

# *Que peut-on attendre d'une prise en charge par un kinésithérapeute ?*

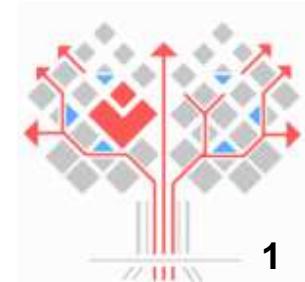
Session A44, CPLF, Lille, 31 janvier 2016



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**Pôle Thorax et Vaisseaux**

**Centre Henri Bazire, Saint  
Julien de Ratz**

**Inserm1055**  
**Biologie Environnementale et**  
**Systémique - BEeSy**



# **Conflits d'intérêts 3 dernières années**

## ***déplacements, conférences, fonds de recherche***

- Actélion
- Astra Zeneca
- Bayer
- Boehringer Ingelheim
- GlaxoSmithKline
- Lilly
- Novartis
- Pfizer
- Pierre Fabre
- Roche
- Sanofi
- *Therakos*
- *PneumRx, Pulmonx, Medwin, Aeris, Holaira*
- *AGIR@dom, Orkyn, Vitalaire, IPS, SOS Oxygène*

# Sommaire

- ***Connaitre les principes physiopathologiques, épidémiologiques et thérapeutiques des pneumopathies interstitielles diffuses***  
*Parcours patient, du diagnostic aux soins palliatifs*
- **Compétences cognitives, comportementales et techniques, contexte souvent pluridisciplinaire**
  - réhabilitation, hors contexte transplantation et av/ap Tx
  - oxygénothérapie, modalités
  - VNI
  - palliation dyspnée

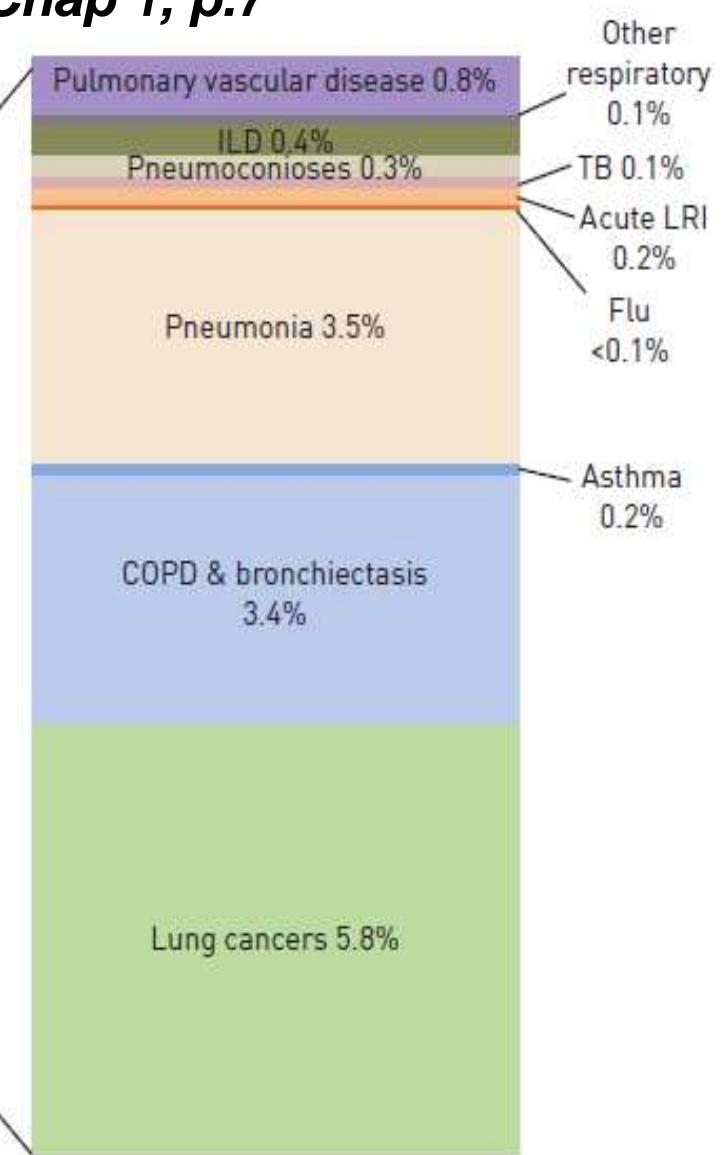
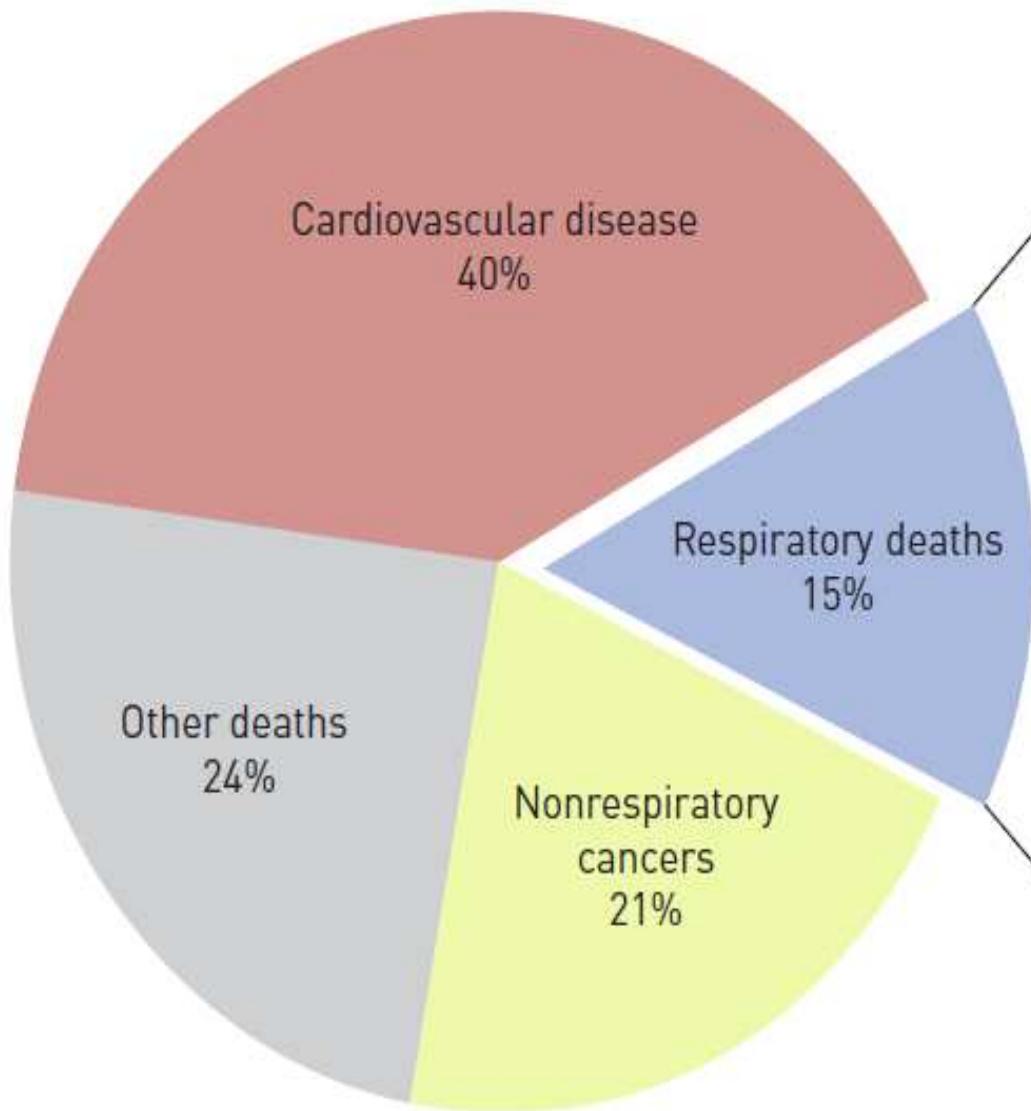
- Médecin consultant Centre Henri Bazire, St Julien de Ratz



- DMD, 2 réunions / mois depuis 10 ans extra, intra CHU

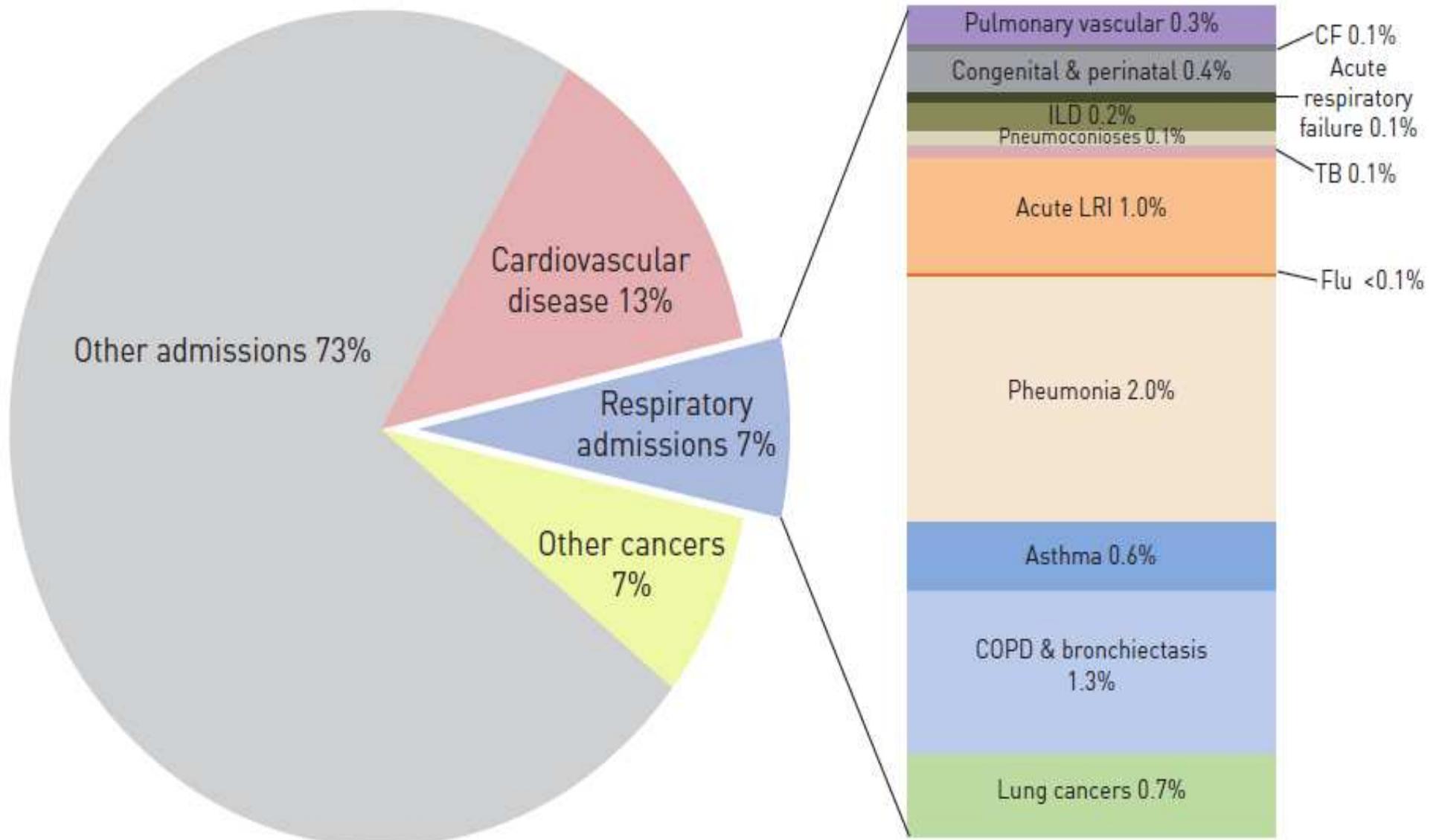
# Epidémiologie – % décès en Europe

*White Book, ERS, ELF 2013, Chap 1, p.7*



# Epidémiologie – % admission en Europe

*White Book, ERS, ELF 2013, Chap 1, p.7*



# Classification

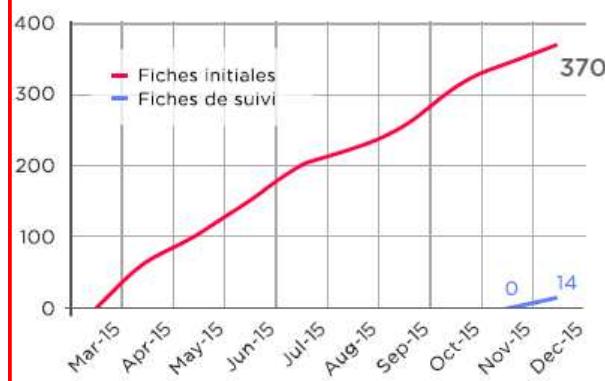
## PNEUMOPATHIES INTERSTITIELLES DIFFUSES

	Cause connue	Contexte défini, cause inconnue	Cause inconnue
<b>Antigène organique</b>	Pneumopathie d'hypersensibilité ± chronique	<b>Connectivites</b> Polyarthrite rhumatoïde, Syndrome Gougerot Sjögren Sclérodermie, Polymyosite et dermatopolymyosite	Fibrose pulmonaire idiopathique (FPI)
<b>Agent minéral</b>	<b>Pneumoconioses</b> Silicose, asbestose, berylliose	<b>Granulomatoses</b> Sarcoidose	Pneumopathie interstitielle non spécifique (PINS)
<b>Médicament</b>	Pneumopathie médicamenteuse	<b>Vascularites</b>	Pneumopathie organisée cryptogénique, bronchiolite respiratoire avec PI, PI desquamative
<b>Insuffisance cardiaque</b>	Œdème interstitiel cardiogénique	<b>Autres</b> Histiocytose X Protéinose alvéolaire, Lymphangioléiomatomateuse, Pneumopathie chronique à éosinophiles	<b>Autres et PI rares</b> (Lymphocytaire, fibroélastose) ou <b>Inclassables</b>
<b>Néoplasie</b>	Lymphangite carcinomateuse, lymphome, carcinome bronchioloalvéolaire		
<b>Infection chronique</b>	Pneumocystose, miliaire tuberculeuse		

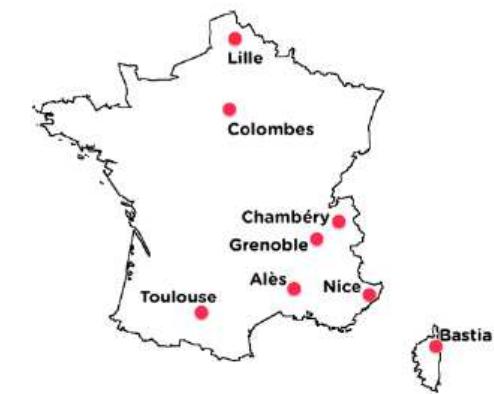
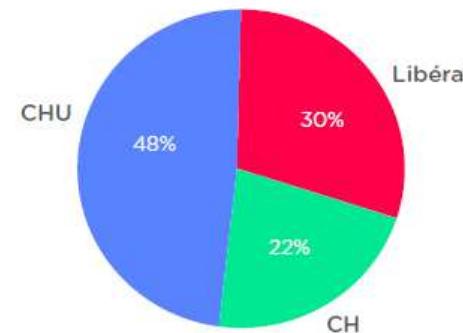
## Colibri-PID

Observatoire web pour faciliter le cheminement diagnostique et le suivi des pneumopathies interstitielles diffuses (PID).

Evolution des inclusions de patients

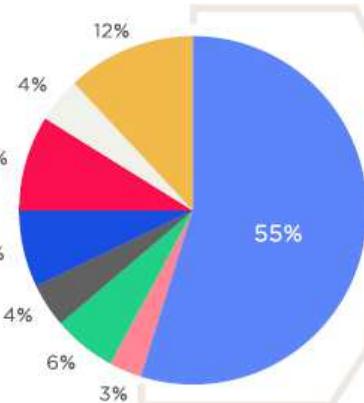


Inclusions selon le lieu d'exercice

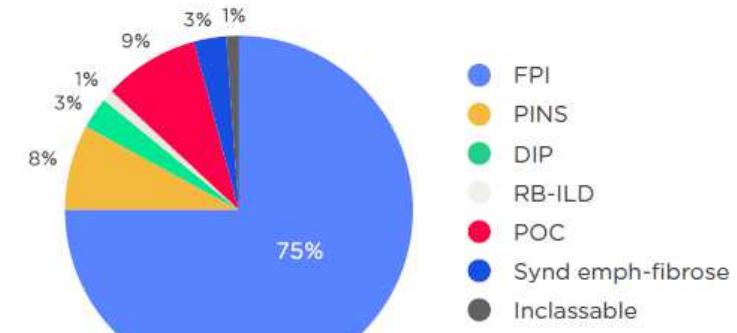


Etiologie des PID\*

- PI idiopathique
- Médicamenteuse
- Professionnelle
- PHS
- Connectivité
- Sarcoïdose
- Cause autre
- PI inclassable



PI idiopathiques\*



[PDF](#) [DOC](#) [PPT](#)  
PDF Word (modifiable) PowerPoint (modifiable)

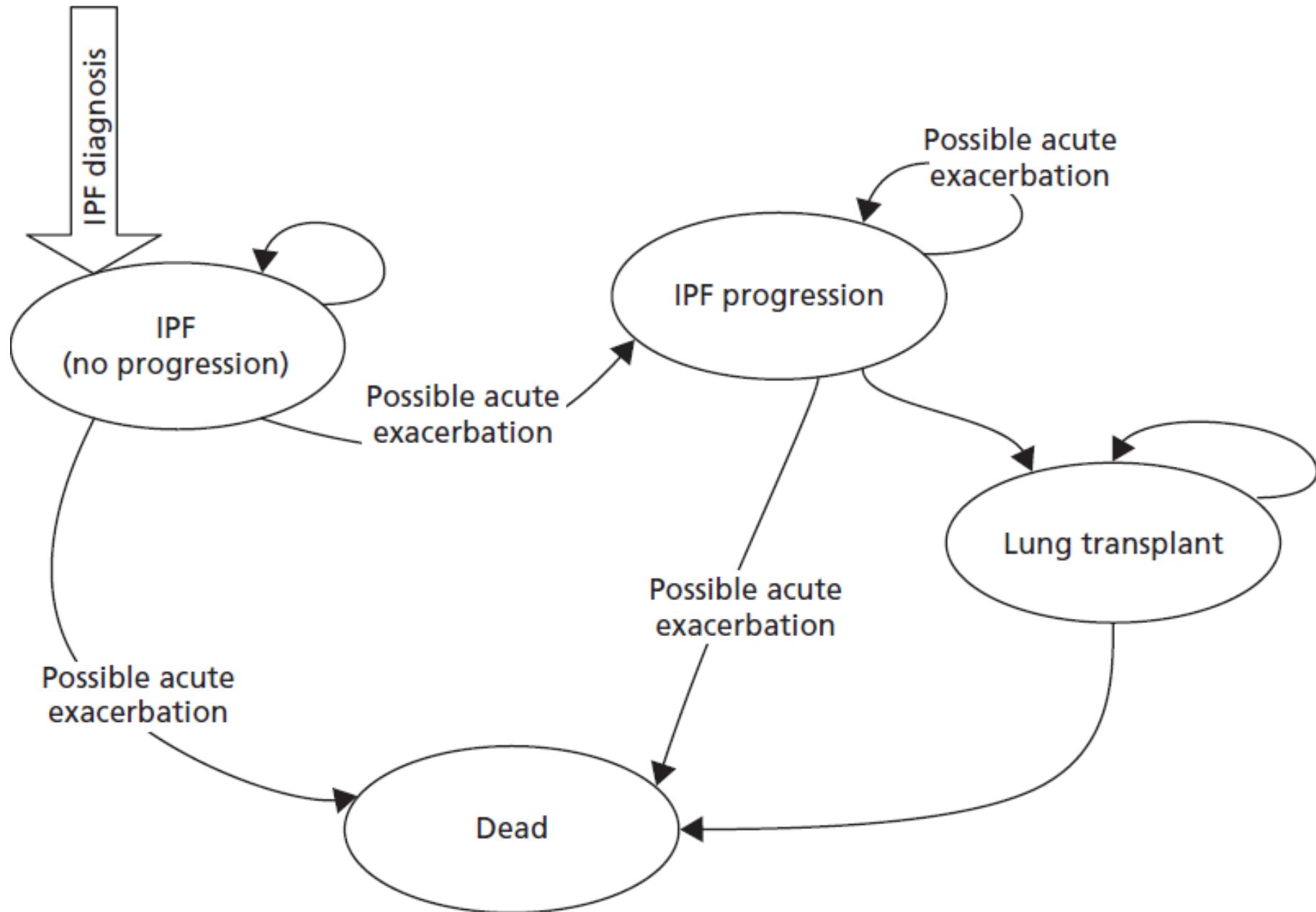
Enregistrer un changement

Assainissement	<input type="radio"/> Dystrophe	<input type="radio"/> Amélioration	<input type="radio"/> Stabilité	<input type="radio"/> Aspiration
Toux	<input type="radio"/> Dystrophe	<input type="radio"/> Amélioration	<input type="radio"/> Stabilité	<input type="radio"/> Aspiration
Expectoration	<input type="radio"/> Dystrophe	<input type="radio"/> Amélioration	<input type="radio"/> Stabilité	<input type="radio"/> Aspiration

### Conclusion

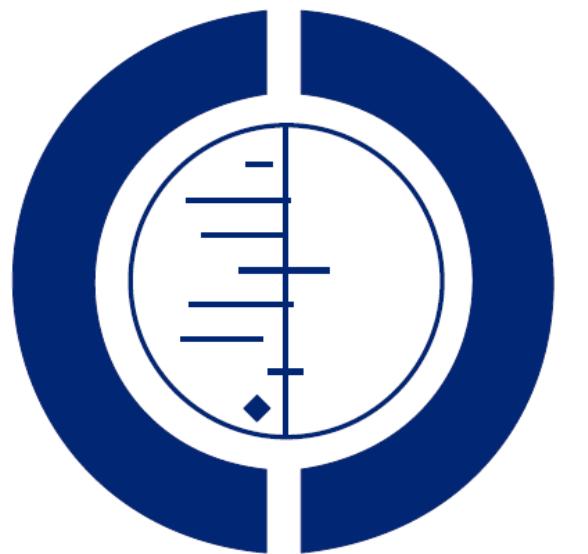
Après 8 mois de fonctionnement, 50 praticiens ont inclus 370 patients, ce qui atteste que Colibri-PID rend service aux utilisateurs. Sous réserve de s'assurer de la fiabilité des données, Colibri-PID devrait permettre de contribuer à la recherche en vraie vie sur les pneumopathies interstitielles diffuses. Chaque praticien peut utiliser Colibri-PID en se connectant sur [www.colibri-pneumo.fr](http://www.colibri-pneumo.fr).

# Parcours patient



# Pulmonary rehabilitation for interstitial lung disease (Review)

Dowman L, Hill CJ, Holland AE



**THE COCHRANE  
COLLABORATION®**

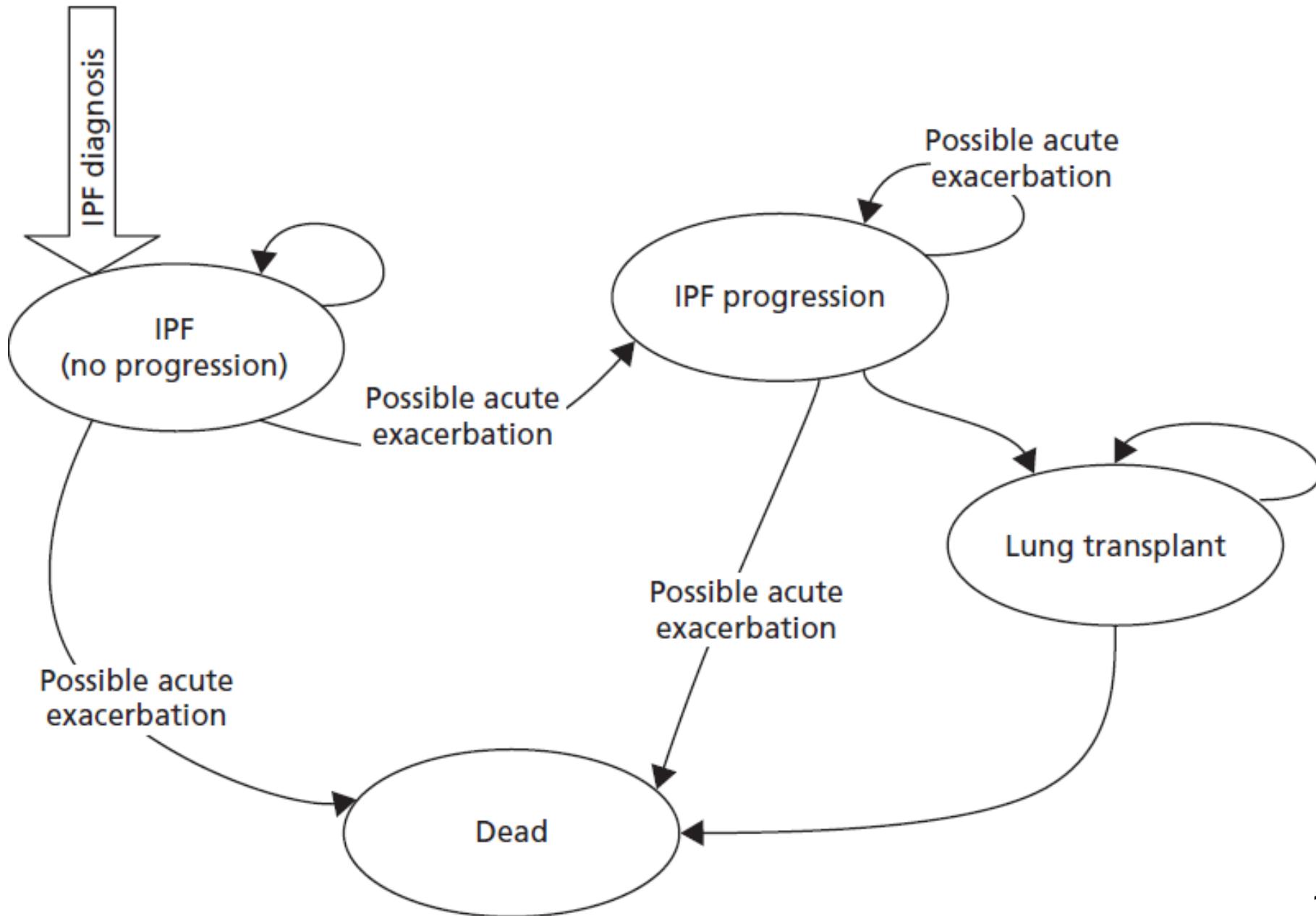
# Réhabilitation PID

Outcomes	Illustrative comparative risks* (95% CI)		Relative effect (95% CI)	Number of participants (studies)	Quality of the evidence (GRADE)
	Assumed risk	Corresponding risk			
	No pulmonary rehabilitation	Pulmonary rehabilitation			
<b>Change in 6-minute walk distance</b> 6-Minute walk test Follow-up: end of rehabilitation (8-12 weeks)	Mean change in 6-minute walk distance ranged across control groups from -4 to 17 metres	Mean change in 6-minute walk distance in the intervention groups was 44 higher (26 to 63 higher)	MD 44.34 (26.04 to 62. 64)	168 (5 studies)	⊕⊕⊕○ moderate <sup>a</sup>
<b>Change in peak oxygen uptake</b> Cardiopulmonary exercise test Follow-up: end of rehabilitation (8-12 weeks)	Mean change in peak oxygen uptake ranged across control groups from -0.02 to 0.4 mL/kg/min	Mean change in peak oxygen uptake in the intervention groups was 1.24 higher (0.46 to 2.03 higher)	MD 1.24 (0.46 to 2.13)	80 (2 studies)	⊕⊕○○ low <sup>b,c</sup>
<b>Change in maximum ventilation</b> Cardiopulmonary exercise test Follow-up: end of rehabilitation (8 weeks)	Mean change in maximum ventilation in control groups was -1.04 L/min	Mean change in maximum ventilation in the intervention groups was 4.71 higher (0.1 to 9.32 higher)	MD 4.71 (0.10 to 9.32)	52 (1 study)	⊕⊕○○ low <sup>d</sup>

# Réhabilitation PID

Outcomes	Illustrative comparative risks* (95% CI)		Relative effect (95% CI)	Number of participants (studies)	Quality of the evidence (GRADE)
	Assumed risk	Corresponding risk			
	No pulmonary rehabilitation	Pulmonary rehabilitation			
Change in dyspnoea score Modified Medical Research Council Dyspnoea Scale Follow-up: end of rehabilitation (8-12 weeks)	Mean change in dyspnoea score ranged across control groups from <b>0.11 to 0.3 points</b>  Follow-up: end of rehabilitation (8-12 weeks)	Mean change in dyspnoea score in the intervention groups was <b>0.60 lower</b> (0.96 to 0.26 lower)	SMD -0.66 (-1.05 to -0.28)	113 (3 studies)	⊕⊕○○ <b>low<sup>a,e</sup></b>
Change in quality of life Chronic Respiratory Disease Questionnaire (total score) Follow-up: end of rehabilitation (8-12 weeks)	Mean change in quality of life in control groups was <b>3.29 points</b>  Follow-up: end of rehabilitation (8-12 weeks)	Mean change in quality of life in the Intervention groups was <b>8.9 higher</b> (3 to 14.8 higher)	SMD 0.59 (0.2 to 0.98)	106 (3 studies)	⊕⊕○○ <b>low<sup>a,e</sup></b>
6-Month survival	<b>74 per 1000</b>	<b>67 per 1000</b> (10 to 353)	RR 0.9 (0.13 to 4.77)	57 (1 study)	⊕⊕○○ <b>low<sup>f</sup></b>
Adverse events Follow-up: 6 months	See comment	See comment	Not estimable	85 (2 studies)	See comment

# Parcours patient



# Réhabilitation PID, pré/post Tx

## Indications pour adresser le patient au centre de Tx

- Histopathologic or radiographic evidence of usual interstitial pneumonitis (UIP) or fibrosing non-specific interstitial pneumonitis (NSIP), regardless of lung function.
- Abnormal lung function: forced vital capacity (FVC) < 80% predicted or diffusion capacity of the lung for carbon monoxide ( $Dl_{CO}$ ) < 40% predicted.
- Any dyspnea or functional limitation attributable to lung disease.
- Any oxygen requirement, even if only during exertion.
- For inflammatory interstitial lung disease (ILD), failure to improve dyspnea, oxygen requirement, and/or lung function after a clinically indicated trial of medical therapy.

# Réhabilitation PID, pré/post Tx

## Indications pour lister le patient pour une transplantation

- Decline in FVC > 10% during 6 months of follow-up (note: a > 5% decline is associated with a poorer prognosis and may warrant listing).
- Decline in DL<sub>CO</sub> > 15% during 6 months of follow-up.
- Desaturation to < 88% or distance < 250 m on 6-minutewalk test > 50 m decline in 6-minute-walk distance over a 6-month period.
- Pulmonary hypertension on right heart catheterization or 2-dimensional echocardiography.
- Hospitalization because of respiratory decline, pneumothorax, or acute exacerbation.

# Réhabilitation PID, pré/post Tx

## *un processus continu*

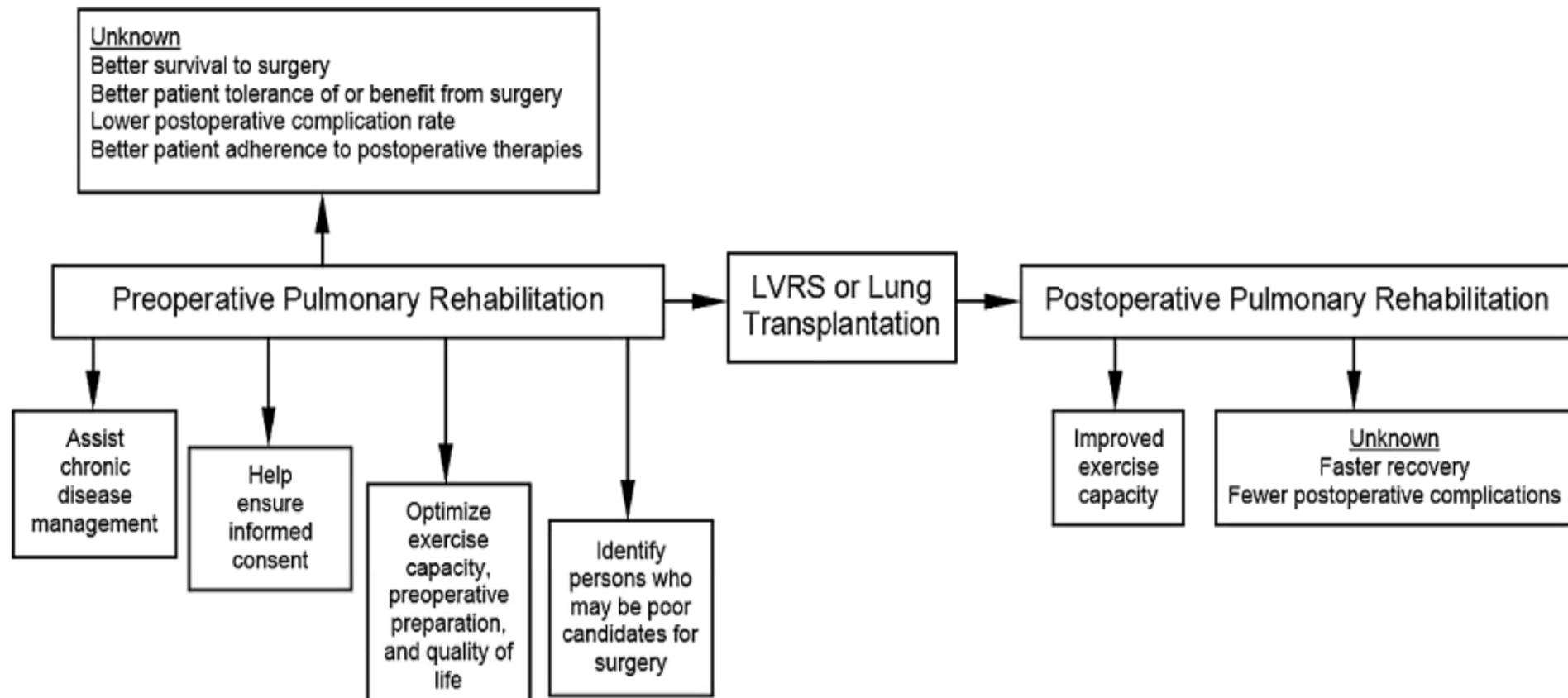


Fig. 1. Schema summarizing the rationale for and benefits of pulmonary rehabilitation for patients who undergo lung volume reduction surgery or lung transplantation.

# Réhabilitation PID, préTx

## *un processus continu*

**IRAD2, Insuffisance Respiratoire Chronique à Domicile**

**Pison et al. Thorax 2011;66:953-60**

Patients	60, 66,6 ± 9,6 ans, IMC 21,5 ± 3,8 62, 65,1 ± 9,6 ans, IMC 21,4 ± 4,0
Durée	12 semaines, 12 mois de suivi
Intervention	Éducation + Exercice + ONS + testostérone orale <i>versus</i> Éducation

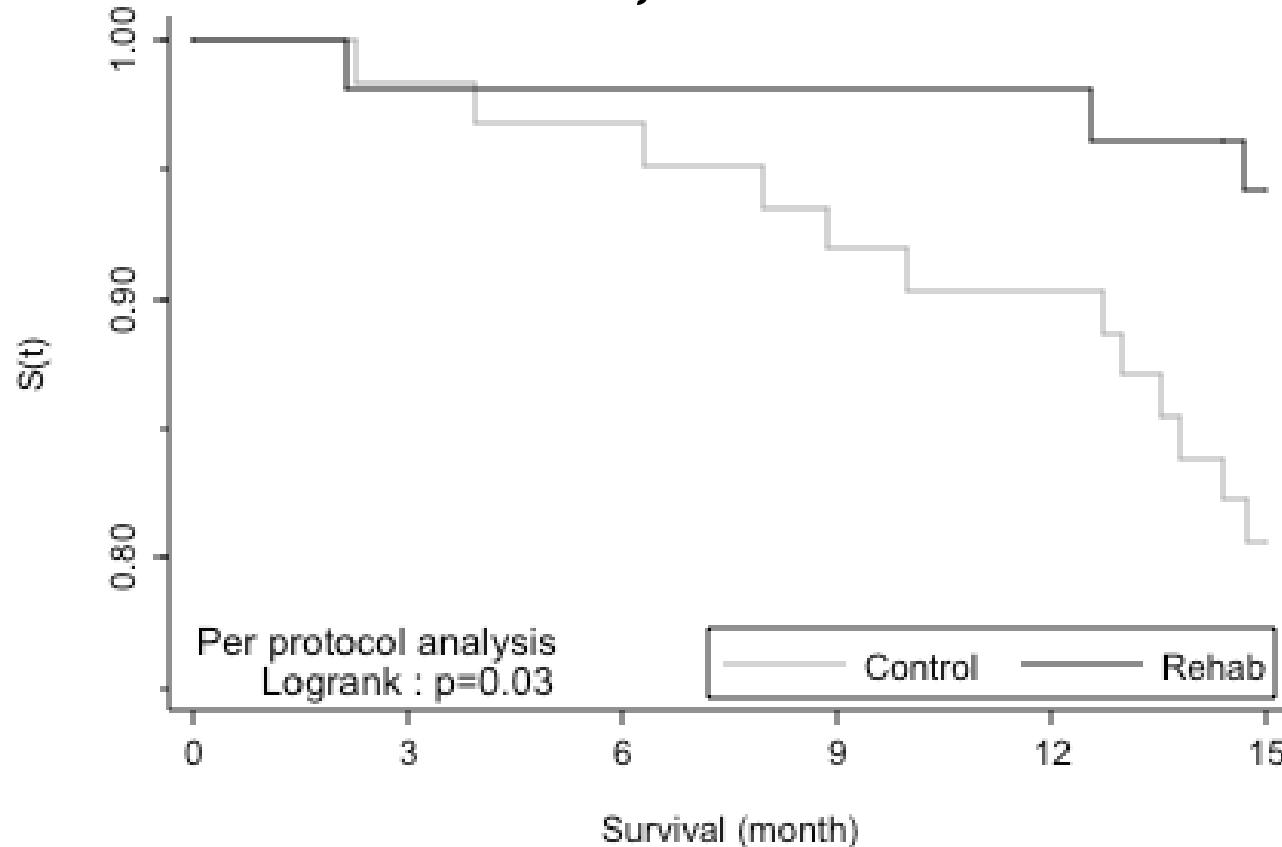
### Résultats

- 3 mois : prise de poids, ↑ masse musculaire, Hb, endurance, Wmax, QdV des femmes
- 15 mois : survie augmentée en per-protocole

# Réhabilitation PID, pré/post Tx

## *un processus continu*

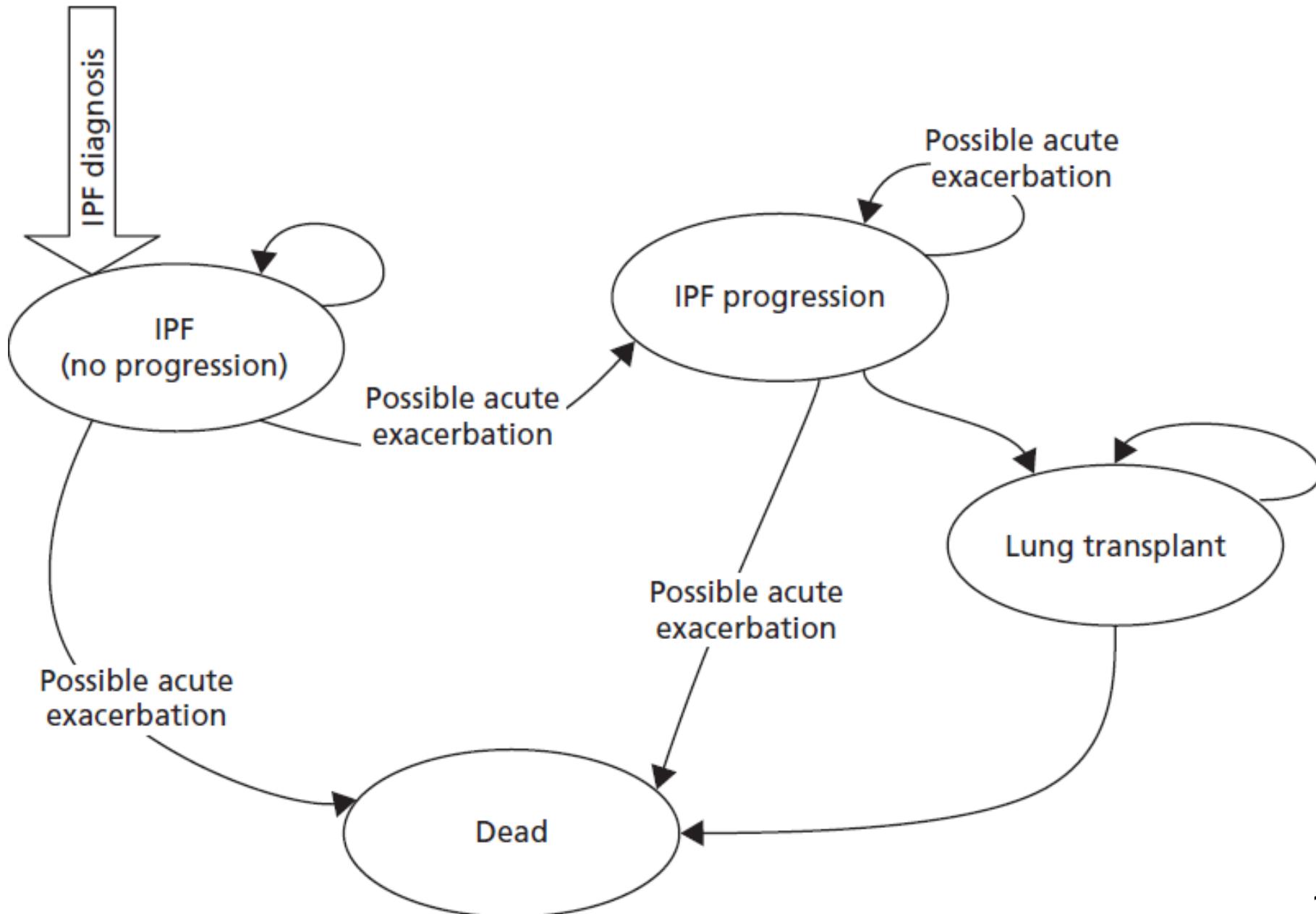
IRAD2, Insuffisance Respiratoire Chronique à Domicile  
*Pison et al. Thorax 2011;66:953-60*

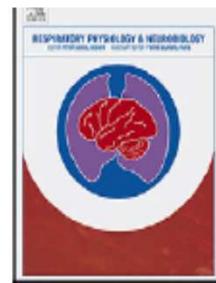


Number at risk

Control	62	61	60	57	56	50
Rehabilitation	52	51	51	51	51	48

# Parcours patient





## Benefits of home-based endurance training in lung transplant recipients

Sabine Vivodtzev<sup>a,h</sup>, Christophe Pison<sup>b,c</sup>, Karen Guerrero<sup>b,d</sup>, Paulette Mezin<sup>e</sup>, Elisabeth Maclet<sup>c</sup>, Jean-Christian Borel<sup>a</sup>, Philippe Chaffanjon<sup>f</sup>, Rachid Hacini<sup>g</sup>, Olivier Chavanon<sup>g</sup>, Dominique Blin<sup>g</sup>, Bernard Wuyam<sup>a,h,\*</sup>

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CHU de Grenoble, Pole Physiologie Rééducation, Clinique de Physiologie, Sommeil & Exercice, BP217, 38043, Grenoble, France

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### ARTICLE INFO

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#### Article history:

Accepted 11 February 2011

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#### Keywords:

Lung transplant recipients  
Cystic fibrosis  
Pulmonary rehabilitation

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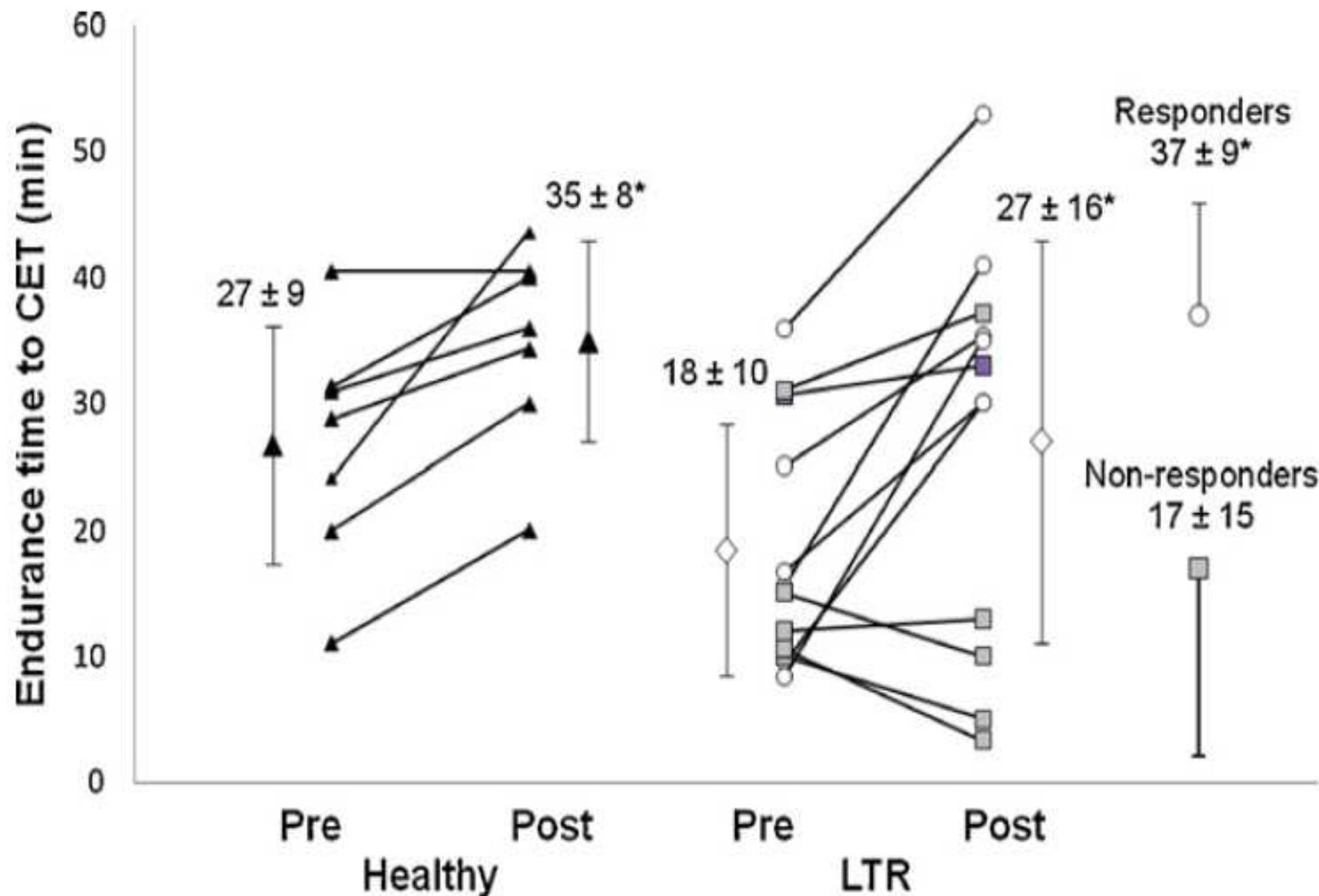
### ABSTRACT

**Background:** To investigate the effect of home-based exercise training on exercise tolerance, muscle function and quality of life in lung transplant recipients (LTR).

**Methods:** Twelve LTR and 7 age-matched healthy subjects underwent exercise training (ET, 12-wk, 3×/wk, 40 min). Peak aerobic capacity ( $\dot{V}O_{2\text{peak}}$ ), endurance time ( $T_{\text{end}}$ ), minute ventilation ( $\dot{V}E$ ) quadriceps strength, percentage of type I fiber (%Ifb), fiber diameters and chronic respiratory questionnaire were assessed before and after ET. A positive response to ET was defined as an improvement in  $T_{\text{end}}$  at least comparable to the mean change observed in healthy subjects.

# Réhabilitation PID, post Tx

## *un processus continu*



# Post transplantation pulmonaire

Am J Transplant 2012, mars

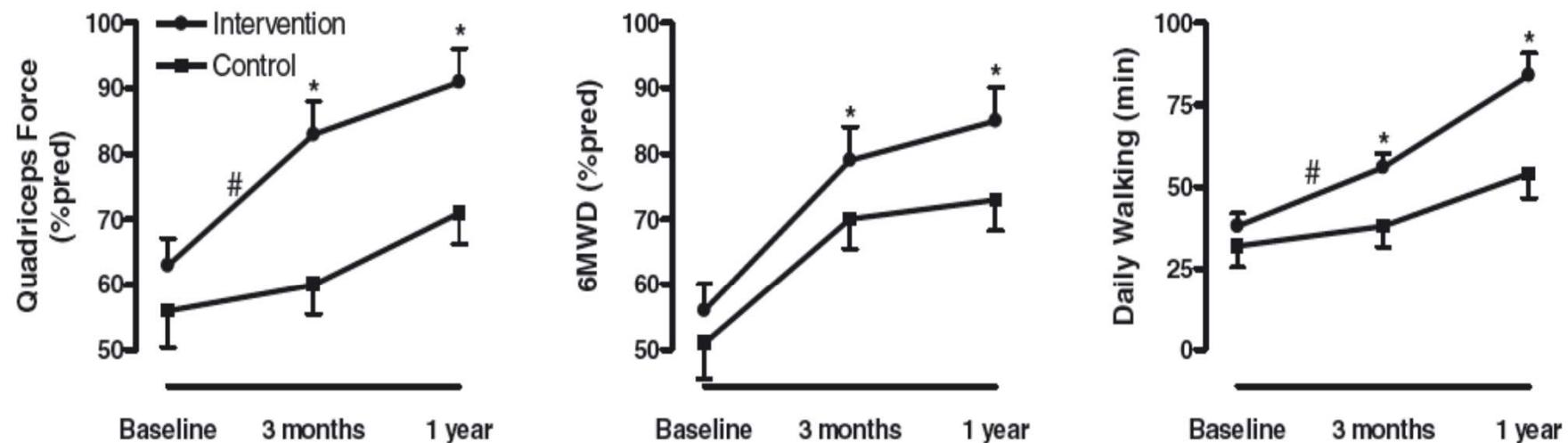
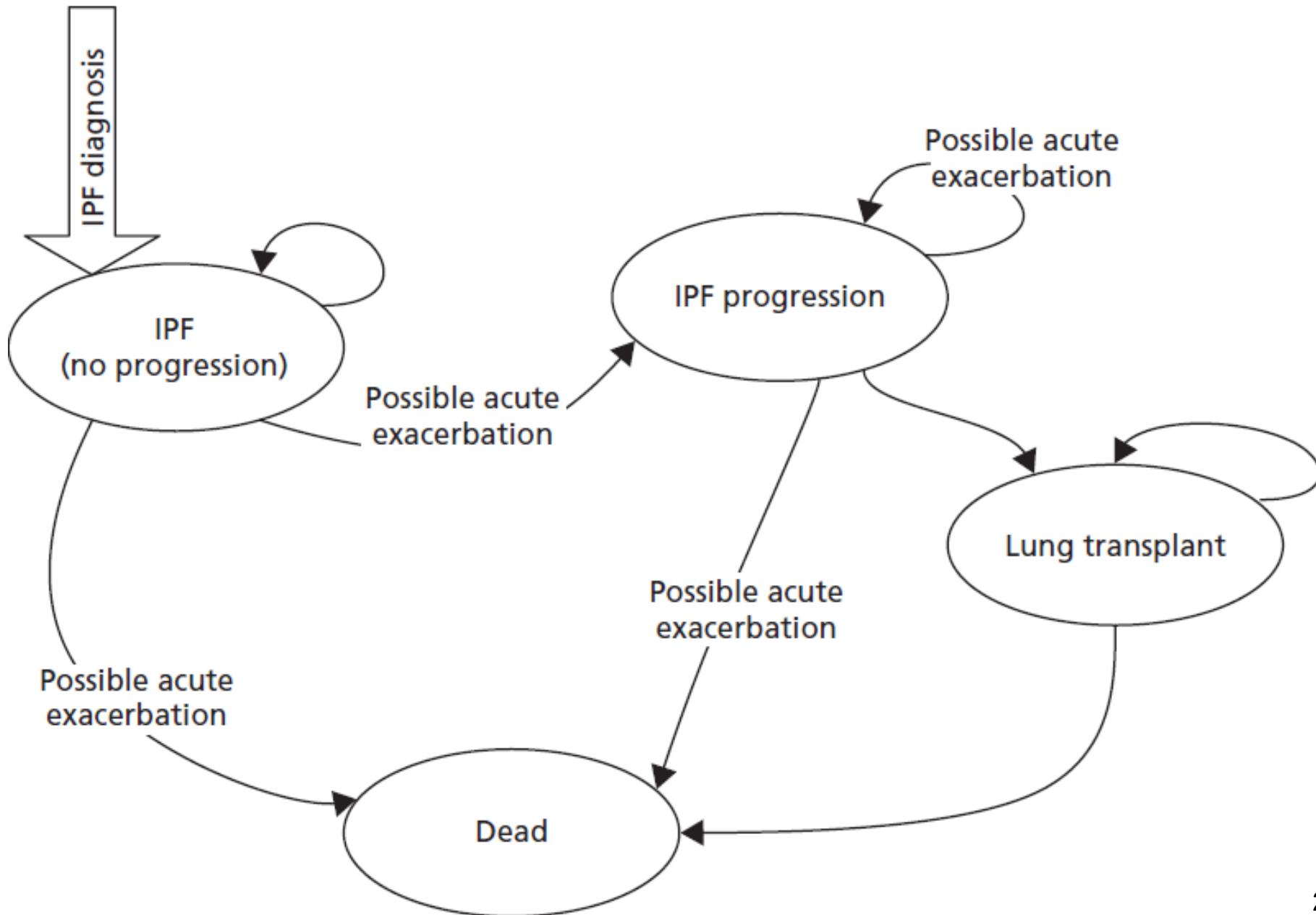


Figure 2: Progression of quadriceps force, 6-minute walking distance (6MWD) and daily walking time during the intervention period (Baseline to 3 months) and during the follow-up period (3 months to 1 year). \*, significant difference between groups; #, significant difference in slopes between groups.

# Parcours patient



# Oxygénothérapie

STUDY PROTOCOL

Open Access

## Protocol for a mixed-methods study of supplemental oxygen in pulmonary fibrosis

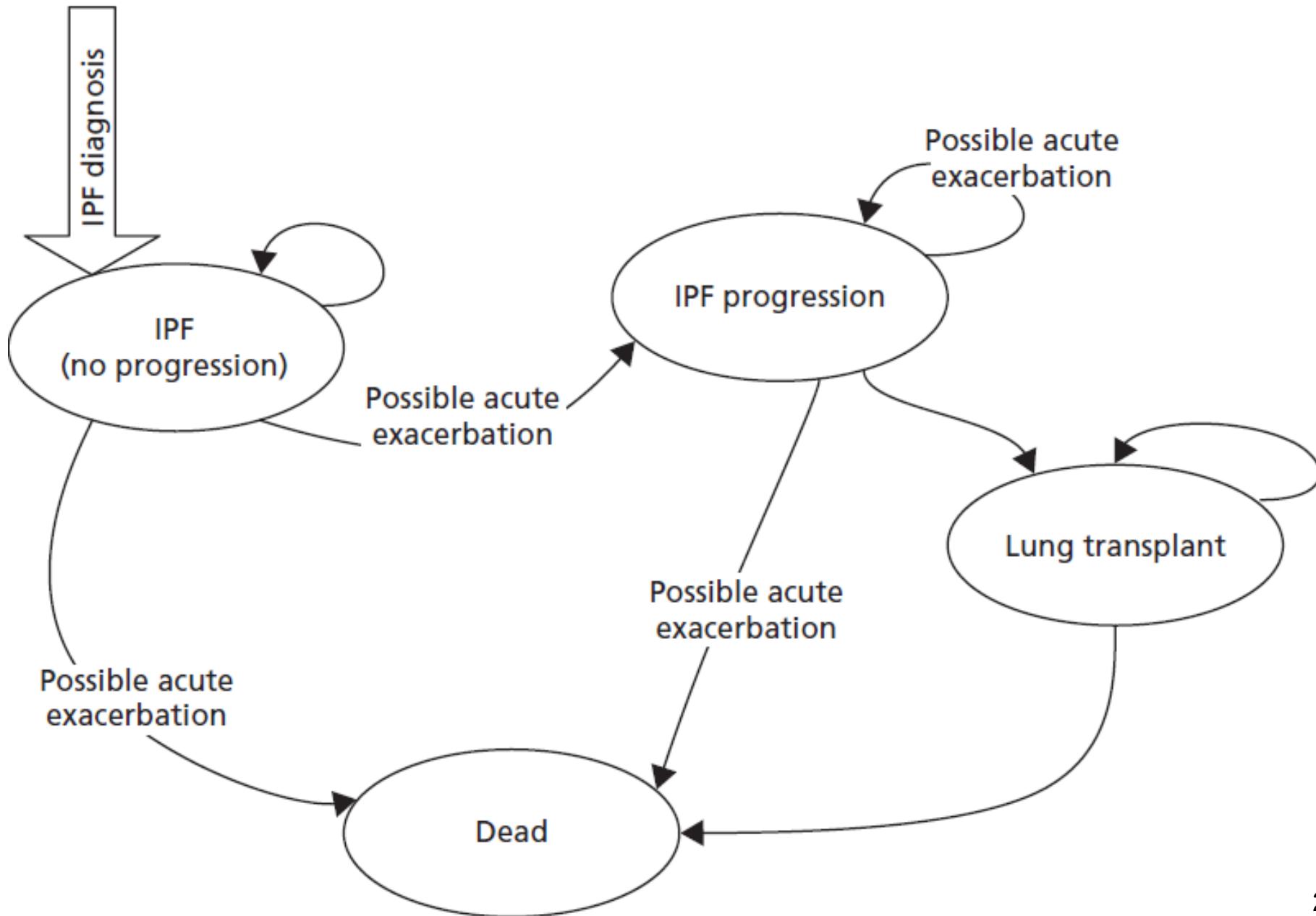
Amanda Belkin<sup>1,3</sup>, Kaitlin Fier<sup>1,3</sup>, Karen Albright<sup>2,3</sup>, Susan Baird<sup>3</sup>, Brenda Crowe<sup>3,4</sup>, Linda Eres<sup>3</sup>, Marjorie Korn<sup>3</sup>, Leslie Maginn<sup>3</sup>, Mark McCormick<sup>3</sup>, Elisabeth D Root<sup>3,5</sup>, Thomas Vierzba<sup>3</sup>, Frederick S Wamboldt<sup>3,6</sup>  
and Jeffrey J Swigris<sup>1,3\*</sup>

### Abstract

**Background:** Little is known about whether or how supplemental oxygen affects patients with pulmonary fibrosis.

**Methods/Design:** A mixed-methods study is described. Patients with pulmonary fibrosis, informal caregivers of pulmonary fibrosis patients and practitioners who prescribe supplemental oxygen will be interviewed to gather data on perceptions of how supplemental oxygen impacts patients. In addition, three hundred pulmonary fibrosis patients who do not use daytime supplemental oxygen will be recruited to participate in a longitudinal, pre-/post- study in which patient-reported outcome (PRO) and activity data will be collected at baseline, immediately before daytime supplemental oxygen is initiated, and then once and again 9–12 months later. Activity data will be collected using accelerometers and portable GPS data recorders. The primary outcome is change in dyspnea from before to one month after supplemental oxygen is initiated. Secondary outcomes include scores from PROs to assess cough, fatigue and quality of life as well as the activity data. In exploratory analyses, we will use longitudinal data analytic techniques to assess the trajectories of outcomes over time while controlling for potentially influential variables.

# Parcours patient



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Published on

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## Pulmonary Rehabilitation and Noninvasive Ventilation in Patients with Hypercapnic Interstitial Lung Disease

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Ursula Schoenheit-Kenn<sup>d</sup> Sandra Winterkamp<sup>d</sup> Klaus Kenn<sup>d</sup>

<sup>a</sup>Department of Cardiology, Pneumology, Angiology and Intensive Care Medicine, University Hospital Aachen, Aachen,

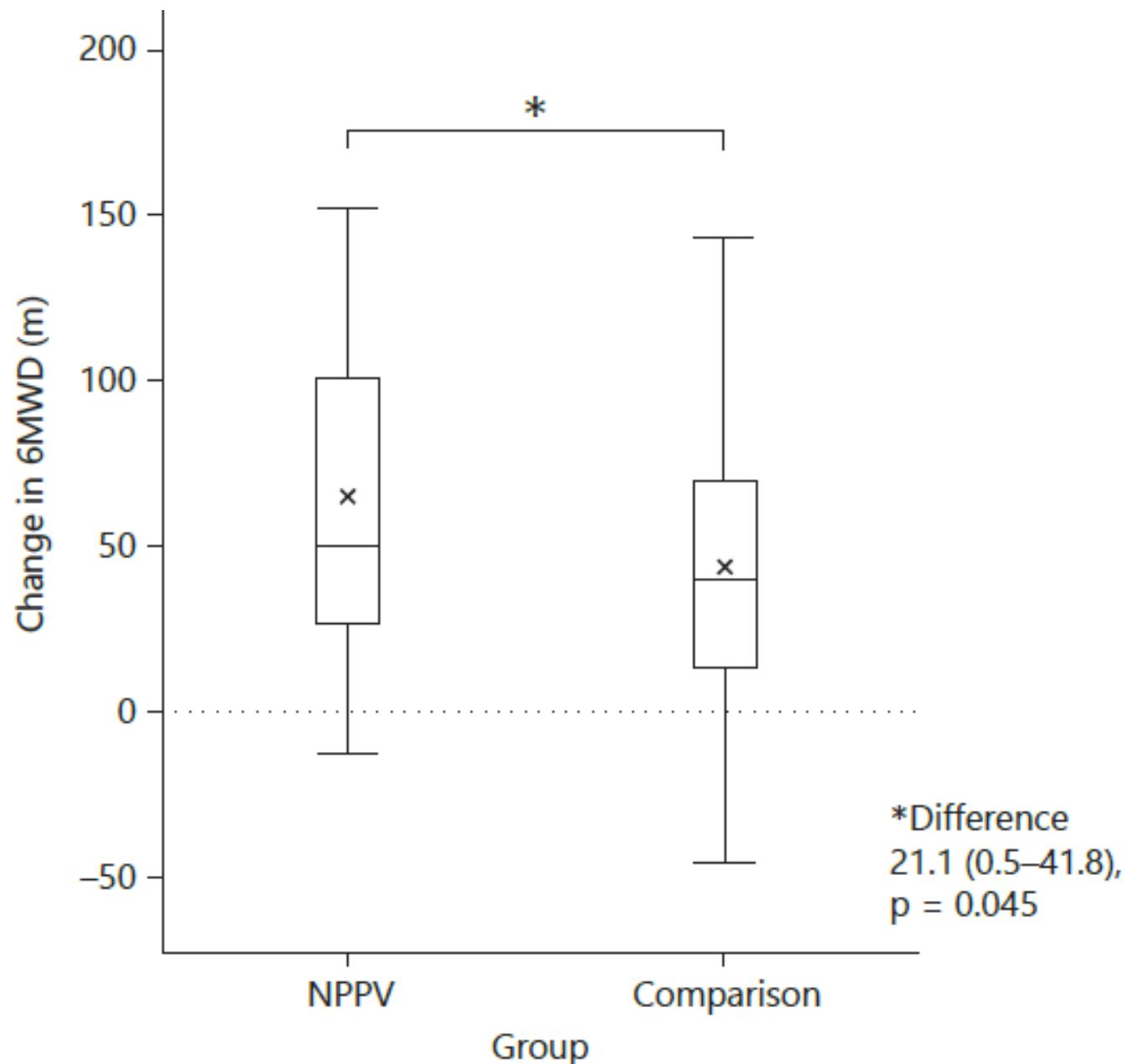
<sup>b</sup>Department of Pneumology and <sup>c</sup>Clinical Trials Unit, University Hospital Freiburg, Freiburg, and

<sup>d</sup>Department of Respiratory Medicine, Schoen-Klinik Berchtesgadener Land, Schoenau am Koenigssee, Germany

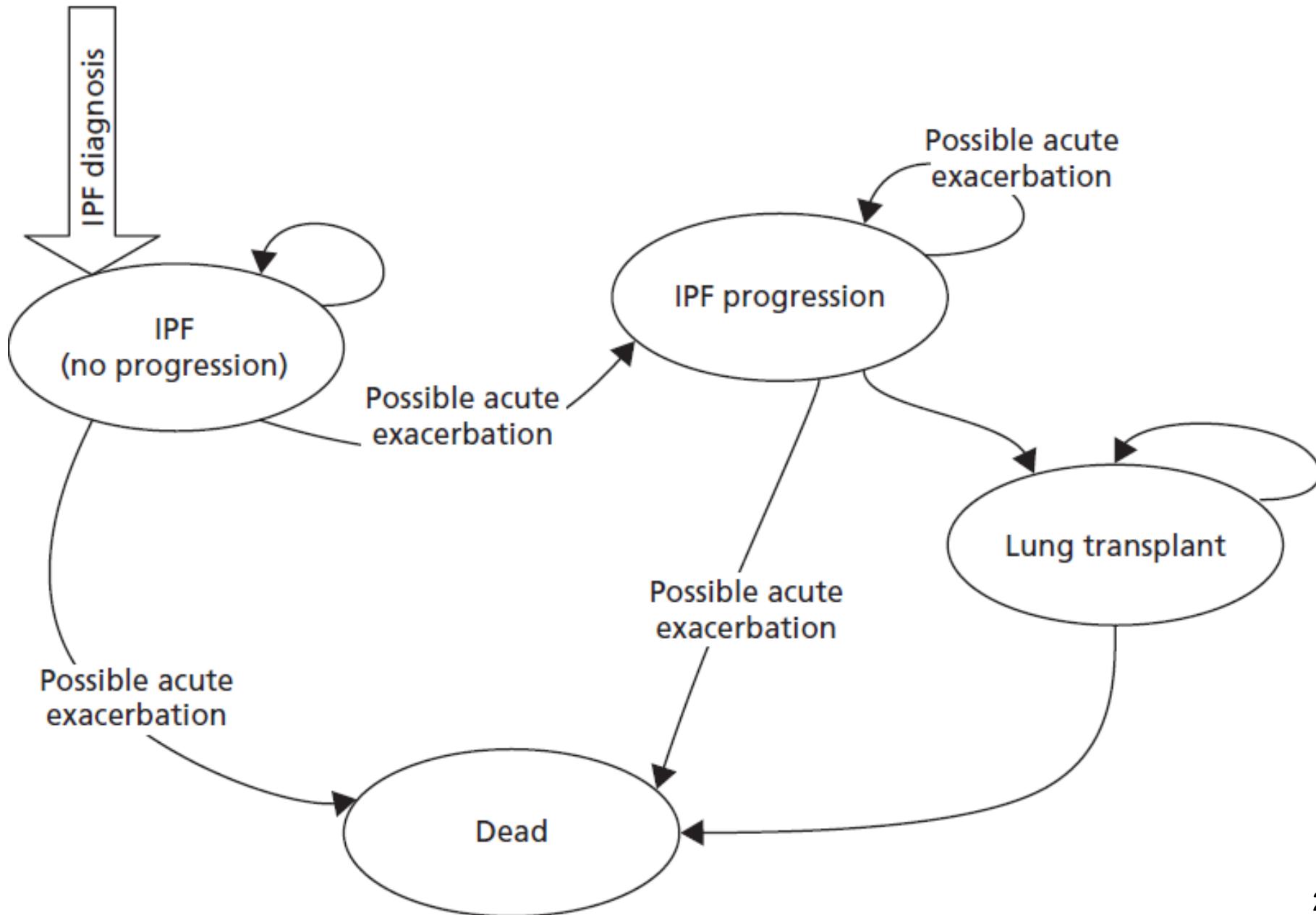
# Ventilation Non Invasive

	NPPV group	Comparison group
Total group	29	319
Idiopathic pulmonary fibrosis	10 (35)	181 (57)
Nonspecific interstitial pneumonia/ cryptogenic organizing pneumonia	2 (7)	18 (6)
Sarcoidosis	2 (7)	43 (14)
Hypersensitivity pneumonitis	8 (28)	49 (15)
Connective tissue disease	3 (10)	11 (3)
Radiogenic/drug-induced	3 (10)	6 (2)
Other	1 (3)	11 (3)

# Ventilation Non Invasive



# Parcours patient



# Palliation – Dyspnée

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An integrated palliative and respiratory care service for patients with advanced disease and refractory breathlessness: a randomised controlled trial



Irene J Higginson, Claudia Bausewein, Charles C Reilly, Wei Gao, Marjolein Gysels, Mendwas Dzingina, Paul McCrone, Sara Booth, Caroline J Jolley, John Moxham

## Summary

**Background** Breathlessness is a common and distressing symptom, which increases in many diseases as they progress and is difficult to manage. We assessed the effectiveness of early palliative care integrated with respiratory services for patients with advanced disease and refractory breathlessness.

*Lancet Respir Med* 2014  
2: 979–87  
Published Online  
October 29, 2014

# Palliation – Dyspnée

Time	Type of contact with clinic	Content of meeting
Week 1	First outpatient clinic visit	<p><b>Before visit:</b> Patients were offered free transport or if required disabled parking for the clinic appointments</p> <p><b>At visit</b></p> <ul style="list-style-type: none"><li>• welcome</li><li>• 6 minute walk test</li><li>• completion of Palliative care Outcome Scale by patient, to aid clinical assessment</li></ul> <p><b>Contact with respiratory medicine physician</b></p> <ul style="list-style-type: none"><li>• explore the symptom of breathlessness and its triggers</li><li>• establish underlying cause of breathlessness</li><li>• optimise disease-orientated management (check medications used correctly, appropriate treatments)</li><li>• review of previous investigations</li><li>• verbal and hand-written handover of notes from respiratory to palliative medicine physician to ensure patients do not have to repeat information</li></ul> <p><b>Contact with palliative medicine physician</b></p>

# Palliation – Dyspnée

## Contact with palliative medicine physician

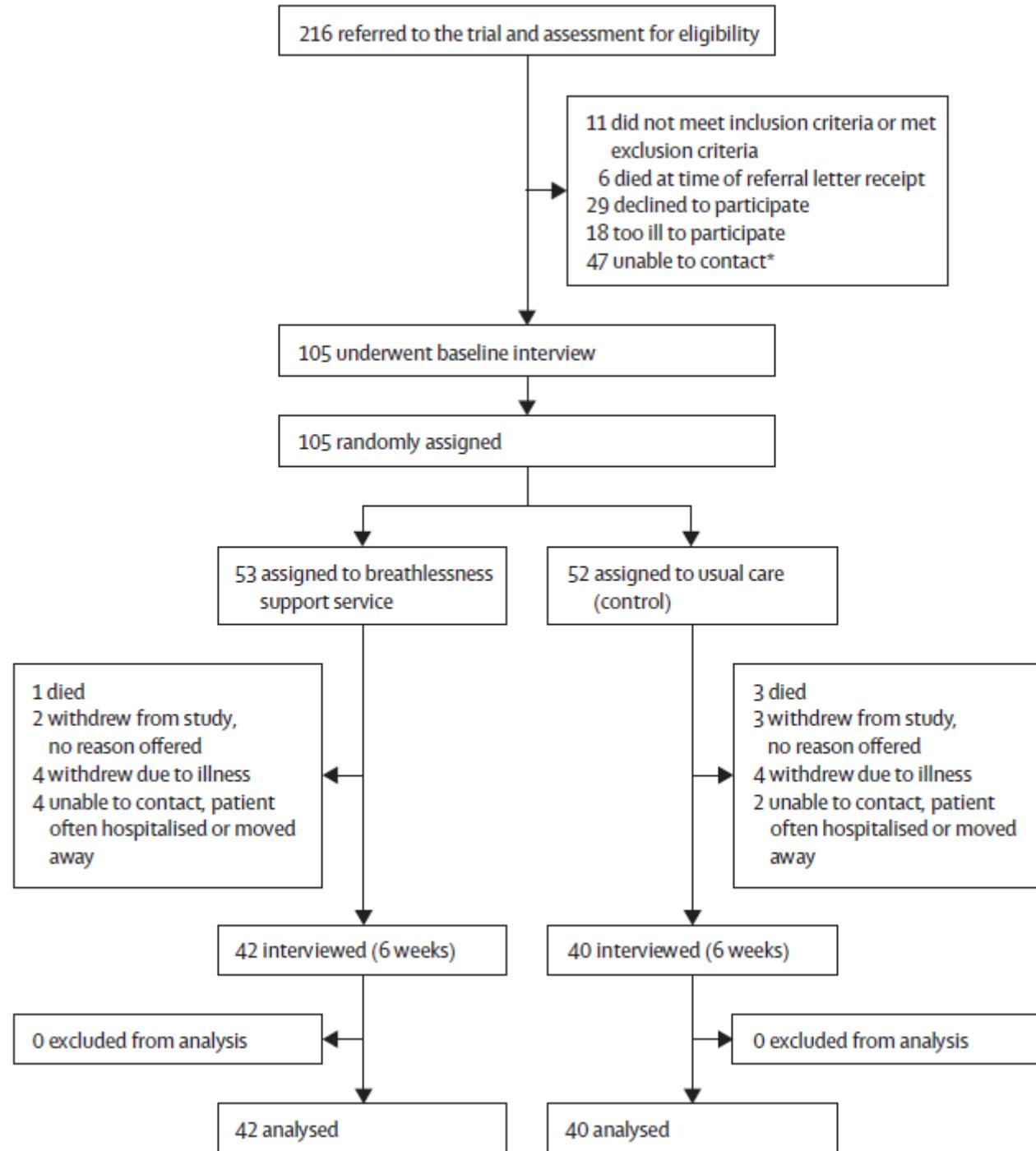
- experience of breathlessness
- development of crises plan
- burden on patient & family
- symptom burden (other than breathlessness), with recommendations to patients and GP of any appropriate treatments
- psychosocial & spiritual issues
- introduction of non-pharmacological measures such as the hand-held fan, water spray
- review together and provide breathlessness pack to take away, with information leaflets on managing breathlessness, a ‘poem’ (a mantra, laminated, to put up in the house and to read and follow when in acute breathlessness, developed by Jenny Taylor at St Christopher’s Hospice), a chart of positions, laminated, to use when in acute breathlessness, fan/water spray

**Following visit** - After each clinic appointment a letter was sent to the patient (to reinforce self-management) summarising the diagnosis, assessment results and plan for treatment, with a copy sent to the referring clinicians and the general practitioner. This and an e-mail were also sent to physiotherapy/occupational therapy to aid their visit. If required, urgent contact/phone call with the GP was made.

Week 2 – 3	Home visit	<p>Based on the patients' needs as assessed during clinic attendance and then home visit:</p> <p><b>Physiotherapy input</b></p> <ul style="list-style-type: none"> <li>• review of the positions of breathlessness</li> <li>• provision of a walking aid</li> <li>• breathing control techniques and anxiety-panic cycle</li> <li>• management of exacerbations in COPD</li> <li>• home programme of exercise (DVD, personalised sheet)</li> <li>• cough minimisation techniques</li> <li>• pacing and fatigue management</li> <li>• sputum clearance techniques</li> <li>• ambulatory oxygen assessments</li> <li>• referral to pulmonary rehabilitation</li> </ul> <p><b>Occupational therapy input</b></p> <ul style="list-style-type: none"> <li>• assessment of Activities of Daily Living (ADL) (mobility/transfers, self-care and domestic ADL)</li> <li>• assessment for aids and minor adaptations and referral for provision of equipment</li> <li>• wheelchair prescription</li> <li>• education on planning, pacing and energy conservation techniques to patients and carers</li> <li>• referral to other community services (local/out of area), as appropriate</li> <li>• assess the need for social support and liaison with the BSS</li> <li>• social worker, as appropriate</li> <li>• liaison with the BSS team regarding interventions and feedback</li> </ul>
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# Palliation – Dyspnée

Week 2-3	Telephone call	<b>Social worker input</b> <ul style="list-style-type: none"><li>• carer assessment including understanding of disease and symptoms &amp; information needs and coping strategies, if indicated at clinic assessment</li></ul>
Week 4 - 5	Second outpatient clinic visit	<b>Contact with palliative medicine physician</b> <ul style="list-style-type: none"><li>• re-evaluation of breathlessness and other symptoms</li><li>• check use of fan, spray, pack, DVD etc, further guidance given</li><li>• change of medications recommended if required, with contact with GP regarding future planned treatments if required</li><li>• referral to medical and/or palliative care services if appropriate</li><li>• discharge from service</li><li>• provided with information on drop-in patient/family information centre for further resources</li></ul> <b>Following visit</b> - After the clinic appointment a letter was sent to the patient (to reinforce self-management) summarising the progress made, further recommendations and plan for treatment, with a copy sent to the referring clinicians and the general practitioner.



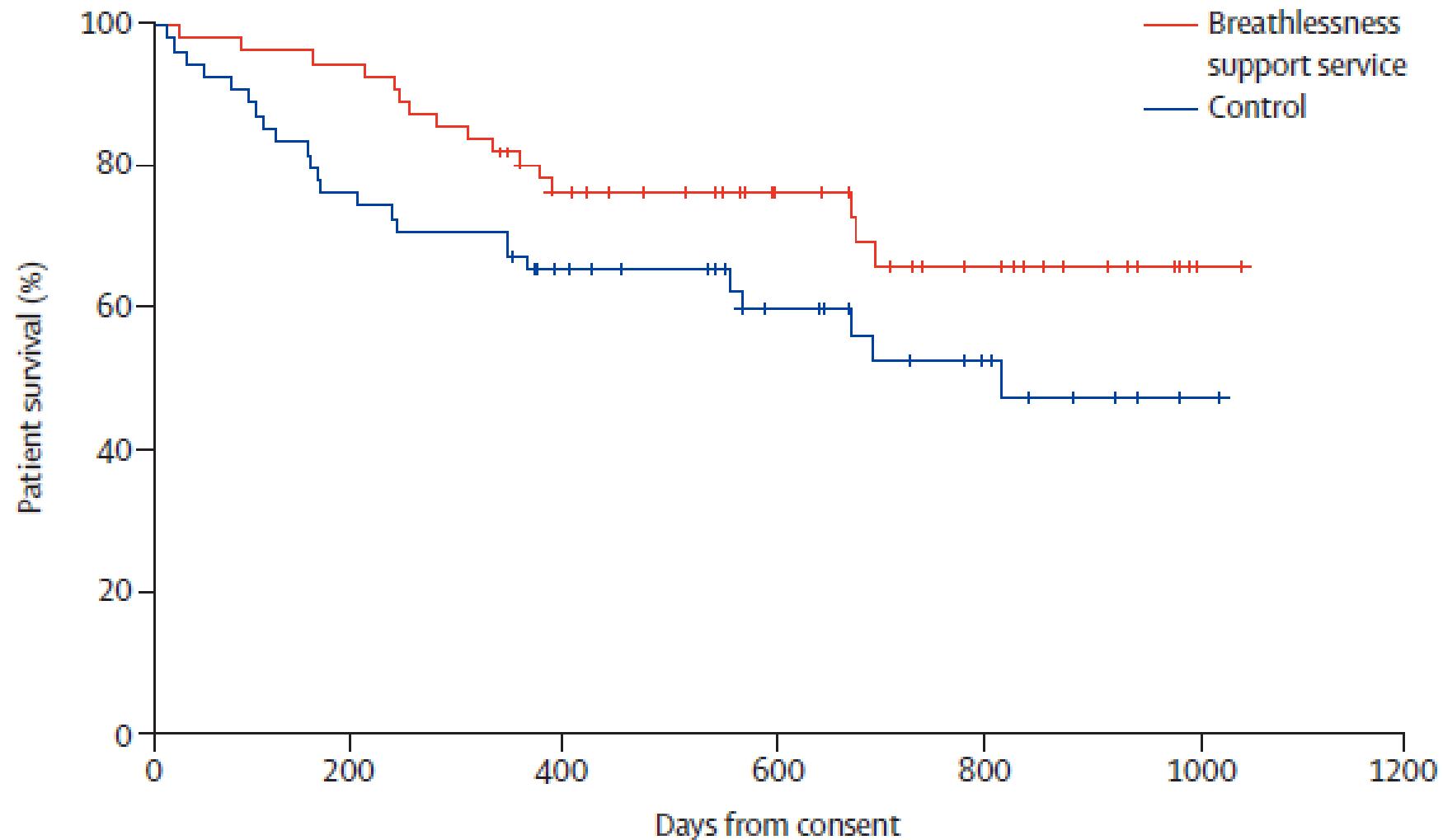
# Palliation – Dyspnée

	Overall (n=105)	Breathlessness support service group (n=53)	Control group (n=52)
Age (years)	67 (10)	66 (11)	68 (11)
Sex			
Men	61 (58%)	28 (53%)	33 (63%)
Women	44 (42%)	25 (47%)	19 (37%)
Diagnosis			
Chronic obstructive pulmonary disease	57 (54%)	29 (55%)	28 (54%)
Cancer*	21 (20%)	11 (21%)	10 (19%)
Interstitial lung disease	19 (18%)	7 (13%)	12 (23%)
Heart failure	5 (5%)	4 (8%)	1 (2%)
Other†	3 (3%)	2 (4%)	1 (2%)
Has carer or family member			
Yes	75 (71%)	38 (72%)	37 (71%)
No	30 (29%)	15 (28%)	15 (29%)
Clinical characteristics			
FEV <sub>1</sub> (L)‡	1.25 (0.70)	1.3 (0.78)	1.2 (0.65)
Predicted FEV <sub>1</sub> (%)‡	46.2 (23.3)	48.0 (24.3)	44.5 (22.4)
VC (L)‡	1.9 (0.96)	2.0 (1.0)	1.8 (0.9)
Predicted VC (%)‡	57.9 (25.7)	59.3 (25.5)	56.6 (26.0)

# Palliation – Dyspnée

	Breathlessness support service group (n=42)	Control group (n=40)	Difference between breathlessness support service and control (95% CI)	p value
Primary outcome (CRQ mastery)*†	4·15 (1·7)	3·57 (1·4)	0·58 (0·01 to 1·15)	0·048
Secondary outcomes				
NRS breathlessness average 24 h‡	5·38 (2·2)	5·71 (2·1)	-0·33 (-1·28 to 0·62)	0·49
NRS breathlessness worst at rest 24 h‡	4·12 (2·8)	4·47 (3·3)	-0·35 (-1·71 to 1·01)	0·61
NRS breathlessness on exertion 24 h‡	7·45 (2·4)	8·18 (1·8)	-0·73 (-1·69 to 0·22)	0·13
CRQ HRQL*	71 (19)	67 (20)	4·21 (-4·52 to 12·94)	0·34
CRQ dyspnoea*†	2·54 (1·1)	2·46 (0·9)	0·08 (-0·38 to 0·52)	0·75
CRQ emotion*†	4·07 (1·3)	3·93 (1·3)	0·14 (-0·42 to 0·71)	0·16
CRQ fatigue*†	3·09 (1·1)	3·07 (1·5)	0·02 (-0·56 to 0·62)	0·93
EQ-5D index*	0·44 (0·31)	0·35 (0·29)	0·092 (-0·23 to 0·4)	0·18
EQ-5D HRQL VAS*	56 (20)	55 (18)	1 (-6·67 to 10·34)	0·67
LCADL total score‡	45 (13)	50 (15)	-5 (-12·22 to 1·02)	0·10
POS total score‡	12·15 (6·8)	12·42 (6·5)	-0·27 (-3·29 to 2·75)	0·86
HADS anxiety‡	9·2 (2·8)	9·1 (2·7)	0·1 (-0·93 to 1·24)	0·78
HADS depression‡	10 (2·8)	11 (2·5)	-1 (-1·82 to 0·30)	0·16

# Palliation – Dyspnée



## Number at risk

	53	50	35	24	13	4
Breathlessness	53	50	35	24	13	4
support service						
Control	52	38	28	18	11	4

# Efficacité et Cout-Efficacité

## Abstract

### The clinical effectiveness and cost-effectiveness of treatments for idiopathic pulmonary fibrosis: a systematic review and economic evaluation

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## Efficacité et Cout-Efficacité

Fourteen studies were included in the review of clinical effectiveness, of which one evaluated azathioprine, three N-acetylcysteine (NAC) (alone or in combination), four pirfenidone, one BIBF 1120, one sildenafil, one thalidomide, two pulmonary rehabilitation, and one a disease management programme.

The model base-case results show increased survival for five pharmacological treatments, compared with best supportive care, at increased cost.

# Efficacité et Cout-Efficacité

**Limitations:** Few direct comparisons of treatments were identified. An indirect comparison through a

Few interventions have any statistically significant effect on IPF and a lack of studies on palliative care approaches was identified. Research is required into the effects of symptom control interventions, in particular pulmonary rehabilitation and thalidomide

once the results of ongoing studies are reported.

**Study registration:** This study is registered as PROSPERO CRD42012002116.

**Funding:** The National Institute for Health Research Health Technology Assessment programme.

# Parcours patient

