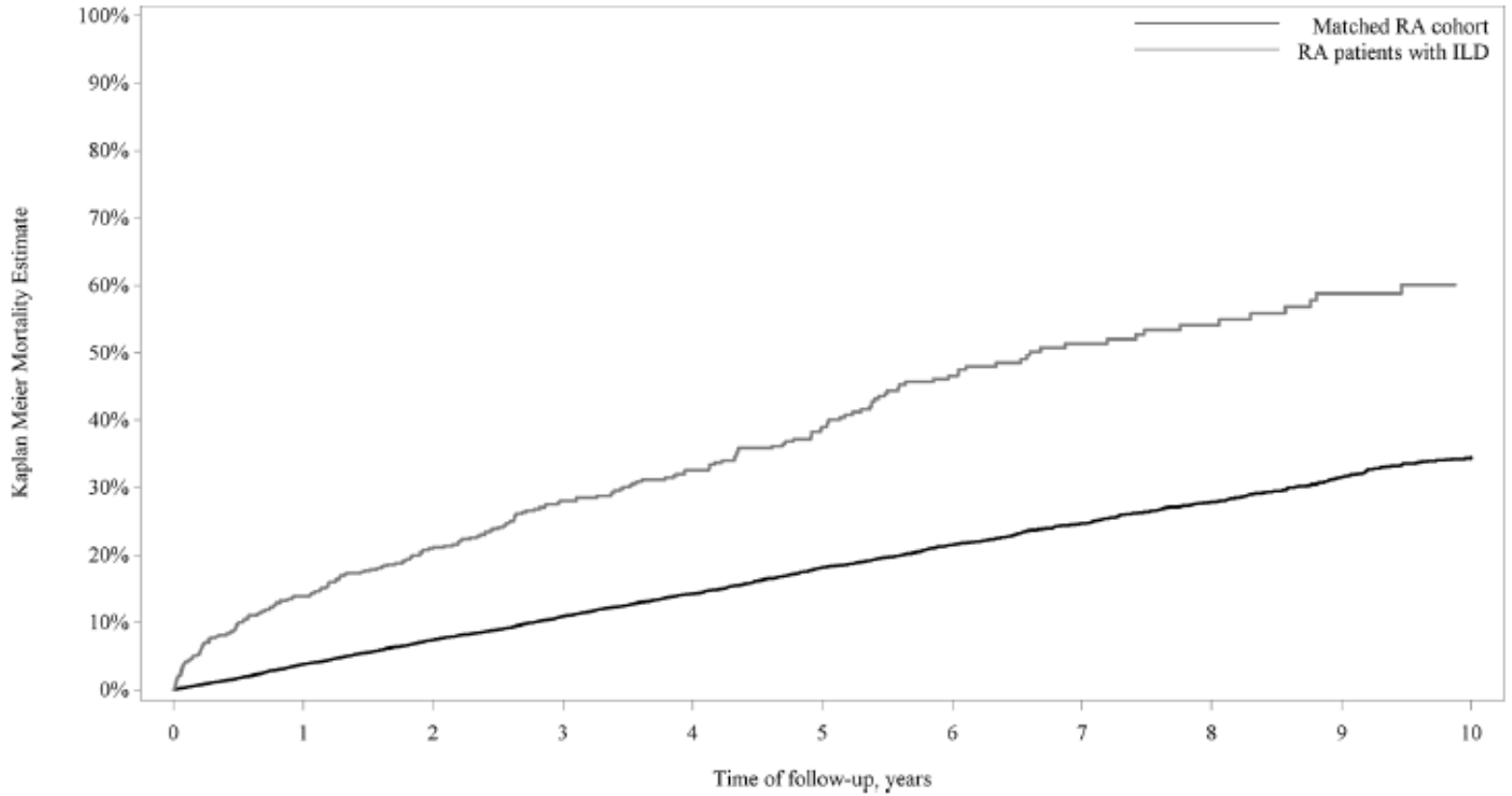
http://www.fda.gov/downloads/ForIndustry/UserFees/PrescriptionDrugUserFee/UCM440829.pdf

DISEASE DIFFERS FROM PATIENT TO PATIENT



King Lancet 2011

Many other ILD's show a similar progressing fibrotic phenotype and also impaired survival



Participants Among patients with RA diagnosed between 2004 and 2016, 679 patients with RA-ILD were matched for birth year, gender and age at RA diagnosis with 11 722 patients with RA but without ILD.

One-year mortality was 13.9% (95% CI, 11.4% to 16.7%) in RA-ILD and 3.8% (95% CI, 3.5% to 4.2%) in non-ILD RA,

Hyldgaard C, et al. Ann Rheum Dis 2017;76:1700-6.

Assayag D, et al. Radiology 2014;270:583-8.

Winstone TA, et al. Chest 2014;146:422-36.

Patterson KC, Strek ME. Ann Am Thorac Soc 2013;10:362-70.

Salisbury ML, et al. Am J Respir Crit Care Med 2017;196:690-699.

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Many other ILD's show a similar progressing fibrotic phenotype and also impaired survival and quality of life

Quality of life in IPF and in other ILD

	IPF (n=108) Mean (SD) N(%)	ILD, non IPF (n=68) Mean (SD) N(%)
K-BILD		
Total [0-100]	51.9 (22.2)	58.7(21.81)
SGRQ		
Total [0-100]	48.9 (20.9)	41.7 (20.8)
EQ-5D-5L		
Index value[-0.329-1.00]	0.66 (0.23)	0.74 (0.19)
Lung function		
FVC % predicted n= 163	72.5 (16.8)	79.1 (20.2)
FEV1 %predicted n=162	75.8 (17.6)	72.9 (19.6)
TLC % predicted n=125	62.3 (13.3)	72.3 (18.0)
TLCOc % predicted n=139	47.1 (15.2)	59.5 (19.2)

Wapenaar et al Chron Respir Dis. 2017 May;14(2):140-150.

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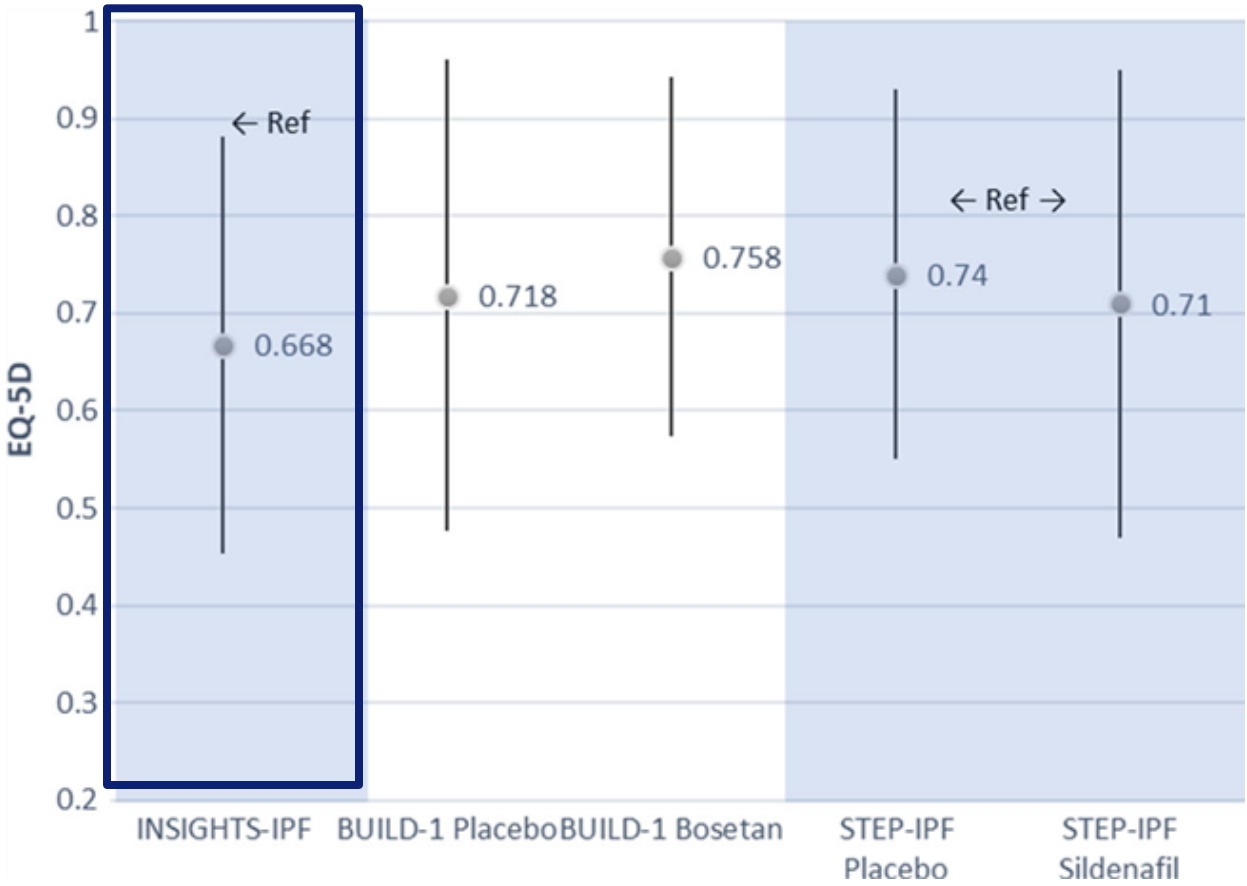
IPF has considerable impact on HRQOL relative to general population levels

Our cohort ILD 0.74
Our cohort IPF 0.66



EuroQol 5-level questionnaire (EQ-5D) in patients with IPF compared with the general population (reference). The lowest and highest of the available intervals are shown in the figure

Diamatopoulos A, et al. The Burden of Illness of Idiopathic Pulmonary Fibrosis: A Comprehensive Evidence Review. PharmacoEconomics 2018

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Quality of life in IPF in clinical practice

German registry
623 Patient with IPF

More severely impaired HRQOL
Mean SGRQ score 48.3
(INPULSIS 39.4 /39.8)

Mean USCD score 47.8
(ASCEND 34.0-36.6)

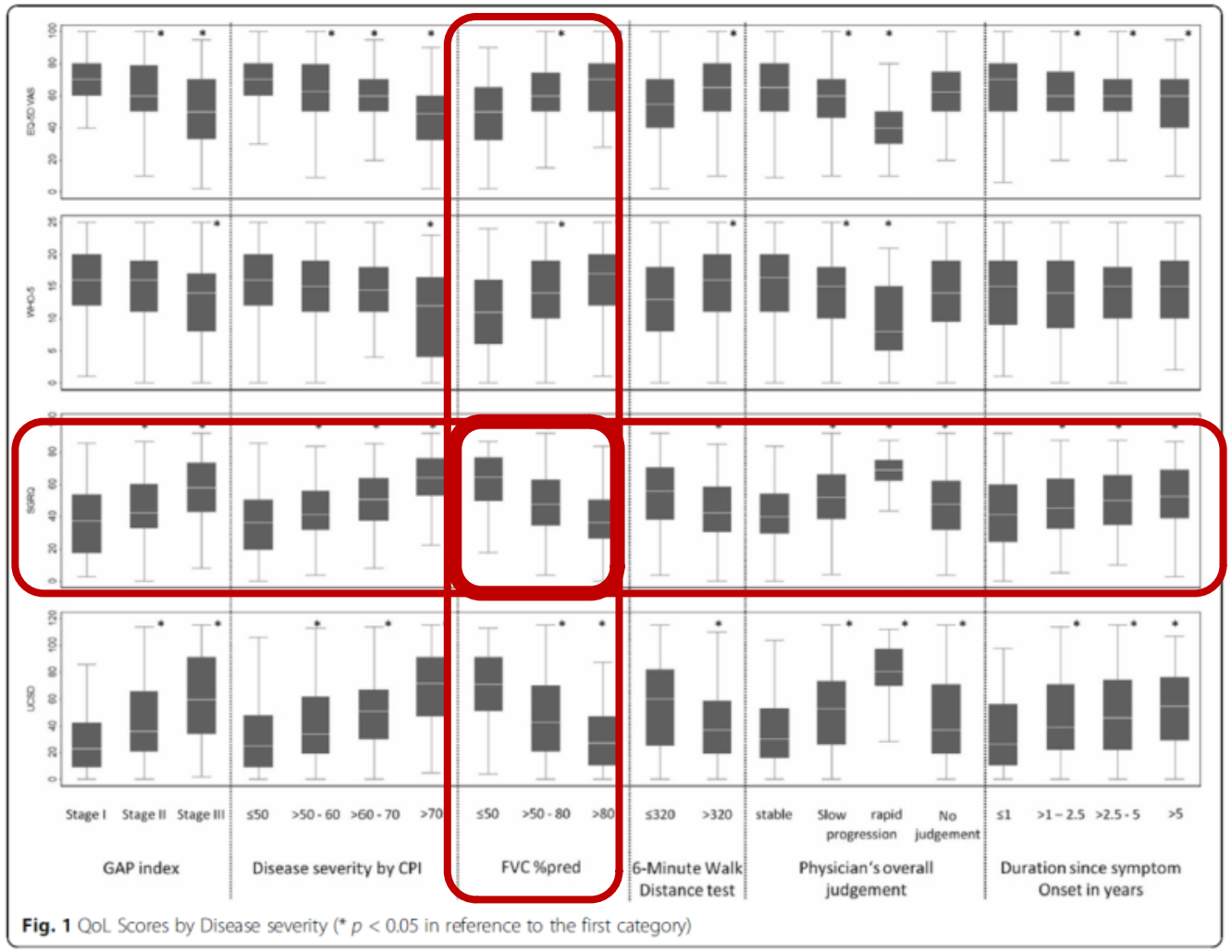

Fig. 1 QoL Scores by Disease severity (* $p < 0.05$ in reference to the first category)

Categorical relationship between QOL and physiological parameters

HRQO, health related quality of life. SGRQ: St George Respiratory Questionnaire. UCSD: University of California Shortness of breath scale. EQ-5d Euroqol 5-level questionnaire

Kreuter M et al. HRQOL in patients with idiopathic pulmonary fibrosis in clinical practice. Respir Res 2017; 18(1): 139.

Physiological outcomes do not correlate well with patient reported outcome measures

Correlation K-BILD and lung function tests

	Lung function		
K-BILD	FEV_1%	FVC%	TLco%
Psychological	0.46	0.38	0.45
Breathlessness and activity	0.55	0.51	0.52
Chest symptoms	0.48	0.45	0.42
Total	0.53	0.47	0.50

Correlation SGRQ and SGRQ-I and pulmonary physiology

	Symptoms		Activity		Impacts		Total	
	SGRQ	SGRQ-I	SGRQ	SGRQ-I	SGRQ	SGRQ-I	SGRQ	SGRQ-I
FVC%	-0.27*	-0.25†	-0.31†	-0.30†	-0.30†	-0.31†	-0.34†	-0.33†
TLco%	-0.23*	-0.25†	-0.34†	-0.33†	-0.38†	-0.36†	-0.38†	-0.37†
6MWD	-0.14	-0.12	-0.32†	-0.30†	-0.24*	-0.26†	-0.28*	-0.28*

1. Patel AS *et al.* The development and validation of the King's Brief Interstitial Lung Disease (K-BILD) health status questionnaire. Thorax 2012;67:804-10
2. Yorke J *et al.* Quantification of dyspnoea using descriptors: development and initial testing of the Dyspnoea-12. Thorax 2010;65:21-26

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Besides lungfunction, dyspnea, cough and depression are major drivers of quality of life

Australian registry shows that there is a strong association between dyspnea, cough, depression and health related quality of life

Table 4 Multivariate predictors of health-related quality of life

Variable	Regression coefficient	Standard error	*P*-value	Partial R^2	Model R^2
UCSD-SOBQ	0.431	0.043	<0.0001	0.709	0.709
Cough VAS	0.202	0.045	<0.0001	0.057	0.766
HADS-D	1.427	0.332	<0.0001	0.036	0.802

HADS, Hospital Anxiety and Depression Scale; UCSD-SOBQ, University of California San Diego Shortness of Breathlessness Questionnaire; VAS, visual analogue scale.

*Health related quality of life was assessed with the St George Respiratory Questionnaire

Glaspole IN et al. Health-related quality of life in idiopathic pulmonary fibrosis: Data from the Australian IPF Registry. Respirology 2017; 22(5): 950-6.

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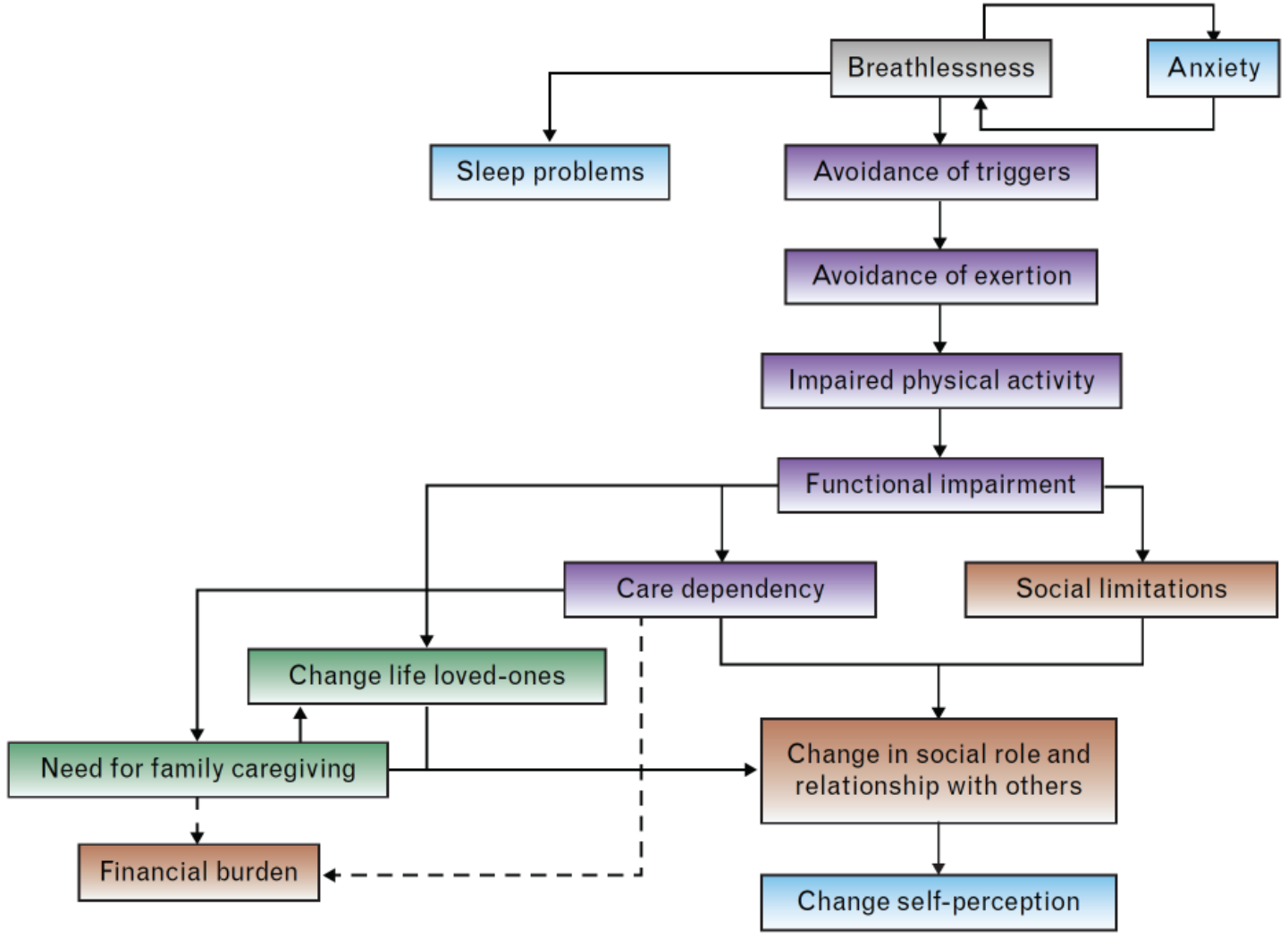
Breathlessnes may have major behavioural, psychological and social consequences

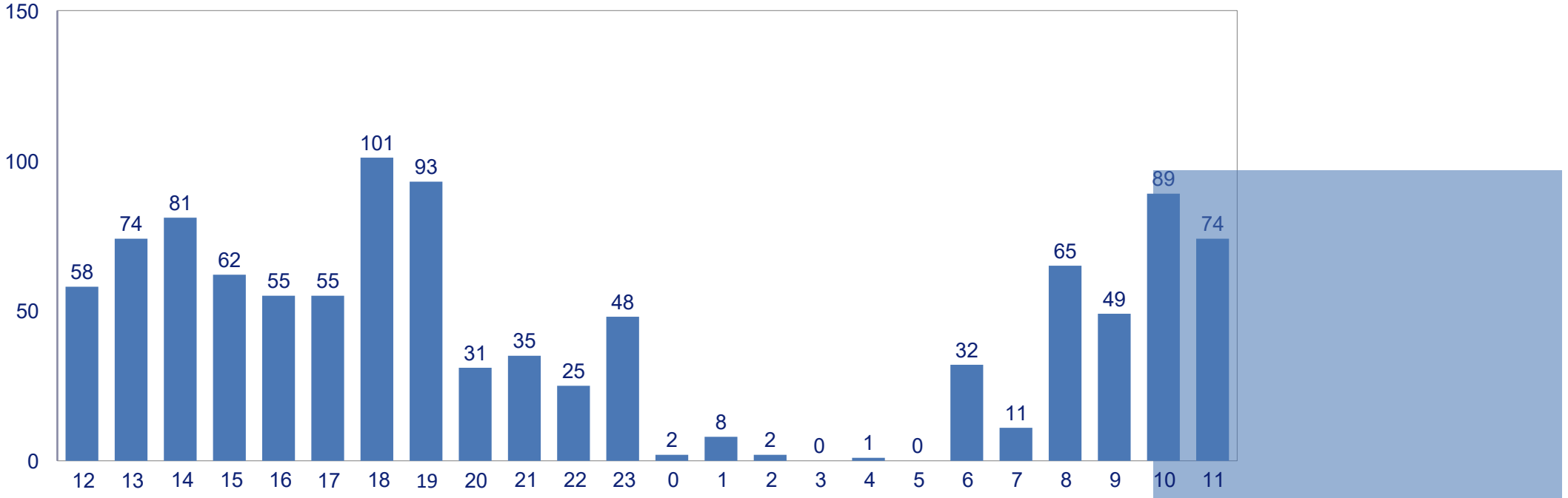


Janssen DJ et al . Psychosocial consequences of living with breathlessness due to advanced disease. Curr Opin Support Palliat Care 2015;9:232-37

Cough in IPF

Cough measurements

Median 24-hour cough-count 520 (91-3394)



In other study patiens with IPF: Average cough 226 /day (36- 946)

Cough independent predictor of disease progression

van Manen MJ et al. Effect of pirfenidone on cough in patients with idiopathic pulmonary fibrosis Eur Respir J 2017;50(4)
Cheng JZ et al. Cough is less common and less severe in systemic sclerosis-associated interstitial lung disease compared to other fibrotic interstitial lung diseases . Respirology 2017; 22(8): 1592-7.
Key AL et al. Objective cough frequency in Idiopathic Pulmonary Fibrosis. Cough 2010;6:4-10
Ryerson CJ et al. Cough predicts prognosis in idiopathic pulmonary fibrosis. Respirology 2011; 16(6): 969-75.

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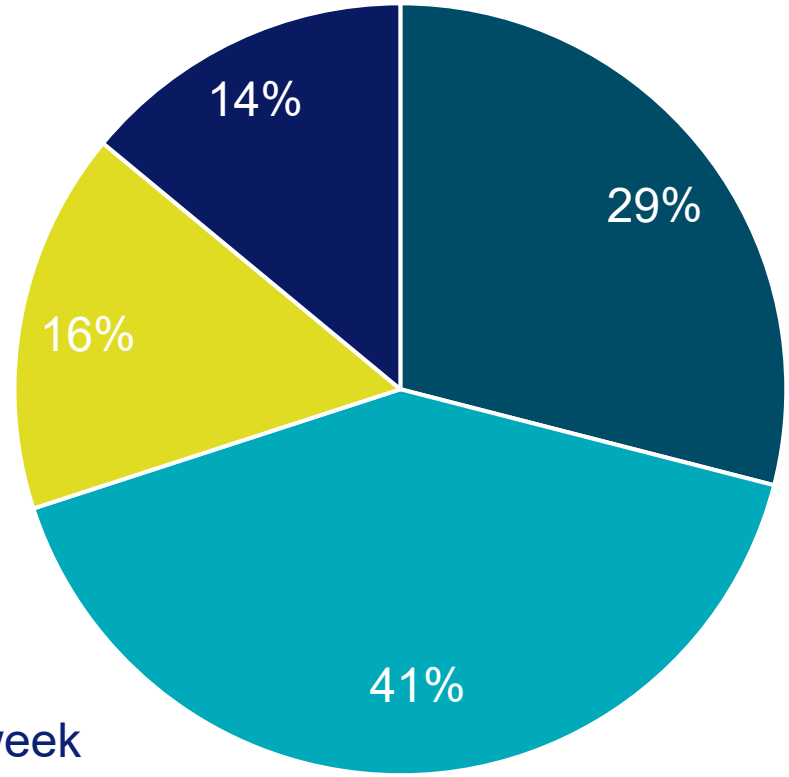
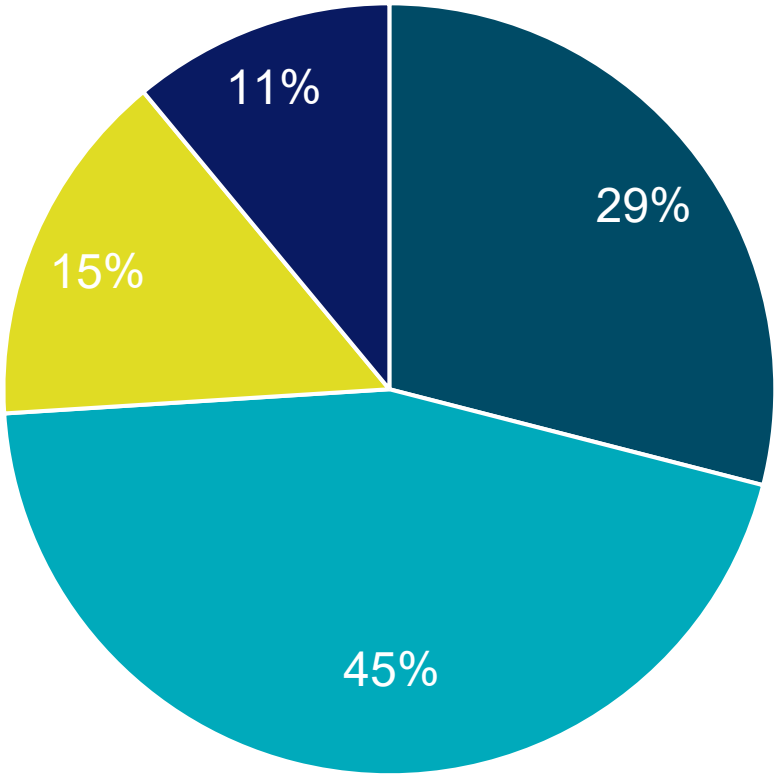
General Anxiety and Distress Score (GAD-1) in patient with IPF and their partners

How often have you been able to relax in the last 2 weeks (patient) ?



n=114

How often have you been able to relax in the last 2 weeks (partners) ?



n=105

- Not at all
- Several days
- More than half of the week
- Almost every day

van Manen MJ et al. What patients with pulmonary fibrosis and their partners think: a live, educative survey in the Netherlands and Germany. ERJ Open Res 2017; 3(1)
Donker T et al. Quick and easy self-rating of Generalized Anxiety Disorder: validity of the Dutch web-based GAD-7, GAD-2 and GAD-SI. Psychiatry Res 2011;188:58-64

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Fatigue, no good studies on prevalence

In Nintedanib trials Fatigue in < 10% of patients placebo arm and treatment arm

Table 2
Adverse events: Pooled data from the TOMORROW and INPULSIS® trials.

N (%)	Nintedanib 150 mg bid (n = 723)	Placebo (n =
Any adverse event(s)	689 (95.3)	456 (89.8)
Most frequent adverse events[a]		
Diarrhoea	445 (61.5)	91 (17.9)
Nausea	176 (24.3)	36 (7.1)
Nasopharyngitis	93 (12.9)	79 (15.6)
Cough	93 (12.9)	75 (14.8)
Vomiting	85 (11.8)	15 (3.0)
Decreased appetite	81 (11.2)	24 (4.7)
Bronchitis	76 (10.5)	56 (11.0)
Progression of IPF[b]	68 (9.4)	72 (14.2)
Upper respiratory tract infection	65 (9.0)	55 (10.8)
Dyspnoea	55 (7.6)	59 (11.6)
Severe adverse event(s)	193 (26.7)	119 (23.4)
Serious adverse event(s)	217 (30.0)	153 (30.1)
Fatal adverse event(s)	38 (5.3)	43 (8.5)
Adverse event(s) leading to treatment discontinuation[c]	149 (20.6)	76 (15.0)
Diarrhoea	38 (5.3)	1 (0.2)
Nausea	17 (2.4)	0 (0.0)
Progression of IPF[a]	15 (2.1)	27 (5.3)
Decreased appetite	11 (1.5)	1 (0.2)
Weight decreased	8 (1.1)	1 (0.2)
Abdominal pain	7 (1.0)	1 (0.2)
Vomiting	7 (1.0)	1 (0.2)
Pneumonia	6 (0.8)	5 (1.0)

Treated set (patients treated with ≥1 dose of trial drug).
[a] Adverse events reported by >10% of patients in either treatment group.
[b] Corresponds to the MedDRA term 'IPF', which included disease worsening and IPF exacerbations.
[c] Adverse events leading to treatment discontinuation in ≥1% of patients in either treatment group by MedDRA preferred term.

In Pirfenidone trials Fatigue in 19% of patients placebo arm and 26% treatment arm

Table 3 Treatment-emergent adverse events*

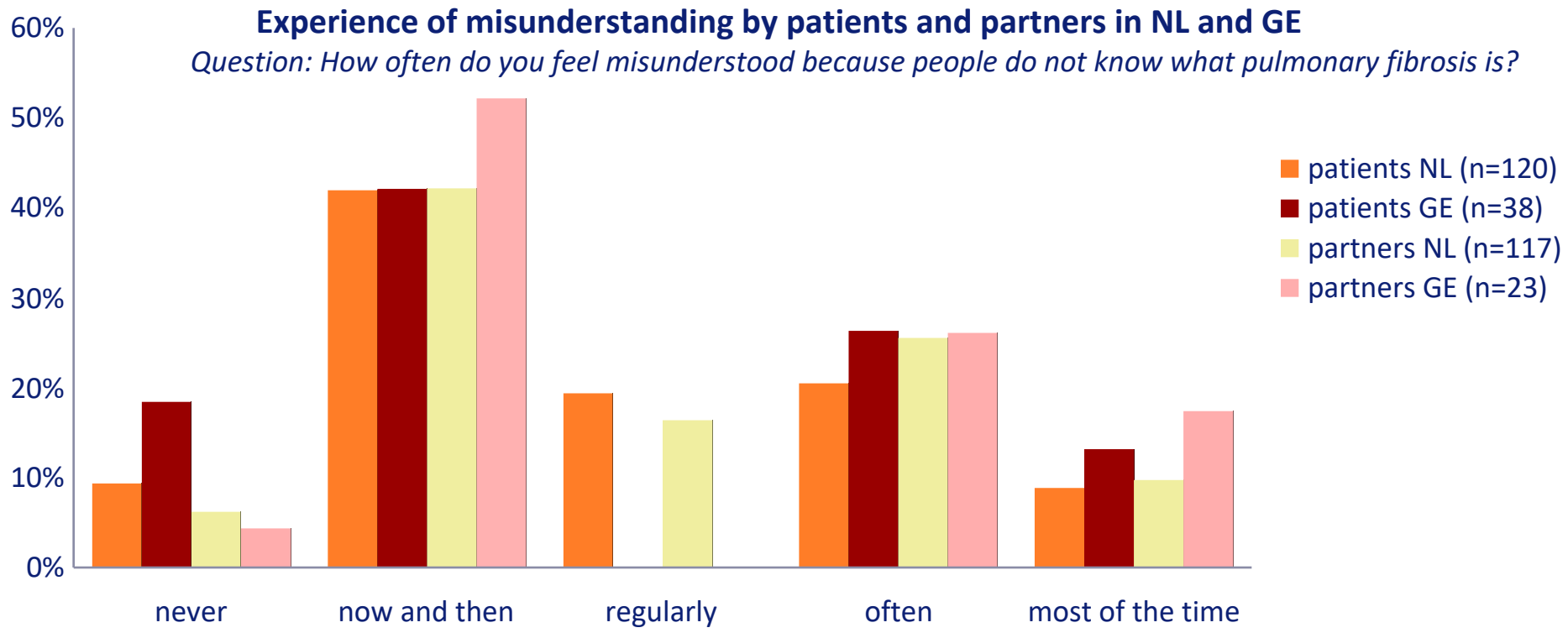
		Phase 3 multinational trials‡	
	Integrated population (N=1299)†	Pirfenidone (N=623)	Placebo (N=624)
Duration of exposure, median (range), years	1.7 (>0, 9.9)	1.0 (>0, 2.3)	1.0 (>0, 2.3)
Treatment-emergent adverse event, %			
Nausea	37.6	36.1	15.5
Cough	35.1	27.8	29.2
Dyspnoea	30.9	16.9	20.2
Upper respiratory tract infection	30.6	26.8	25.3
Idiopathic pulmonary fibrosis	29.3	13.0	19.9
Fatigue	28.2	26.0	19.1
Diarrhoea	28.1	25.8	20.4
Rash	25.0	30.3	10.3
Bronchitis	23.8	14.1	15.4
Headache	21.6	22.0	19.2
Nasopharyngitis	21.3	16.7	17.9
Dizziness	21.2	18.0	11.4
Dyspepsia	18.4	18.5	6.9
Vomiting	15.9	13.3	6.3
Weight decreased	15.6	10.1	5.4
Back pain	15.4	10.4	10.4
Anorexia	15.2	13.0	5.0

*Occurring in ≥15% of patients in the cumulative clinical database.
†Includes two patients in study 002 with a diagnosis of "pulmonary fibrosis."
‡CAPACITY (studies 004 and 006) and ASCEND (study 016).

Fatigue TEAEs tended to be more frequent yet shorter in duration in patients with IPF who received pirfenidone vs. placebo

Richeldi L et al. Nintedanib in patients with idiopathic pulmonary fibrosis: Combined evidence from the TOMORROW and INPULSIS trials. Respir Med 2016; 113: 74-9.
Lancaster L et al. Safety of pirfenidone in patients with idiopathic pulmonary fibrosis: integrated analysis of cumulative data from 5 clinical trials. BMJ Open Respir Res 2016; 3(1): e000105
Costabel U et al. Fatigue in patients with idiopathic pulmonary fibrosis (IPF) from the pooled pirfenidone (PFD) Phase III trials. [Abstract]. Eur Respir J 2017; 50: A4875

IPF is rare and unknown; patients and partners feel misunderstood



van Manen M et al. Eur Respir Research 2017

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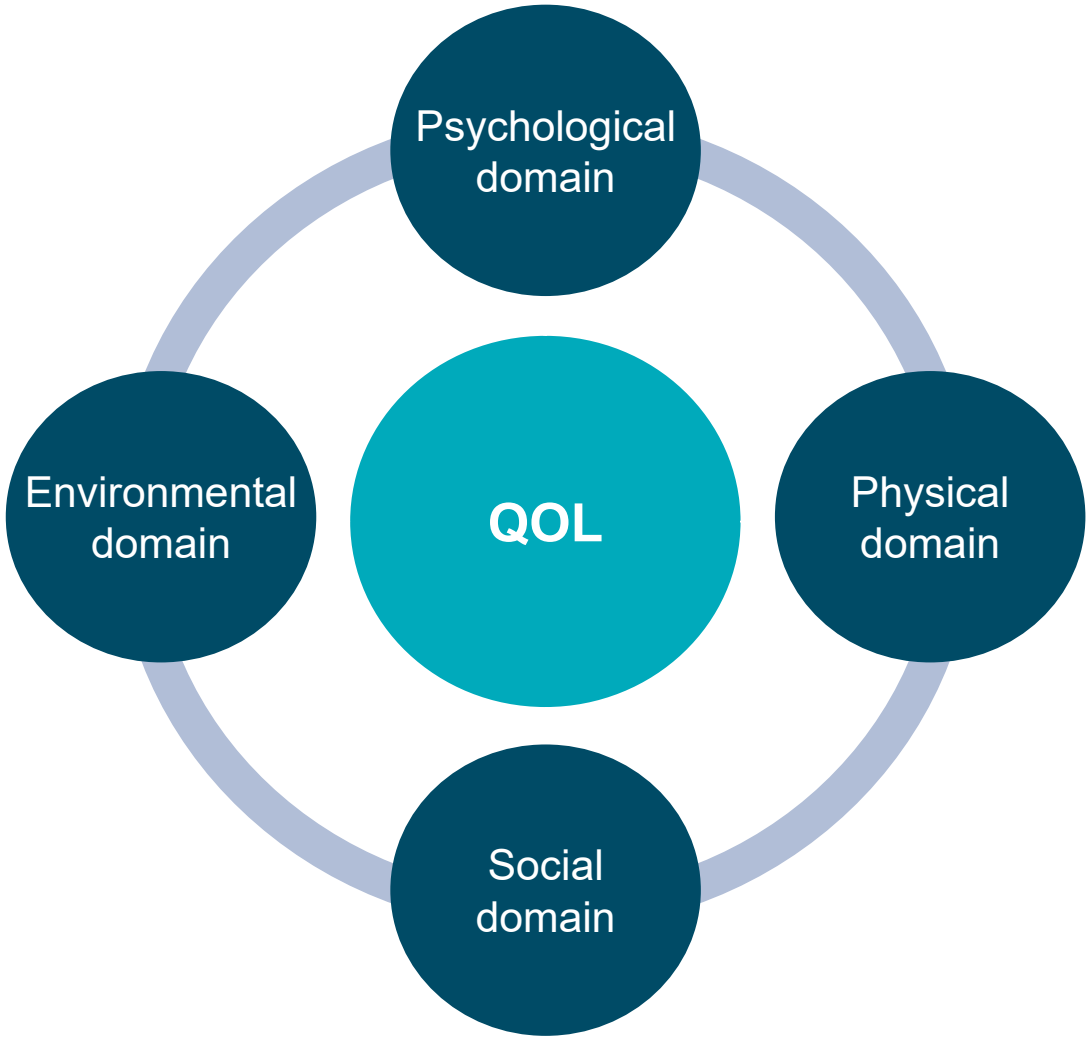
Determinants of quality of life are multiple and personal



Symptoms, reactions and perceptions interact

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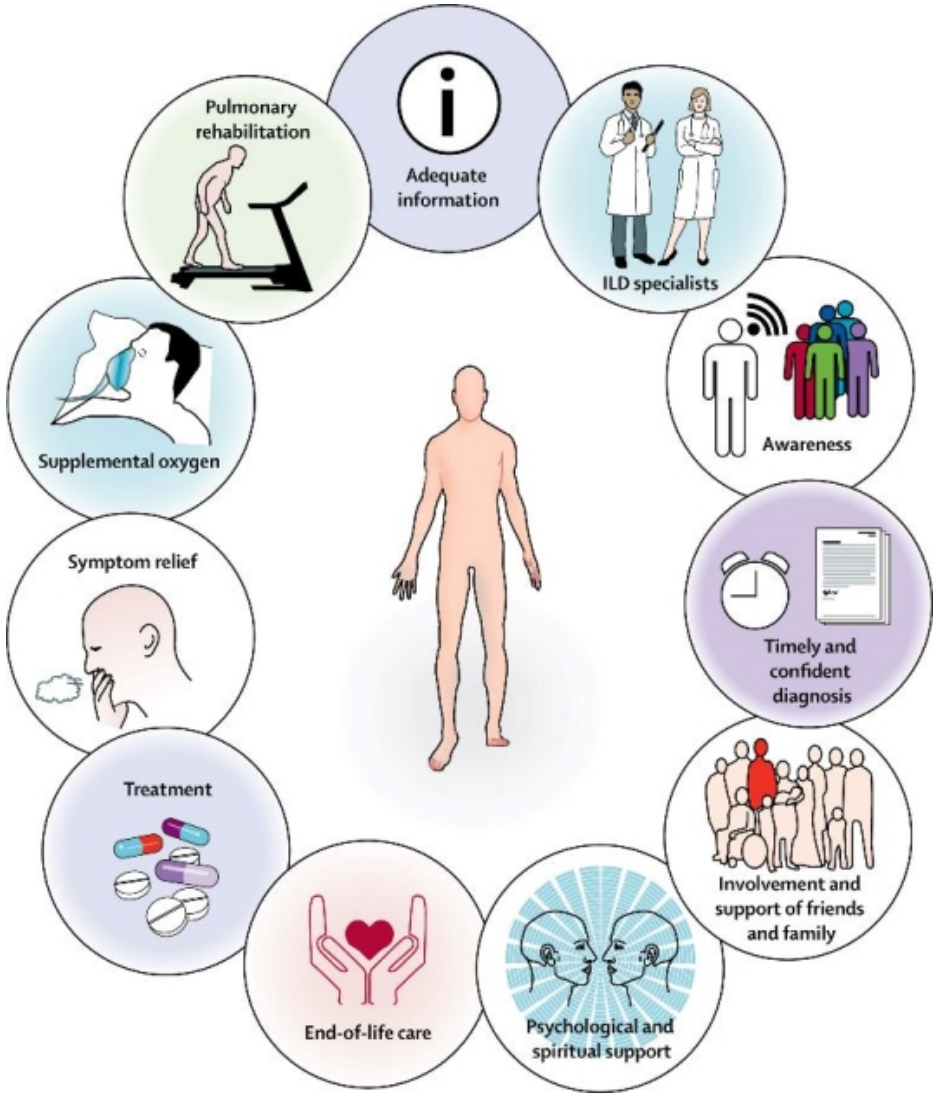
van Manen MJ et al. Optimizing quality of life in patients with idiopathic pulmonary fibrosis. Ther Adv Respir Dis 2017; 11(3)157-169

Is it possible to improve QoL in a progressive fibrosing ILD's ?

Needs for patients



In addition to disease-modifying treatment, complementary approaches are required to improve QoL or slow down deterioration in QoL

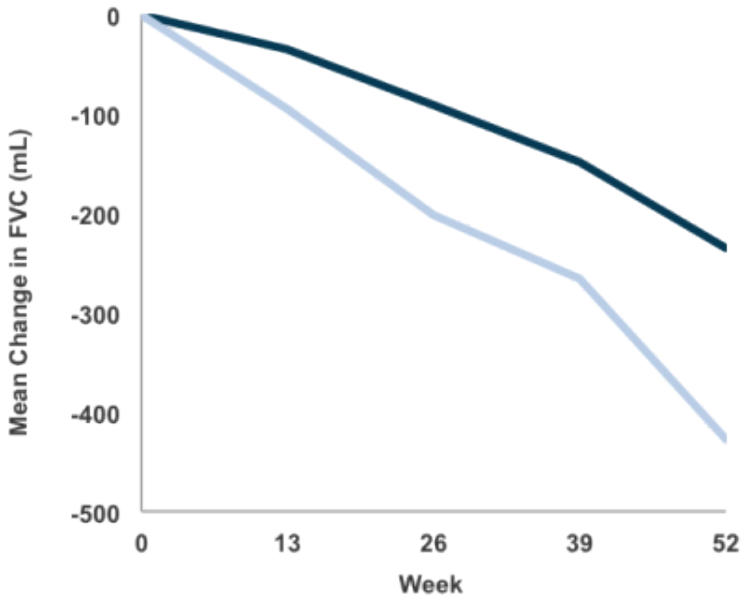
Kreuter M et al. Palliative care in interstitial lung disease: living well Lancet Respir Med 2017 Dec;5(12):968-980

- **The impact of disease:** patient and partner's needs
- **Therapeutic needs and palliative care go hand in hand**
- **Holistic approach to ILD care:** ABCDE of ILD care

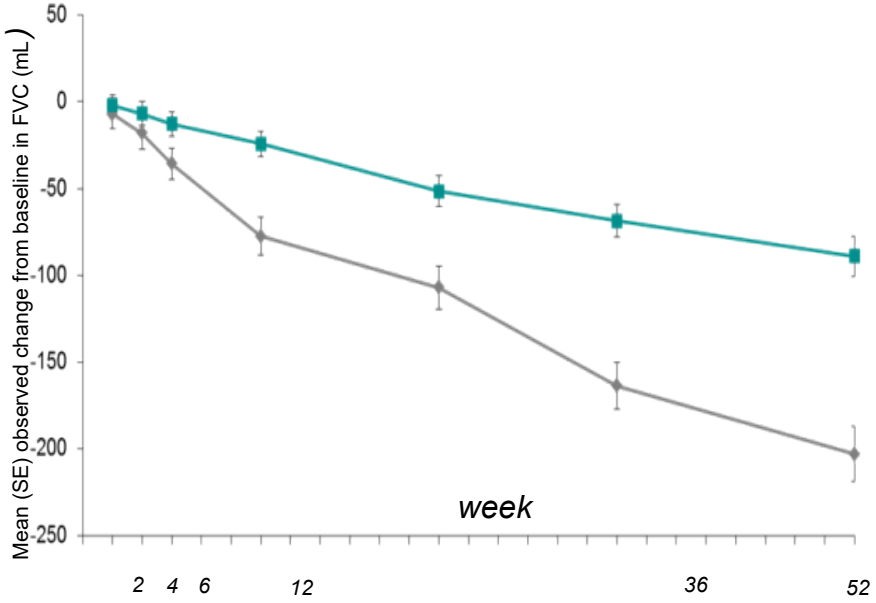
In a fatal progressive diseases, prolonging life at an acceptable quality is what most people strive for

SINCE 2014 TWO DRUGS THAT SLOW DOWN DISEASE DECLINE AND IMPROVE SURVIVAL

Pirfenidone : Mean Change in FVC Volume (mL)



Nintedanib: Mean Change in FVC Volume (mL)



A disease-centered model assumes that treating the disease treats the patient and thus improves quality and quantity of life

King TE et al. N Engl J Med 2014;370:2083-2092
Richeldi L et al. N Engl J Med 2014;370:2071-2082

No convincing effect of treatment on health related quality of life in the positive IPF trials

Trial	Treatment	PRO	Outcome
CAPACITY 1&2 (Noble 2011)	Pirfenidone	UCSD SGRQ WHO-QOL	ns ns ns
BIBF-1120 (Richeldi 2011)	Nintedanib	SGRQ	150 mg bid p<0.01
ASCEND (King 2014)	Pirfenidone	UCSD	ns
INPULSIS (Richeldi 2014)	Nintedanib	SGRQ	ns, INPULSIS 2 p=0.02

However, in pirfenidon and ninterdanib trials there was a positive effect on respectively UCSD-SOB and SGRQ in more severe groups

Noble PW et al. Pirfenidone for idiopathic pulmonary fibrosis: analysis of pooled data from three multinational phase 3 trials. Eur Respir J 2016;47:243-53
Richeldi et al. Efficacy of a tyrosine kinase inhibitor in idiopathic pulmonary fibrosis. N Engl J Med 2011; 365(12): 1079-87
King TE et al. A phase 3 trial of pirfenidone in patients with idiopathic pulmonary fibrosis. N Engl J Med 2014; 370(22): 2083-92.
Richeldi et al. Efficacy and safety of nintedanib in idiopathic pulmonary fibrosis. N Engl J Med 2014; 370(22): 2071-82.

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Palliative care should be explained to patients

Patients were asked "What does palliative care mean to you?"

- "...I fear it to some extent because I don't know exactly what it means..."
- "It's a bit scary—having someone explain might help"
- "...I would appreciate if somebody would talk to me about it..."
- "...Is palliative care not solely a euphemism for dying soon..."
- "...Before I was sent to the palliative care ward I was frightened to be demoted for dying but at the same time frightened to suffocate. Palliative care took away my breathlessness and my fears..."
- "...My main goal of therapy for my chronic disease is to maintain quality of life at the best achievable level for the time I have left..."
- "...I did not know that palliative care is reimbursed, many people think they have to pay for it themselves..."
- "...I do not want to be labelled as a doomed man - that´s why I do not like to be sent to palliative care..."
- "...Professional carers that understand the needs of patients with fibrosis and administer help that make patients feel better..."
- "...I would not like it but if I need it, I would take it..."
- "...Dignity, pain free and of great benefit to patients and families..."
- "Palliative care always meant end of life to me but if it helps my quality of life for whatever time I have left, I would be happy to look into it"

ATS definition of the goal of palliative care:

'to prevent and relieve suffering by controlling symptoms and to provide other support to patients and families in order to maintain and improve their quality of living'

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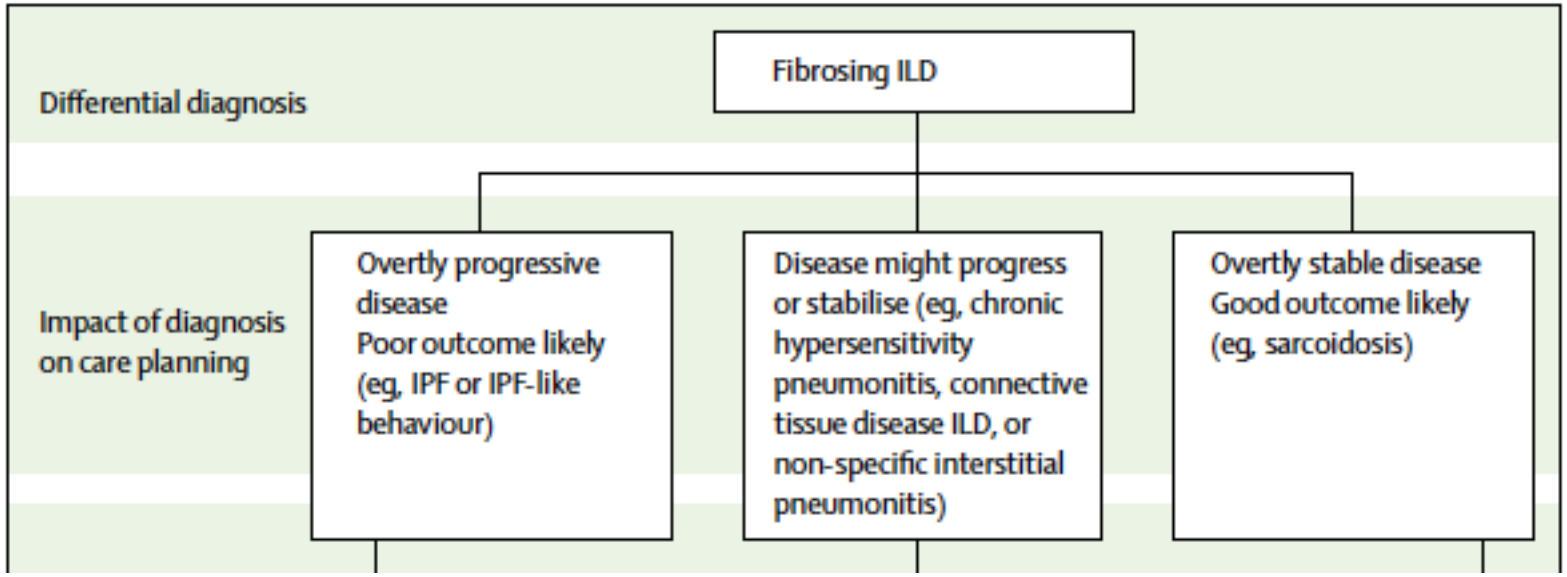
Kreuter M et al. Palliative care in interstitial lung disease: living well Lancet Respir Med 2017;5(12):968-980
Lanken PN et al. An official American Thoracic Society clinical policy statement: palliative care for patients with respiratory diseases and critical illnesses. Am J Respir Crit Care Med 2008; 177(8): 912-27.

Not all fibrosing ILD's are equal

timing and tailoring are key



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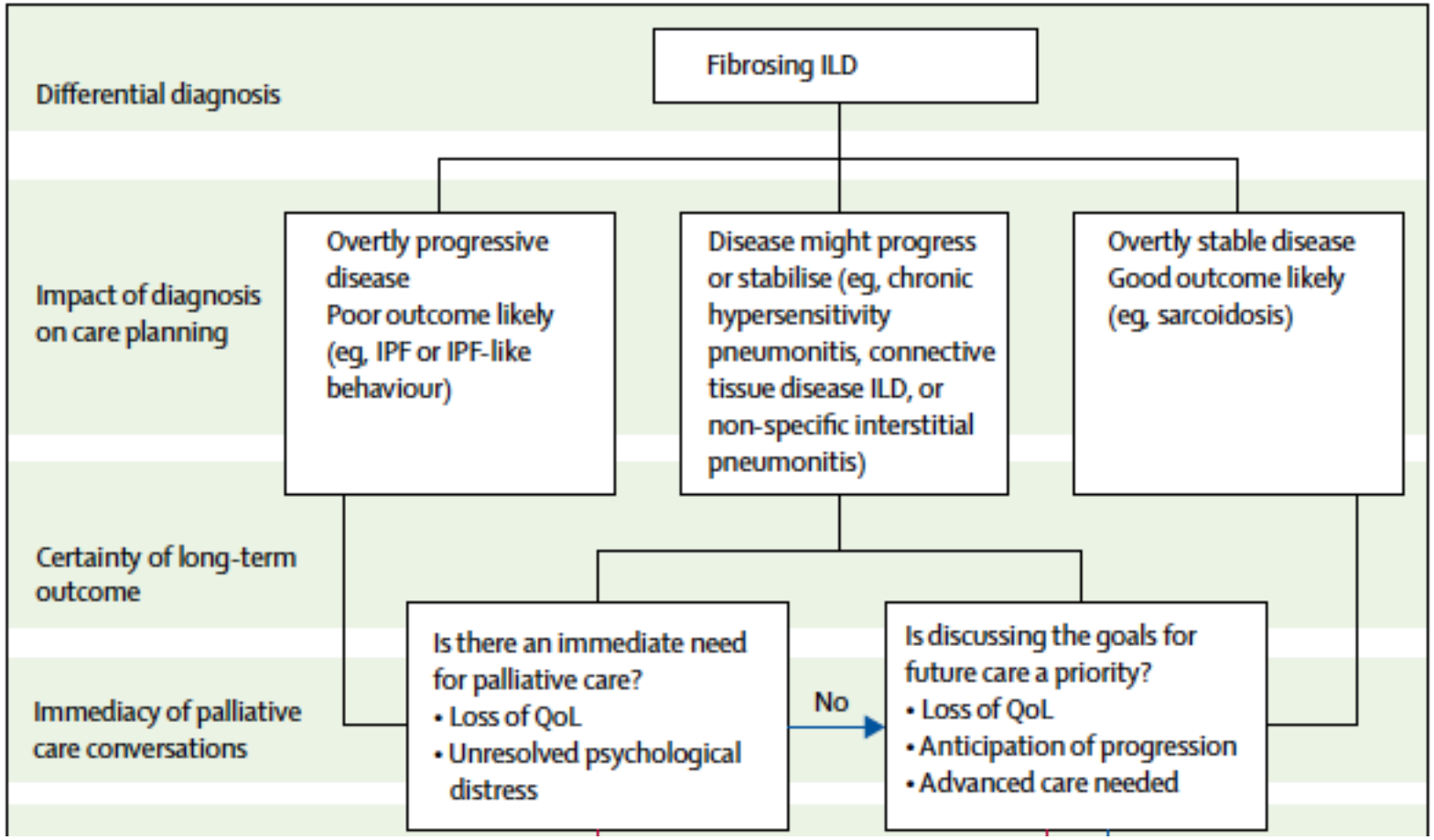

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Not all fibrosing ILD's are equal

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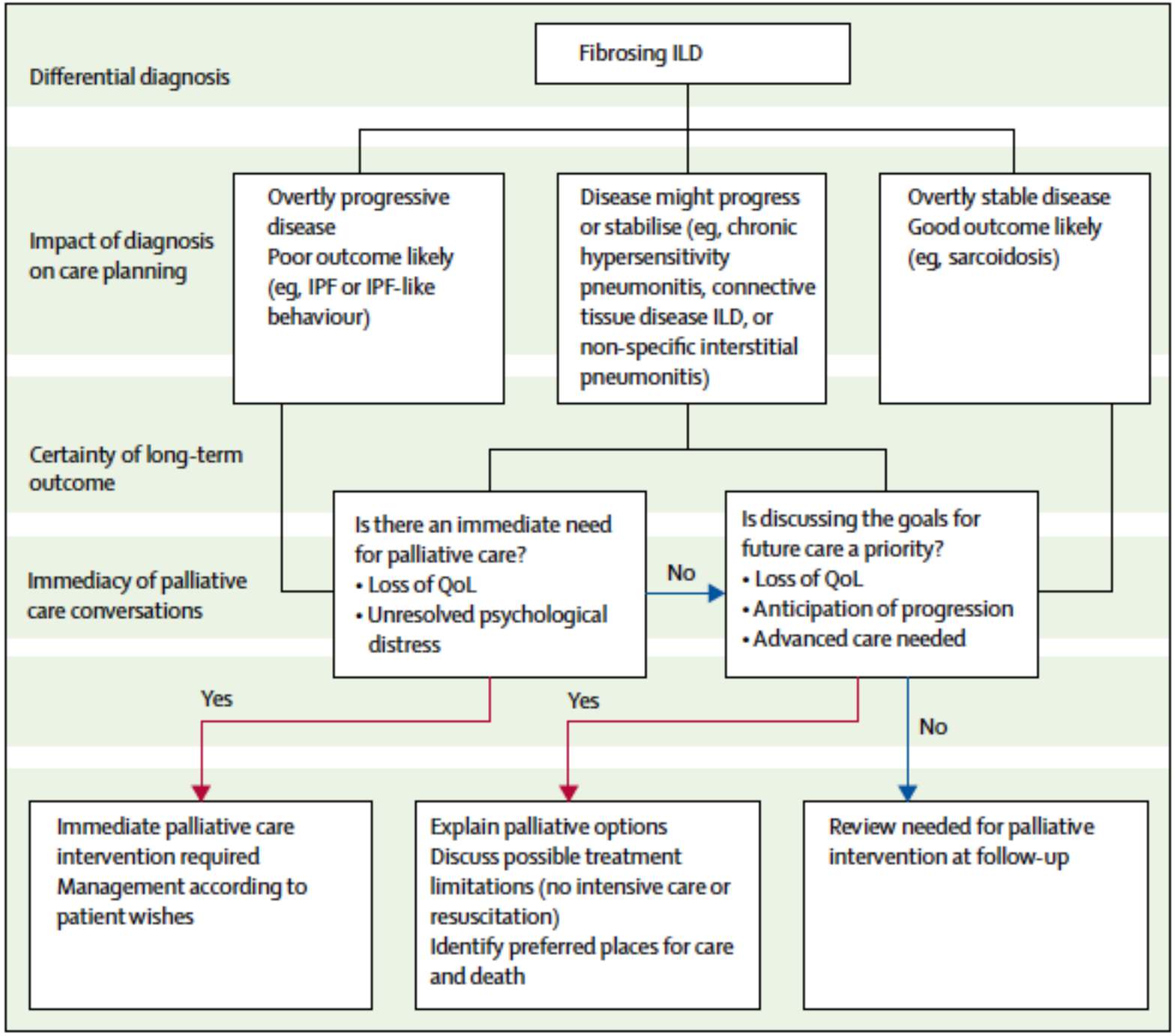

Kreuter M et al. Palliative care in interstitial lung disease: living well
Lancet Respir Med 2017;5(12):968-980

Management of dyspnea

- Treat, when possible, co-morbities that contribute to dyspnea
- Supplemental oxygen : small studies suggesting benefit
- Hand held fan
- Opioids
- Benzodiazepines
- Sildenafil?

Mc Donald . Exercise desaturation and oxygen therapy in ILD and COPD: Similarities, differences and therapeutic relevance. Respirology 2018 [Epub ahead of print].
Visca et al. AmbOx: A Randomised Controlled, Crossover Trial Evaluating the Effects of Ambulatory Oxygen on Health Status in Patients with Fibrotic Interstitial Lung Disease. [Abstract]. Am J Respir Crit Care Med 2017; 195: A7603.
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Johnson MJ et al. A Mixed-Methods, Randomized, Controlled Feasibility Trial to Inform the Design of a Phase III Trial to Test the Effect of the Handheld Fan on Patients With Refractory Breathlessness. J Pain Symptom Manage 2016; 51(5): 807-15.
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Boyden JY et al. Nebulized medications for the treatment of dyspnea: a literature review. J Aerosol Med Pulm Drug Deliv 2015; 28(1): 1-19.
Simon ST et al. Benzodiazepines for the relief of breathlessness in advanced malignant and non-malignant diseases in adults. Cochrane Database Syst Rev 2016; 10: CD007354.
Zisman DA et al. A controlled trial of sildenafil in advanced idiopathic pulmonary fibrosis. N Engl J Med 2010; 363(7): 620-8.

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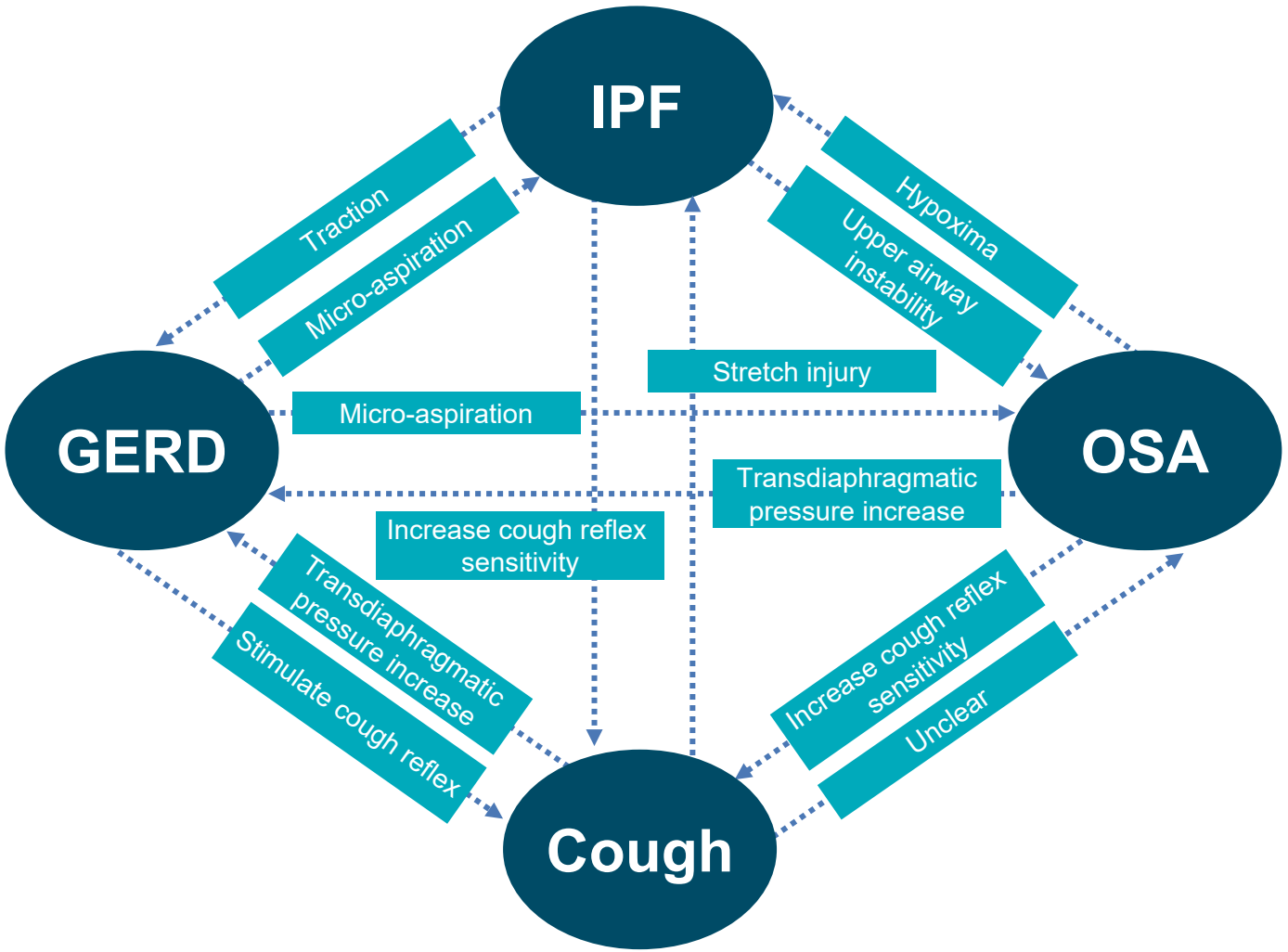
Cough : Complex interplay



First look at other causes for cough

Most common ones being

- GERD
- OSA
- Emphysema
- Ace inhibitor use
- Chronic sinusitis
- Lung Cancer
- Infection

van Manen MJ et al. Cough in idiopathic pulmonary fibrosis. Eur Respir Rev 2016; 25(141): 278-86.
Vigeland JR et al. Etiology and treatment of cough in IPF. Respir Med 2017; 123: 98-104.

GERD : gastroesophageal reflux disease
OSA : obstructive sleep apnoea

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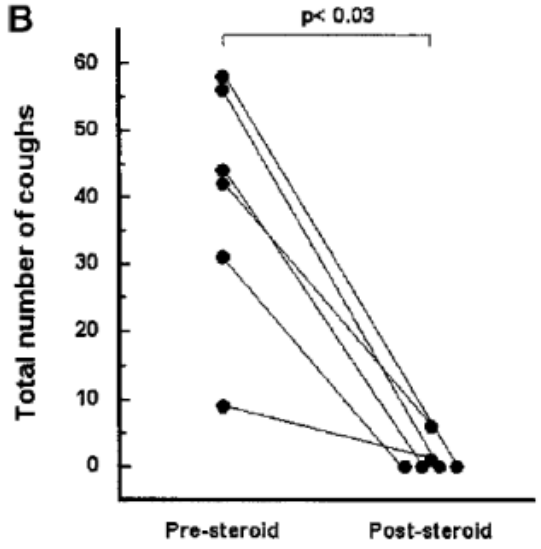
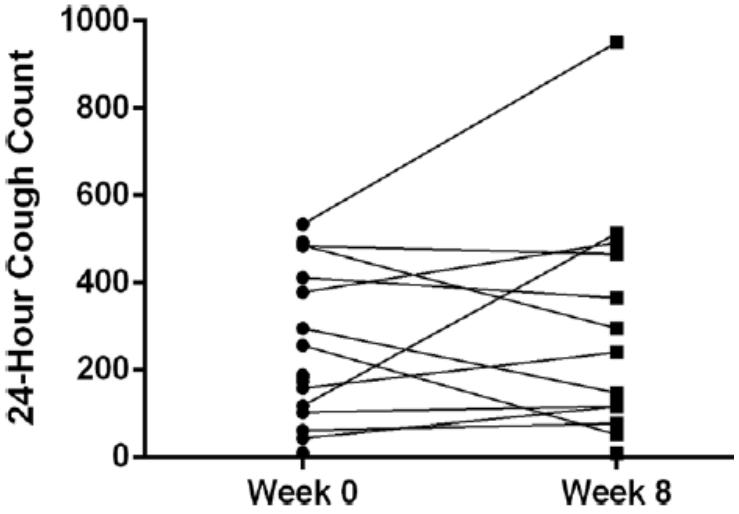
Often tried in daily practice but only anecdotal evidence in IPF-cough

- Opioids
- Different over-the-counter cough suppressants
- Codeine
- Steroids
- Acid suppression therapy

Cough counts
Pre- and post-steroids



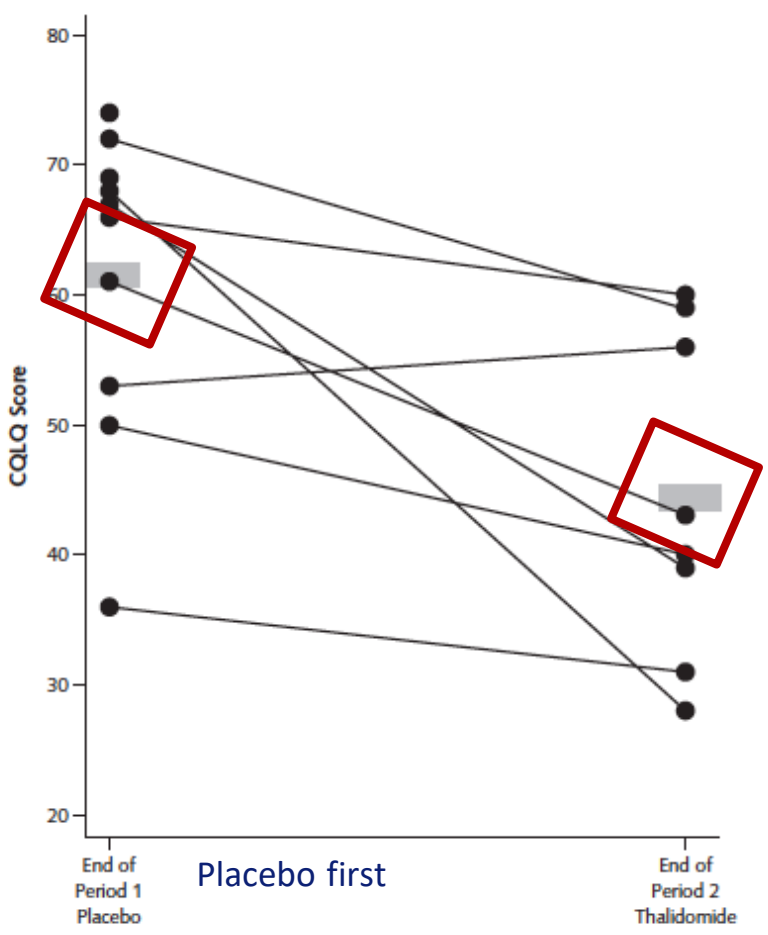
24-hour cough count before and after treatment with acid suppressant medication; p=0.70

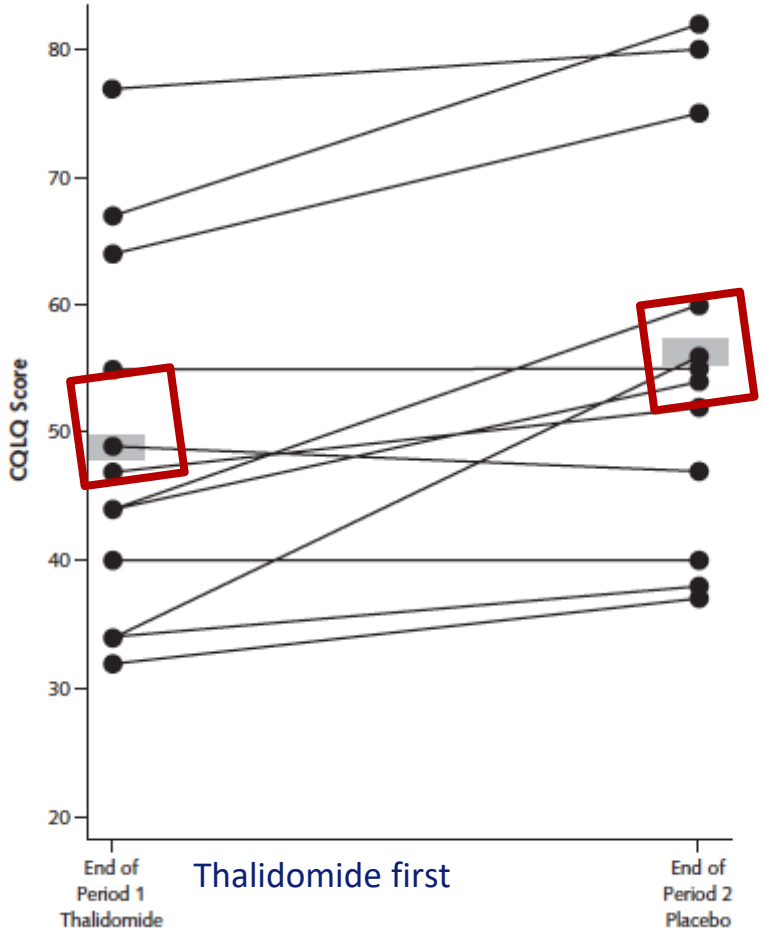


Saunders P et al. Cough in fibrotic lung disease: An unresolved challenge. Respirology 2017; 22(8): 1491-2.
Vigeland CL et al. Etiology and treatment of cough in idiopathic pulmonary fibrosis. Respir Med 2017; 123: 98-104.
Hope-Gill BD et al. A study of the cough reflex in idiopathic pulmonary fibrosis. Am J Respir Crit Care Med 2003; 168(8): 995-1002.
Kilduff CE et al. Effect of acid suppression therapy on gastroesophageal reflux and cough in idiopathic pulmonary fibrosis: an intervention study. Cough 2014; 10: 4.

Thalidomide for cough in IPF

Thalidomide decreased Cough Quality of Life Questionnaire (CQLQ) score





Caution :

20 patients (98 screened)

77% side effects

Thalidomide might ↑ risk for thromboemboli

Horton MR et al. Thalidomide for the treatment of cough in idiopathic pulmonary fibrosis: a randomized trial. Ann Intern Med 2012; 157(6): 398-406.
Bennett CL et al. Thalidomide- and lenalidomide-associated thromboembolism among patients with cancer. Jama 2006; 296(21): 2558-60.

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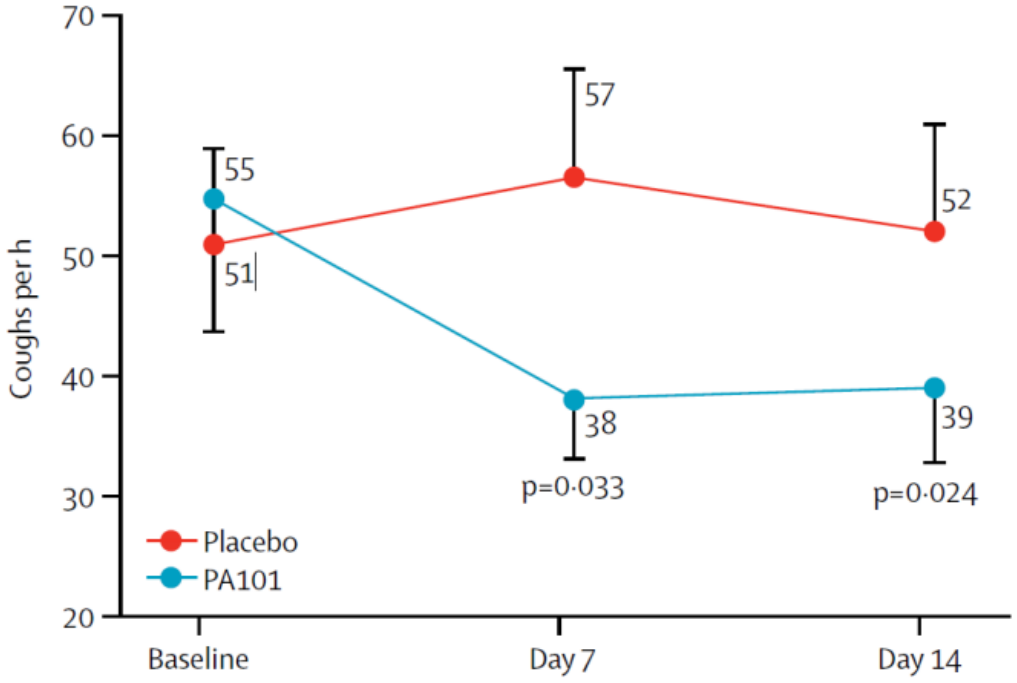
Significant improvement in refractory chronic cough with inhaled PA101 in patients with IPF

Phase II trial results

- Well tolerated, adverse events comparable with placebo
- Statistically significant 31% reduction in daytime cough frequency at Day 14 vs baseline
- QoL and cough severity scores improved with PA101



Error bars show standard error of the mean
Please note that PA101 has not been approved for the treatment of IPF

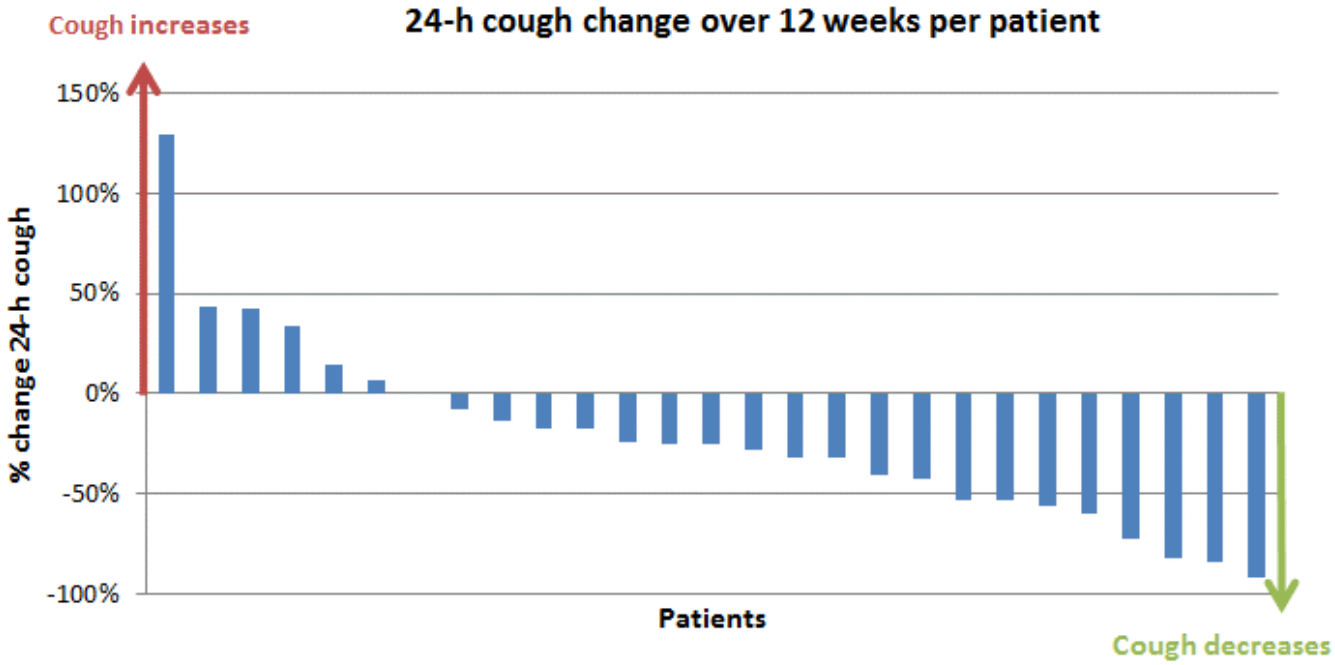
Day 14 between-group comparison (PA101–placebo)		
Total score	2·2 (–0·76 to 5·19, p=0·11)	..
Psychological	3·9 (0·37 to 7·40, p=0·032)	..
Breathlessness and activities	0·5 (–7·24 to 8·21, p=0·87)	..
Chest symptoms	8·7 (1·38 to 15·99, p=0·027)	..

Data are mean (SD) or least-squares mean difference (95% CI, p value).
K-BILD=King's Brief Interstitial Lung Disease Questionnaire. IPF=idiopathic pulmonary fibrosis.

SD, standard deviation

Birring SS et al. A novel formulation of inhaled sodium cromoglicate (PA101) in idiopathic pulmonary fibrosis and chronic cough. Lancet Respir Med 2017; 5(10): 806-15.

Observational study on effect of pirfenidone on cough in IPF showed a decrease in cough



Effect of 12 weeks' pirfenidone treatment on cough measures, linear mixed model analysis		
	Change in cough# (95% CI)	**P-value#**
24-hour cough, %	–34% (–48, –15)	0.002
LCQ, points	2.0 (1.0, 3.0)*	<0.001
VAS cough, mm	–19 (–28, –10)	<0.0001
VAS urge to cough, mm	18 (–26, –10)	<0.0001

#Analysed using a linear mixed model, change in %, points or mm
*Minimal clinical important difference for chronic cough is 1.3

LCQ, Leicester Cough Questionnaire

van Manen MJ et al. Effect of pirfenidone on cough in patients with idiopathic pulmonary fibrosis. Eur Respir J 2017; 50(4).

Fatigue treatment

- Treatment of co-morbidities
  - sleep disorders
  - anxiety / depression
  - Side effect of medication
- No evidence for pharmacological treatment
- No research done (yet) in F-ILD

Pulmonary rehabilitation

- Safe for people with ILD
- Improves short-term exercise capacity, dyspnoea, quality of life
- Little evidence available regarding long-term benefits

Dowman L et al. Pulmonary rehabilitation for interstitial lung disease. Cochrane Database Syst Rev 2014; (10): CD006322.
Ferreira A et al. Pulmonary rehabilitation in interstitial lung disease: benefits and predictors of response. Chest 2009; 135(2): 442-7.
Gomes-Neto M et al. Impact of Pulmonary Rehabilitation on Exercise Tolerance and Quality of Life in Patients With Idiopathic Pulmonary Fibrosis: a systematic review and meta-analysis. J Cardiopulm Rehabil Prev 2018.
Spruit MA et al. An official American Thoracic Society/European Respiratory Society statement: key concepts and advances in pulmonary rehabilitation. Am J Respir Crit Care Med 2013; 188(8): e13-64.

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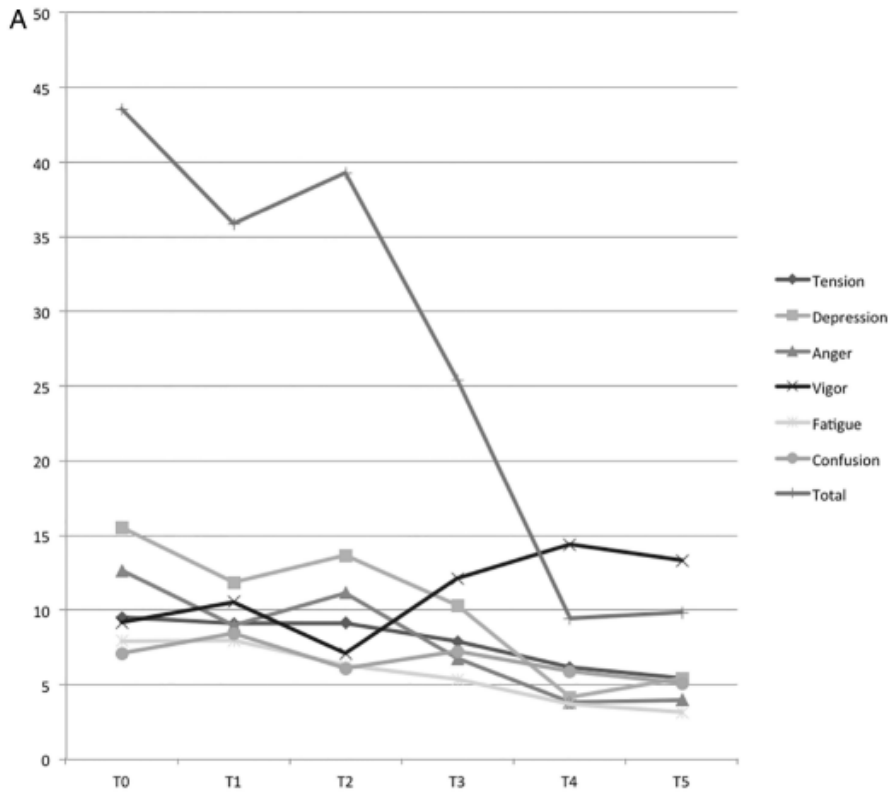
Relieving anxiety and distress in IPF; pilot data

Mindfullness improved mood
in 17 patients with ILD



Patient and Partner Empowerment Program resulted in improvement of anxiety and distress

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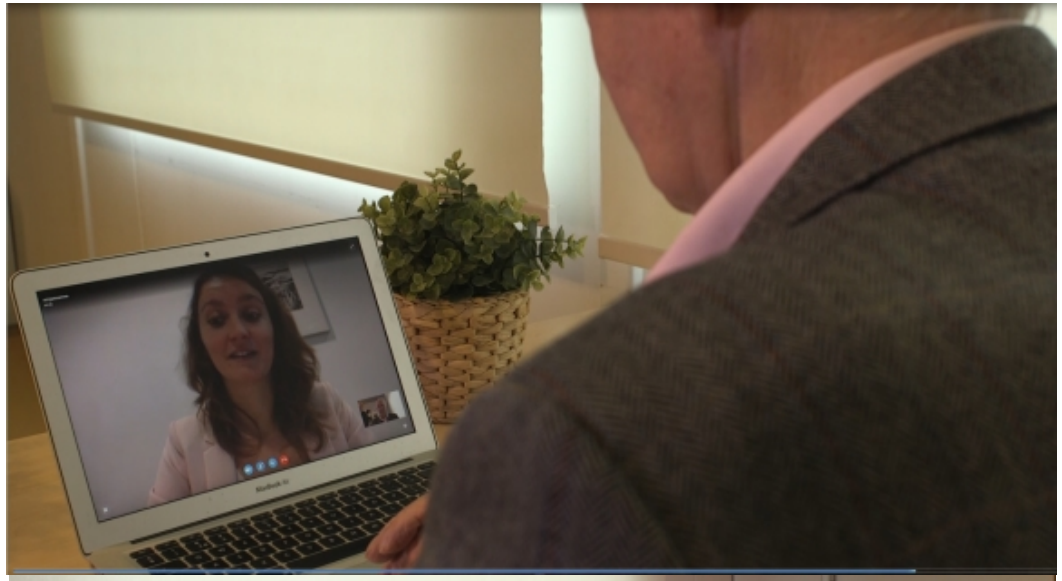
Sgalla G et al. Mindfulness-based stress reduction in patients with interstitial lung diseases: a pilot, single-centre observational study on safety and efficacy. BMJ Open Respir Res 2015; 2(1): e000065.
van Manen M et al. Patient and partner empowerment programme for idiopathic pulmonary fibrosis. Eur Respir J, 2017 Apr 26;49(4)

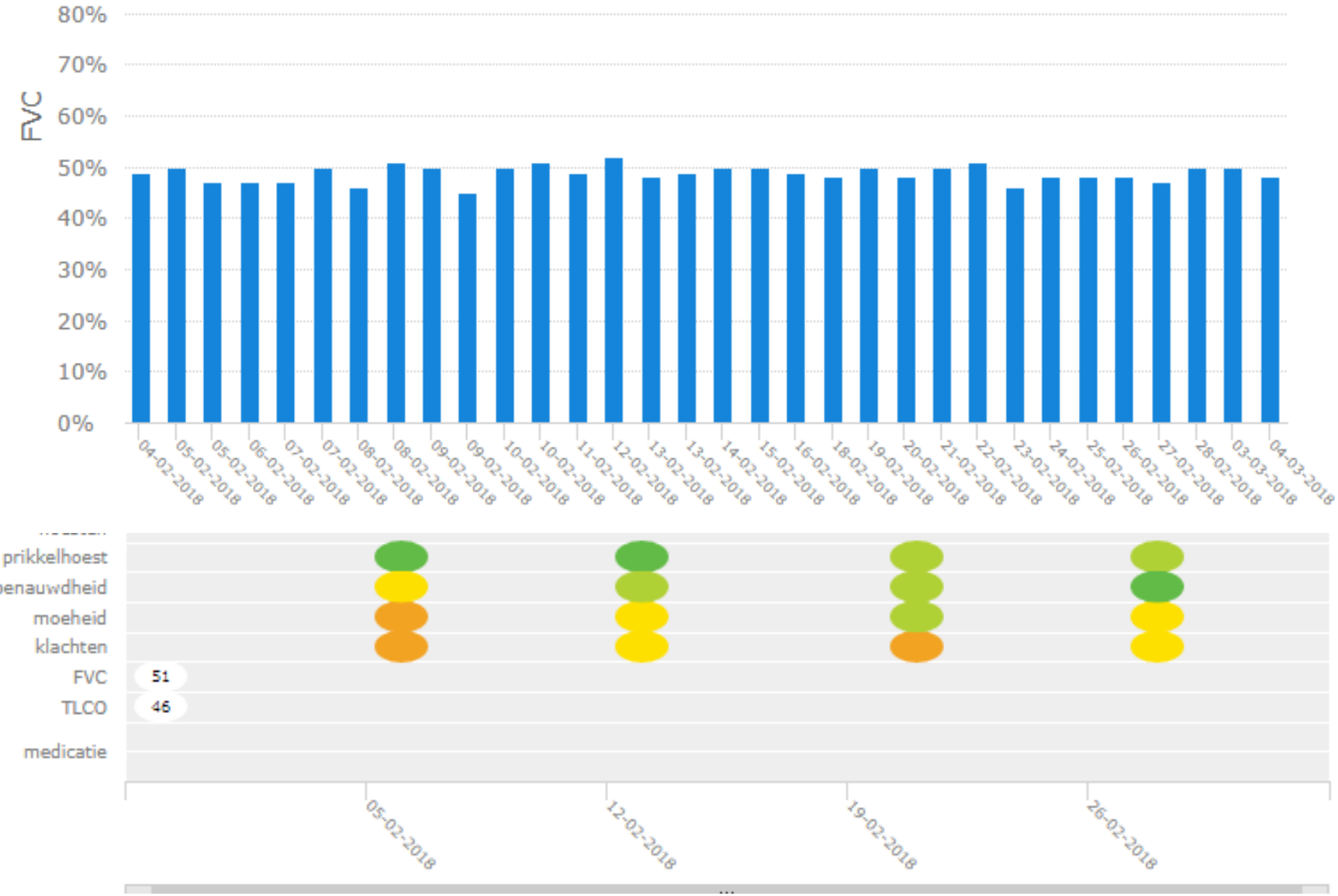
Improving quality of life is also being there for patients and families with your team; in realtime, but nowadays also online



Moor C et al, Eur Respir J. 2018 Jan 18.

Shown with permission of patient

Patient as partner in care



Daily homespirometry, Real time sent to hospital

Weekly symptom and AE scores
Real time sent to hospital

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- **The impact of disease:** patient and partner's needs
- **Therapeutic needs and palliative care go hand in hand**
- **Holistic approach to ILD care:** ABCDE of ILD care

The ABCDE of ILD care

Assess

Patients' needs and values

Patients as partners in care

Include caregivers

Kreuter M et al. Palliative care in interstitial lung disease: living well. Lancet Respir Med 2017; 5(12): 968-80.
van Manen et al. Optimizing quality of life in patients with idiopathic pulmonary fibrosis. Ther Adv Respir Dis 2017; 11(3): 157-69.

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The ABCDE of ILD care

Assess	Backing
Patients' needs and values Patients as partners in care Include caregivers	Education Self-management • Dietary support Support groups Patient advocacy groups Pulmonary rehabilitation Prevention • Stop smoking • Vaccination Discuss trial options

Kreuter M et al. Palliative care in interstitial lung disease: living well. Lancet Respir Med 2017; 5(12): 968-80.
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The ABCDE of ILD care

Assess	Backing	Comfort care and Comorbidities
Patients' needs and values Patients as partners in care Include caregivers	Education Self-management • Dietary support Support groups Patient advocacy groups Pulmonary rehabilitation Prevention • Stop smoking • Vaccination Discuss trial options	Comfort-care • Dyspnoea • Cough • Fatigue • Depression/anxiety Other palliative options Comorbidities • Cardiovascular • OSA • Lung cancer • Emphysema • GERD

Kreuter M et al. Palliative care in interstitial lung disease: living well. Lancet Respir Med 2017; 5(12): 968-80.
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The ABCDE of ILD care

Assess	Backing	Comfort care and Comorbidities	Disease-modifying treatment
Patients' needs and values Patients as partners in care Include caregivers	Education Self-management • Dietary support Support groups Patient advocacy groups Pulmonary rehabilitation Prevention • Stop smoking • Vaccination Discuss trial options	Comfort-care • Dyspnoea • Cough • Fatigue • Depression/anxiety Other palliative options Comorbidities • Cardiovascular • OSA • Lung cancer • Emphysema • GERD	Antifibrotic drugs • Pirfenidone • Nintedanib Lung transplantation (if patient is eligible)

Kreuter M et al. Palliative care in interstitial lung disease: living well. Lancet Respir Med 2017; 5(12): 968-80.
van Manen et al. Optimizing quality of life in patients with idiopathic pulmonary fibrosis. Ther Adv Respir Dis 2017; 11(3): 157-69.

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The ABCDE of ILD care

Assess	Backing	Comfort care and Comorbidities	Disease-modifying treatment	End-of-life care
Patients' needs and values Patients as partners in care Include caregivers	Education Self-management • Dietary support Support groups Patient advocacy groups Pulmonary rehabilitation Prevention • Stop smoking • Vaccination Discuss trial options	Comfort-care • Dyspnoea • Cough • Fatigue • Depression/anxiety Other palliative options Comorbidities • Cardiovascular • OSA • Lung cancer • Emphysema • GERD	Antifibrotic drugs • Pirfenidone • Nintedanib Lung transplantation (if patient is eligible)	Timing of discussion Discuss • Fears • Palliative options • Practical needs • Preferred place of dying • Preferred way of dying Discuss treatment limits • About resuscitation • About intubation

Kreuter M et al. Palliative care in interstitial lung disease: living well. Lancet Respir Med 2017; 5(12): 968-80.
van Manen et al. Optimizing quality of life in patients with idiopathic pulmonary fibrosis. Ther Adv Respir Dis 2017; 11(3): 157-69.

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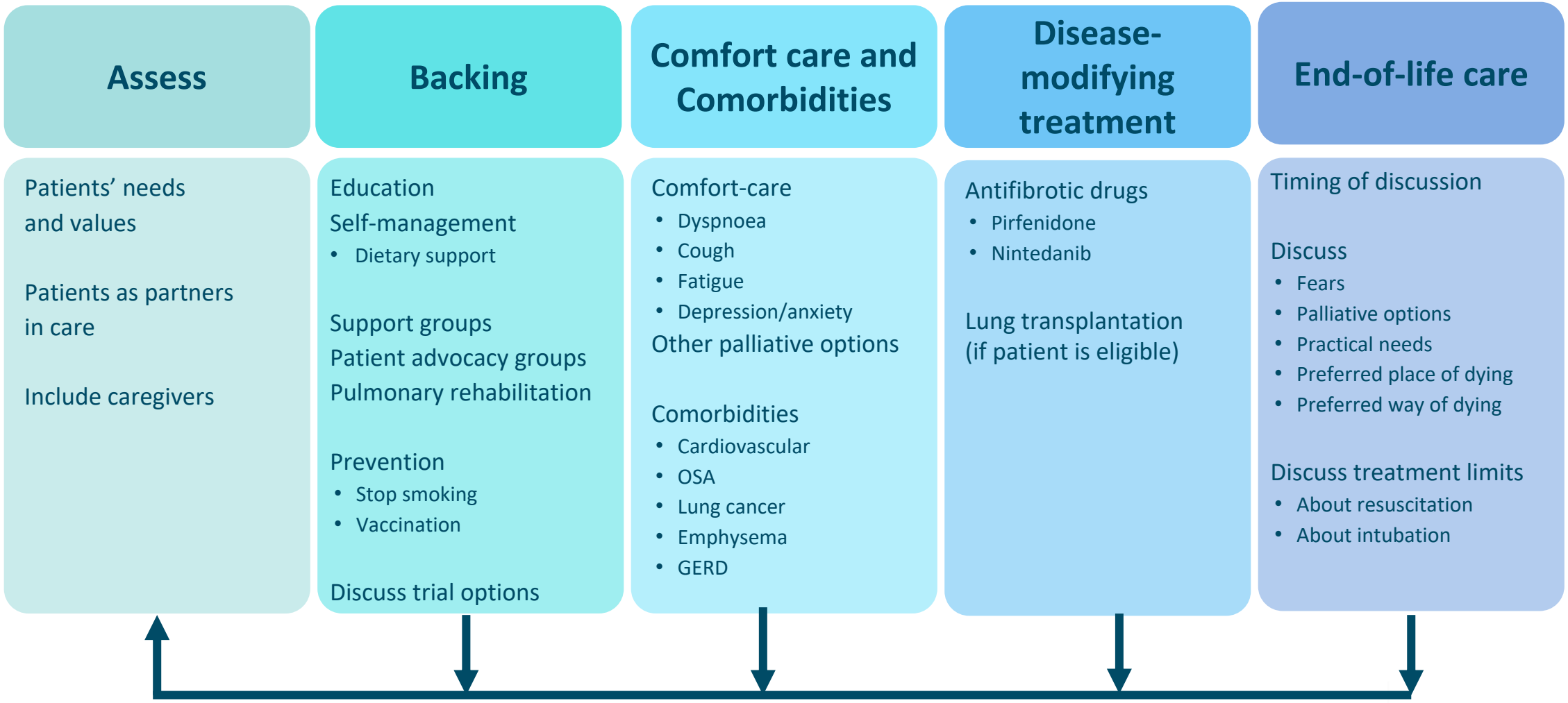
A process of constant re-assessment of disease as well as patient needs and wishes



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van Manen et al. Optimizing quality of life in patients with idiopathic pulmonary fibrosis. Ther Adv Respir Dis 2017; 11(3): 157-69.

Comprehensive care for patients with pulmonary fibrosis

- IPF, but also other progressive fibrotic diseases, have a major impact on patient's and partner's lives
- Palliative care is about relieving symptoms and providing support, this requires complementary strategies besides disease-centered management
- Care in ILD is a process of constant re-assessment of disease as well as patient's needs and wishes
- There is a great lack of good studies into treatment of symptoms and improving quality of life in IPF/ILD