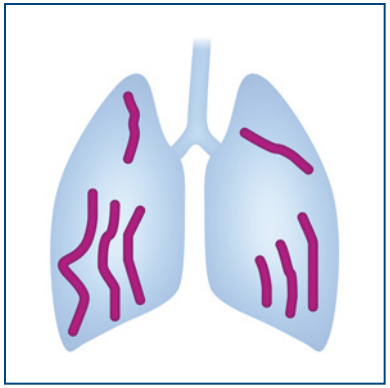
Pneumo Update Europe 2018

15 - 16 June, Budapest

Interstitial Lung Diseases



Luca Richeldi, Italy

Diagnosis of IPF

Diagnostic criteria for idiopathic pulmonary fibrosis: a Fleischner Society White Paper

David A Lynch, Nicola Sverzellati, William D Travis, Kevin K Brown, Thomas V Colby, Jeffrey R Galvin, Jonathan G Goldin, David M Hansell, Yoshikazu Inoue, Takeshi Johkoh, Andrew G Nicholson, Shandra L Knight, Suhail Raoof, Luca Richeldi, Christopher J Ryerson, Jay H Ryu, Athol U Wells

Lynch et al.,Lancet Resp Med, 2018; 6: e7

CLINICAL CHECKLIST FOR ALTERNATIVE DIAGNOSES

General

- What are the severity, duration, and pace of the primary respiratory symptoms?

Systemic autoimmune disease

- Are symptoms or signs of a systemic autoimmune disorder present?
- Are serological findings suggestive of an autoimmune disorder? Eg, rheumatoid arthritis, systemic sclerosis, polymyositis and dermatomyositis, Steven-Johnson syndrome, or mixed-connective tissue disease.

Other systemic disease (sarcoid, immune-system abnormalities)

- Is there evidence of other organ involvement?

Hypersensitivity pneumonitis

- Does the patient have a clinically relevant exposure to an antigen, generally inhaled, known to result in the development of hypersensitivity pneumonitis?
- Do they have pets, including birds?
- What are they exposed to in their home or work environment? Is there water damage?
- Is the exposure clinically significant?
- Is the intensity clinically significant?
- Is there a temporal association between the exposure and symptom onset?

Occupational and environmental lung disease

- Does the patient work in an occupation known to be at risk for the development of lung disease?
- What do they do in their current job and previous jobs?
- What avocational exposures exist?

Drug-induced lung disease

- Does the patient use any medicines, herbs, vitamins, supplements, or recreational drugs that could account for the presence of lung disease?

Specific genetic syndromes

- Is there a family history of lung fibrosis?
- Is there evidence of premature graying, cryptogenic cirrhosis, aplastic anaemia, myelodysplasia, macrocytosis, or thrombocytopenia?

Lynch et al.,Lancet Resp Med, 2018; 6: e7

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Lynch et al.,Lancet Resp Med, 2018; 6: e7

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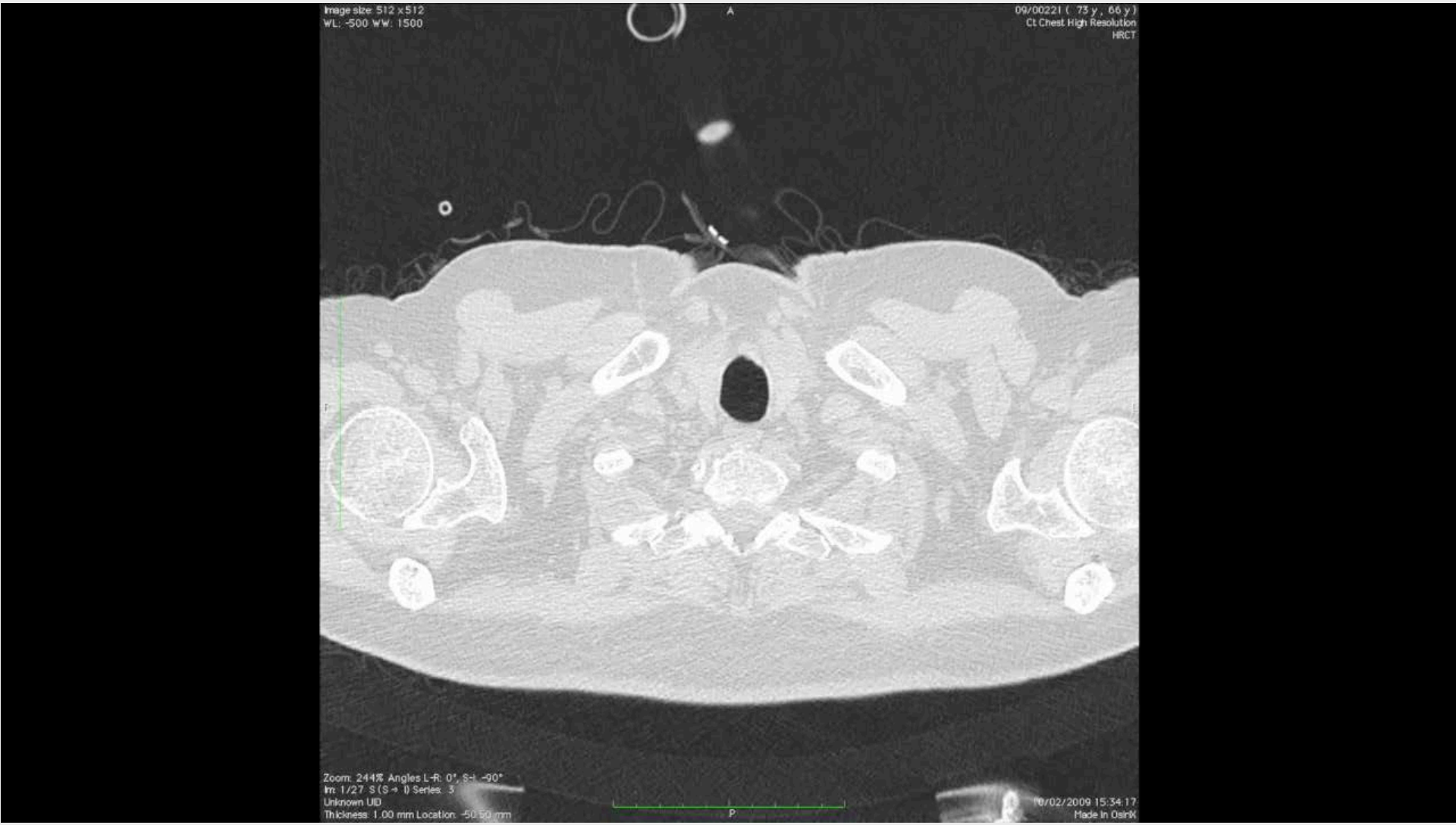
CHEST HRCT



Courtesy Simon LF Walsh, London

Richeldi L et al, *Lancet* 2017; 389: 1941-1952

DIAGNOSTIC CATEGORIES OF UIP BASED ON CT PATTERNS

	Typical UIP CT pattern	Probable UIP CT pattern	CT pattern indeterminate for UIP	CT features most consistent with non-IPF diagnosis
Distribution	Basal predominant (occasionally diffuse), and subpleural predominant; distribution is often heterogeneous	Basal and subpleural predominant; distribution is often heterogeneous	Variable or diffuse	Upper-lung or mid-lung predominant fibrosis; peribronchovascular predominance with subpleural sparing
Features	Honeycombing; reticular pattern with peripheral traction bronchiectasis or bronchiolectasis*; absence of features to suggest an alternative diagnosis	Reticular pattern with peripheral traction bronchiectasis or bronchiolectasis*; honeycombing is absent; absence of features to suggest an alternative diagnosis	Evidence of fibrosis with some inconspicuous features suggestive of non-UIP pattern	Any of the following: predominant consolidation, extensive pure ground glass opacity (without acute exacerbation), extensive mosaic attenuation with extensive sharply defined lobular air trapping on expiration, diffuse nodules or cysts

Lynch et al.,Lancet Resp Med, 2018; 6: e7

HISTOPATHOLOGICAL CRITERIA FOR UIP IN IPF (UIP-IPF)

	Definite UIP-IPF	Probable UIP-IPF	Indeterminate for UIP-IPF	Features most consistent with an alternative diagnosis
General comments	Patients show features with all four criteria, and do not show features that might suggest an alternative diagnosis (eg, non-UIP)	Patients show either honeycomb fibrosis only, or a severe fibrosing process that falls short of showing all four criteria for definite UIP-IPF and do not show features that might suggest an alternative diagnosis	Patients show evidence of a fibrosing process but with features that are more in favour of either a non-UIP pattern, or UIP in a setting other than IPF	Patients show either a UIP pattern with ancillary features strongly suggesting an alternative diagnosis, or a non-UIP pattern (see cell below)
Specific criteria	Dense fibrosis causing architecture remodelling with frequent honeycombing; patchy lung involvement by fibrosis; subpleural or paraseptal distribution, or both; fibroblast foci at the edge of dense scars	Honeycomb fibrosis only or; dense fibrosis causing architecture remodelling with frequent honeycombing; patchy lung involvement by fibrosis; fibroblast foci at the edge of dense scars may or may not be present	Patients have less compelling histological changes than those classified by the final column (eg, occasional foci of centrilobular injury or scarring, rare granulomas or giant cells, only a minor degree of lymphoid hyperplasia or diffuse inflammation, or diffuse homogenous fibrosis favouring fibrotic non-specific interstitial pneumonia); these features, and the differential diagnoses they call to mind, become part of the multidisciplinary discussion and decision with regard to a multidisciplinary diagnosis of IPF, or not	Non-UIP pattern: patients with features of other fibrotic disorders—eg, fibrotic hypersensitivity pneumonitis, fibrotic non-specific interstitial pneumonia, fibrosing organising pneumonia, pleuroparenchymal fibroelastosis, pulmonary Langerhans cell histiocytosis, or smoking-related interstitial fibrosis; UIP pattern with ancillary features strongly suggesting an alternative diagnosis: eg, prominent diffuse alveolar damage or organising pneumonia (consider acute exacerbation of UIP), granulomas, (consider hypersensitivity pneumonitis, sarcoid, infection), marked interstitial inflammatory cell infiltrate away from areas of UIP (consider hypersensitivity pneumonitis)

SAME CATEGORIES FOR IMAGING AND PATHOLOGY

Lynch et al.,Lancet Resp Med, 2018; 6: e7

Role of cryobiopsy

- Tissue specimen larger than transbronchial biopsy, but usually smaller than surgical biopsy. Also, not typically subpleural.
- Diagnostic accuracy for all ILD about 80%, may be lower for UIP
- Diagnostic yield and complication rate is variable and depend on the experience of the operator
- Experience is higher in Europe
- Role remains **unclear**

Lynch DA et al, *Lancet Resp Med* 2018

PATHWAYS TO A CONFIDENT WORKING MULTIDISCIPLINARY DIAGNOSIS OF IPF

When can one make a confident diagnosis of IPF without biopsy?

- Clinical context of IPF*, with CT pattern of typical or probable UIP

When is a diagnostic biopsy necessary to make a confident diagnosis of IPF?

- Clinical context of IPF* with CT pattern either indeterminate or suggestive of an alternative diagnosis
- Clinical context indeterminate for IPF† with any CT pattern

When is multidisciplinary diagnosis necessary in the context of suspected IPF?

- When the clinical context or the CT pattern, or both, are indeterminate; the outcome of multidisciplinary discussion will be a decision whether to perform an additional clinical evaluation, bronchoalveolar lavage, or diagnostic biopsy, or some combination of these procedures
- After biopsy, to integrate the clinical, imaging, and histological features
- To re-review patients in whom the longitudinal course of disease is discordant with the previously established multidisciplinary diagnosis
- When diagnostic tissue is not available, to consider a working diagnosis of IPF

What should be done when diagnostic tissue is not available?

- Multidisciplinary diagnosis with consideration of the patient's age, sex, smoking status, findings on bronchoalveolar lavage, and longitudinal disease behaviour
- In this context, a working diagnosis of IPF can be made in the presence of a progressive fibrosing interstitial pneumonia, and in the absence of an alternative explanation; the level of diagnostic confidence of such a working diagnosis should be recorded, and the diagnosis should be reviewed at regular intervals, since it might change over time

Lynch et al.,Lancet Resp Med, 2018; 6: e7

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Clinical context of IPF includes *all* of the following:

- **older than 60 years**
- **absence of clinically significant environmental or medication exposure**
- **no evidence of connective tissue disease**

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Lynch et al.,Lancet Resp Med, 2018; 6: e7

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- Clinical context of IPF* with CT pattern either indeterminate or suggestive of an alternative diagnosis
- Clinical context indeterminate for IPF† with any CT pattern

Clinical context *indeterminate* for IPF includes *any* of the following:

- **aged 60 years or younger**
- **potentially significant environmental or medication exposure**
- **evidence of connective tissue disease**

Lynch et al.,Lancet Resp Med, 2018; 6: e7

PATHWAYS TO A CONFIDENT WORKING MULTIDISCIPLINARY DIAGNOSIS OF IPF

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Lynch et al.,Lancet Resp Med, 2018; 6: e7

RECOMMENDATIONS FOR MULTIDISCIPLINARY DIAGNOSIS CONFERENCES

	Desirable features
Frequency	Weekly to monthly, depending on volume of patients
Patient selection	Focus on patients with disease that is not fully characterised, and those with suspicion of a non-IPF aetiology (eg, hypersensitivity pneumonitis, connective tissue disease); in experienced groups with a high volume of patients, those with typical features might not require review; selected patients might also be re-reviewed on follow-up
Nature of conference	Direct contact or telemedicine; pathology and radiology should be directly visualised
Participants	Clinician, radiologist, and pathologist with interest or experience in ILD; if not experienced, linkage to an experienced group is needed (eg, electronic transmission of images, review of slides, telephone or e-mail discussion of clinical aspects); a rheumatologist is often helpful
Goals of meeting	Diagnosis, management plan, review of disease progression
Documentation	First choice multidisciplinary diagnosis (including "unclassifiable disease"), realistic differential diagnoses, likely reversibility; recommendations on additional diagnostic tests
Communication	Final multidisciplinary diagnosis recorded in case notes and communicated in discharge statements; could also include list of conference participants, clinical, radiological, and pathological diagnoses, and management recommendations

Lynch et al.,Lancet Resp Med, 2018; 6: e7

Summary

- Fleischner Society diagnostic criteria permit diagnosis of IPF *in a wider spectrum of patients:*
  - with probable UIP CT pattern
  - with indeterminate CT patterns
    - if a lung biopsy is performed *or*
    - if a confident working diagnosis can be made

Lynch DA et al, *Lancet Resp Med* 2018

Therapy of IPF

CONCEPTUAL MODEL OF IPF ACROSS AN INDIVIDUAL'S LIFE COURSE

- Genetic predisposition
- Smoking, occupational dust exposure, and other risk factors
- Subclinical disease (Velcro-type crackles)
- Onset of symptoms
- Diagnosis (typically age 60–70 years)
- Disease-modifying therapy
- Comprehensive management

Richeldi L et al, *Lancet* 2017; 389: 1941-1952

Panel: Therapies identified in clinical trials as harmful, ineffective, or effective in the treatment of idiopathic pulmonary fibrosis

Potentially harmful therapies

- Ambrisentan[81]
- Everolimus[82]
- Prednisolone, azathioprine, acetylcysteine[9]
- Warfarin[83]

Potentially ineffective therapies

- Bosentan[84]
- Imatinib[85]
- Macitentan[86]
- Acetylcysteine[87]
- Sildenafil[88]

Effective disease-modifying therapies

- Nintedanib[89]
- Pirfenidone[90,91]

Lancet Resp Med, 2018; 6: e7

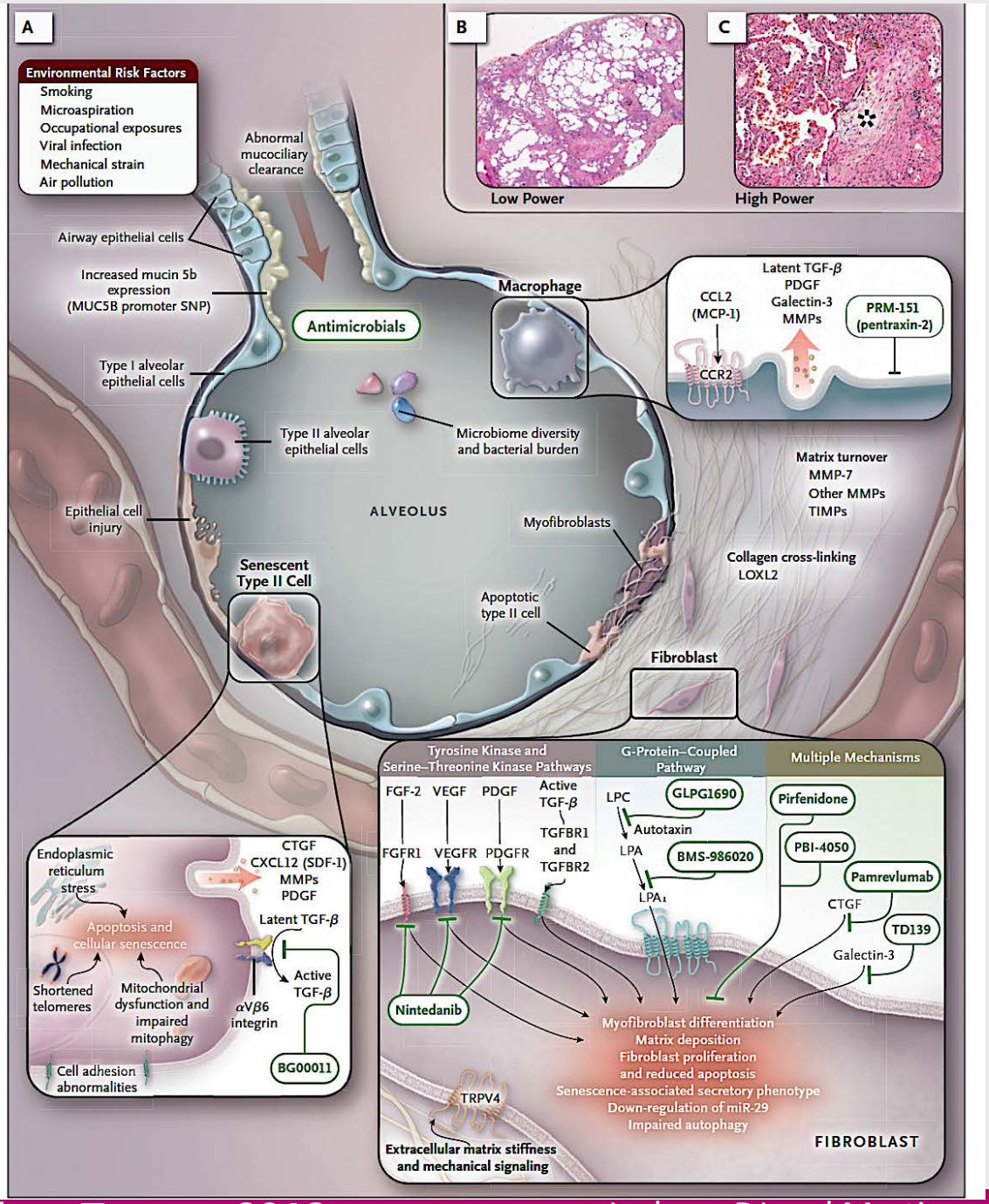
STATE OF THE ART

Dan L. Longo, M.D., *Editor*

Idiopathic Pulmonary Fibrosis

David J. Lederer, M.D., and Fernando J. Martinez, M.D.

Lederer DJ and Martinez FJ, *N Engl J Med* 2018



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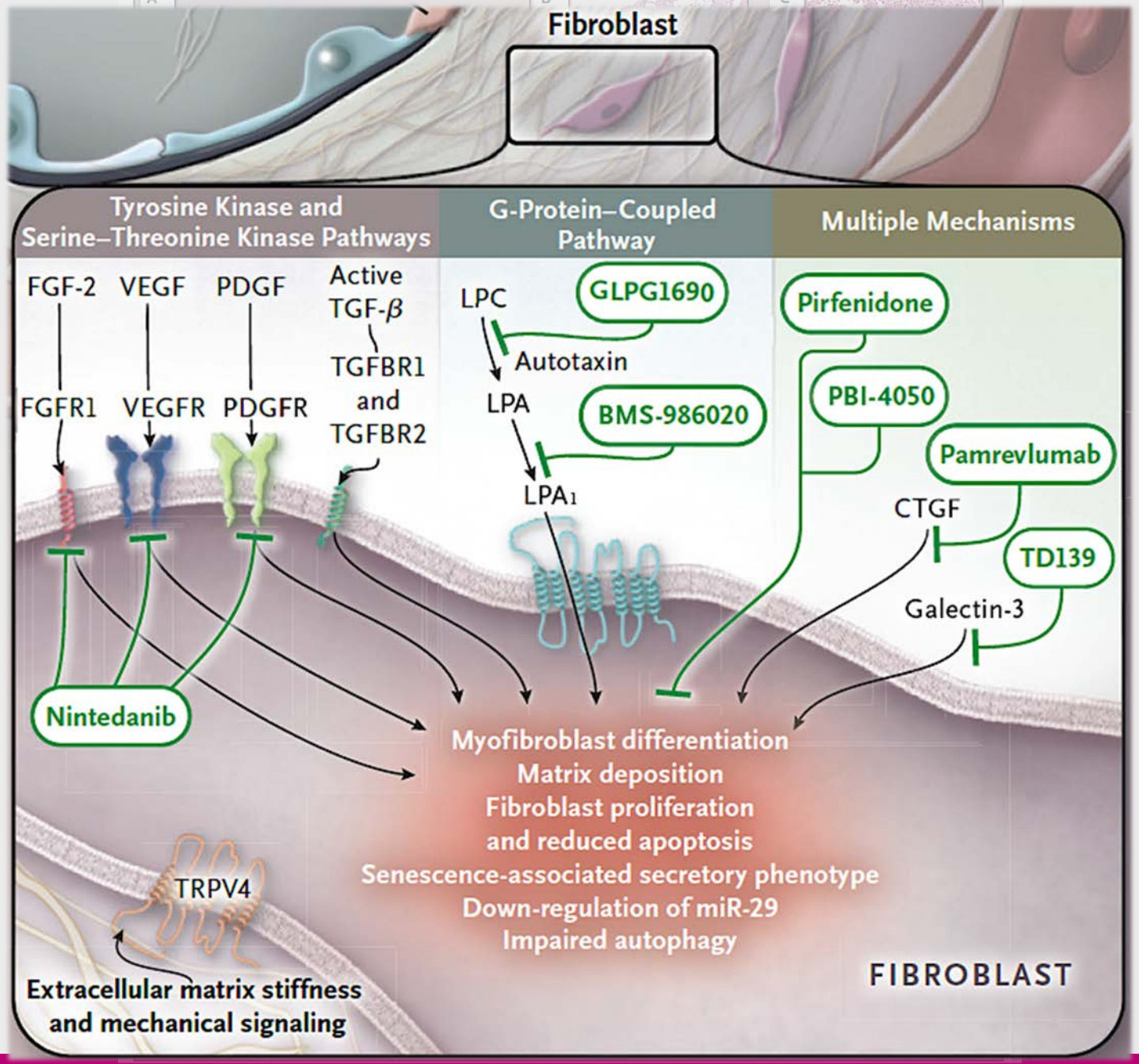

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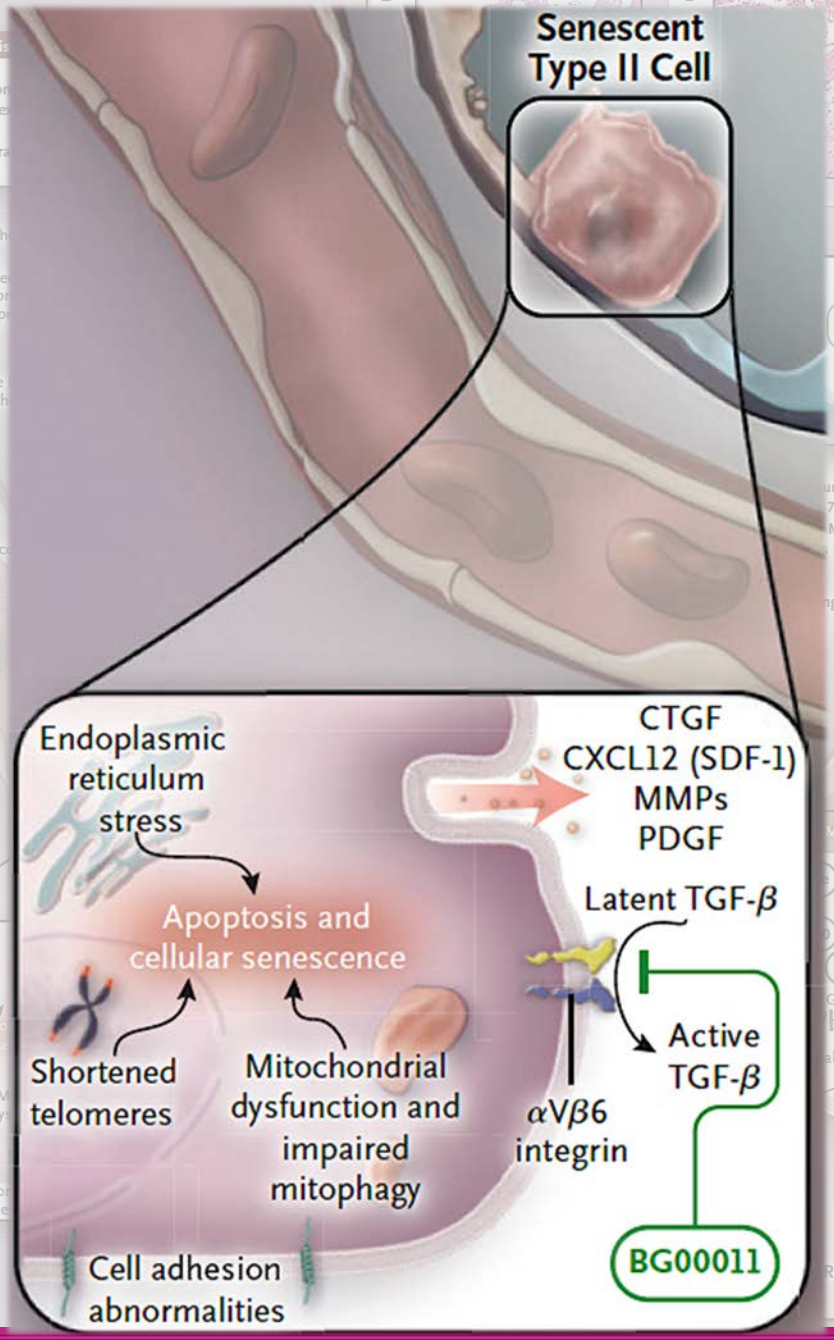

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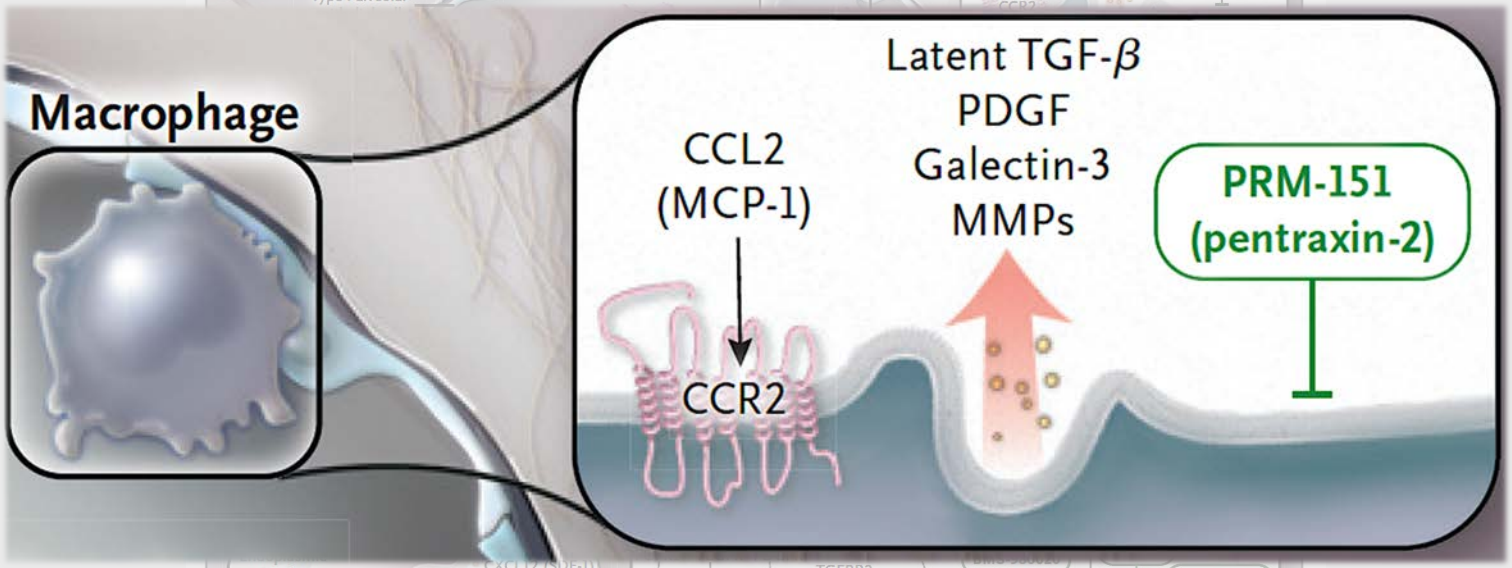


Lederer DJ and Martinez FJ, *N Engl J Med* 2018

ATS 2018 San Diego

Research

JAMA | Preliminary Communication

Effect of Recombinant Human Pentraxin 2 vs Placebo on Change in Forced Vital Capacity in Patients With Idiopathic Pulmonary Fibrosis

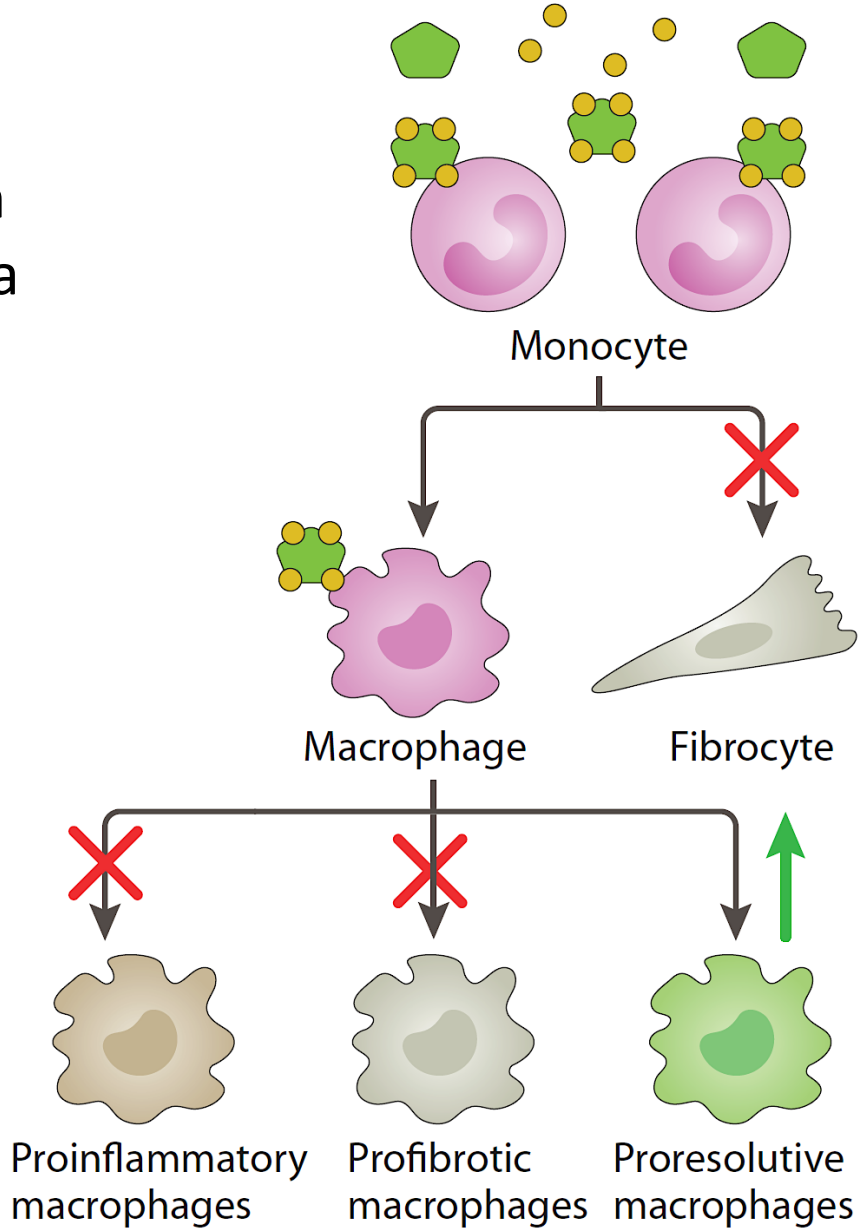
A Randomized Clinical Trial

Ganesh Raghu, MD; Bernt van den Blink, MD, PhD; Mark J. Hamblin, MD; A. Whitney Brown, MD; Jeffrey A. Golden, MD; Lawrence A. Ho, MD; Marlies S. Wijsenbeek, MD; Martina Vasakova, MD, PhD; Alberto Pesci, MD; Danielle E. Antin-Ozerkis, MD; Keith C. Meyer, MD; Michael Kreuter, MD; Hugues Santin-Janin, PhD; Geert-Jan Mulder, MD; Brian Bartholmai, MD; Renu Gupta, MD; Luca Richeldi, MD

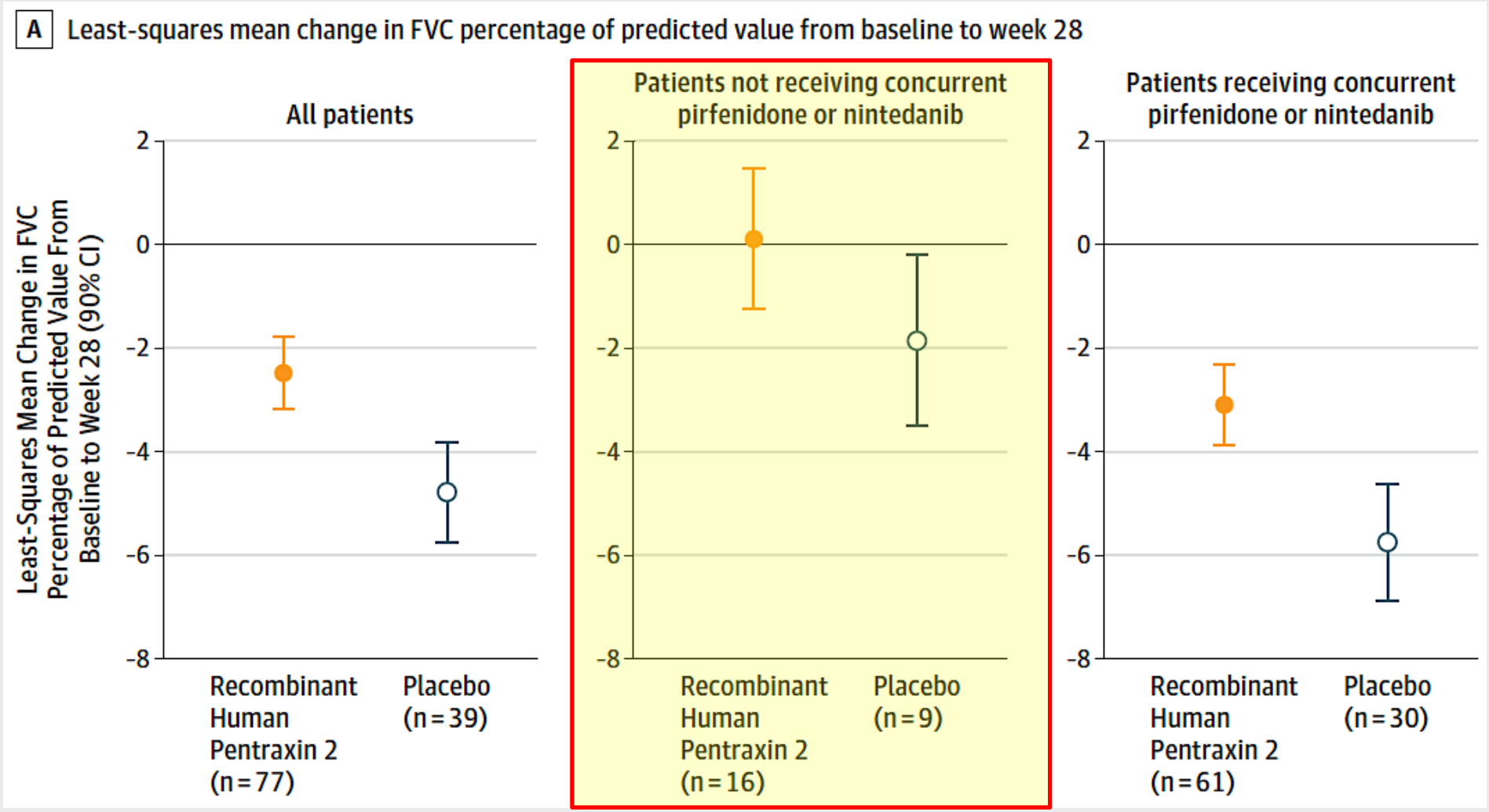
Raghu et al., JAMA. 2018;319(22):2299-2307

PENTRAXIN

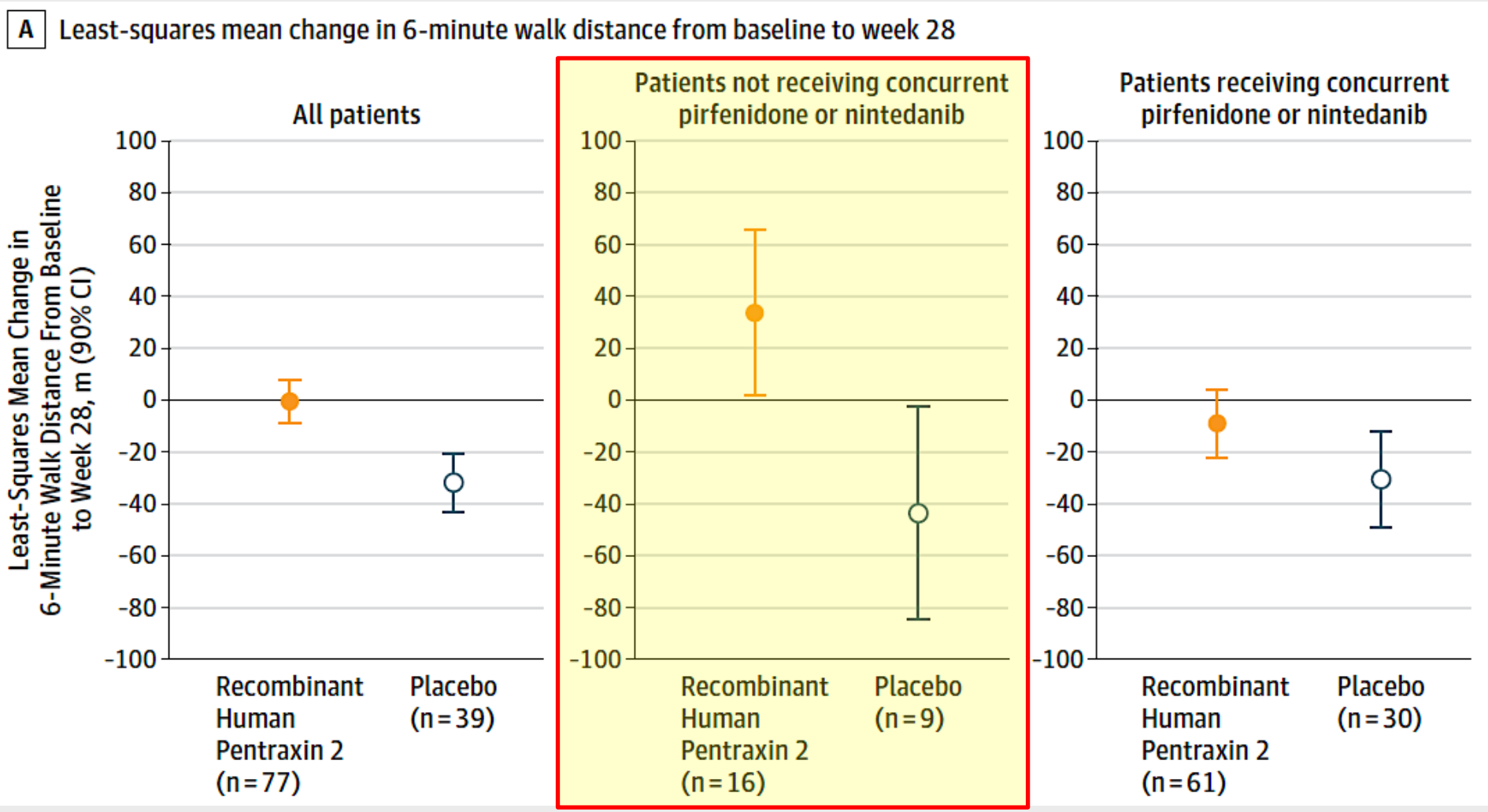
- PTX-2 (aka Serum Amyloid P) is an **innate immune regulatory** plasma protein.
- PTX-2 binds Danger-Associated Molecular Patterns (**DAMPs**), which have specificity for damaged tissue.
- PTX-2 binds to **monocyte and macrophage** FcγR, blocks fibrocyte and profibrotic / proinflammatory macrophage production.



Raghu et al., JAMA. 2018;319(22):2299-2307



JAMA doi:10.1001/jama.2018.6129, Published online May 20, 2018



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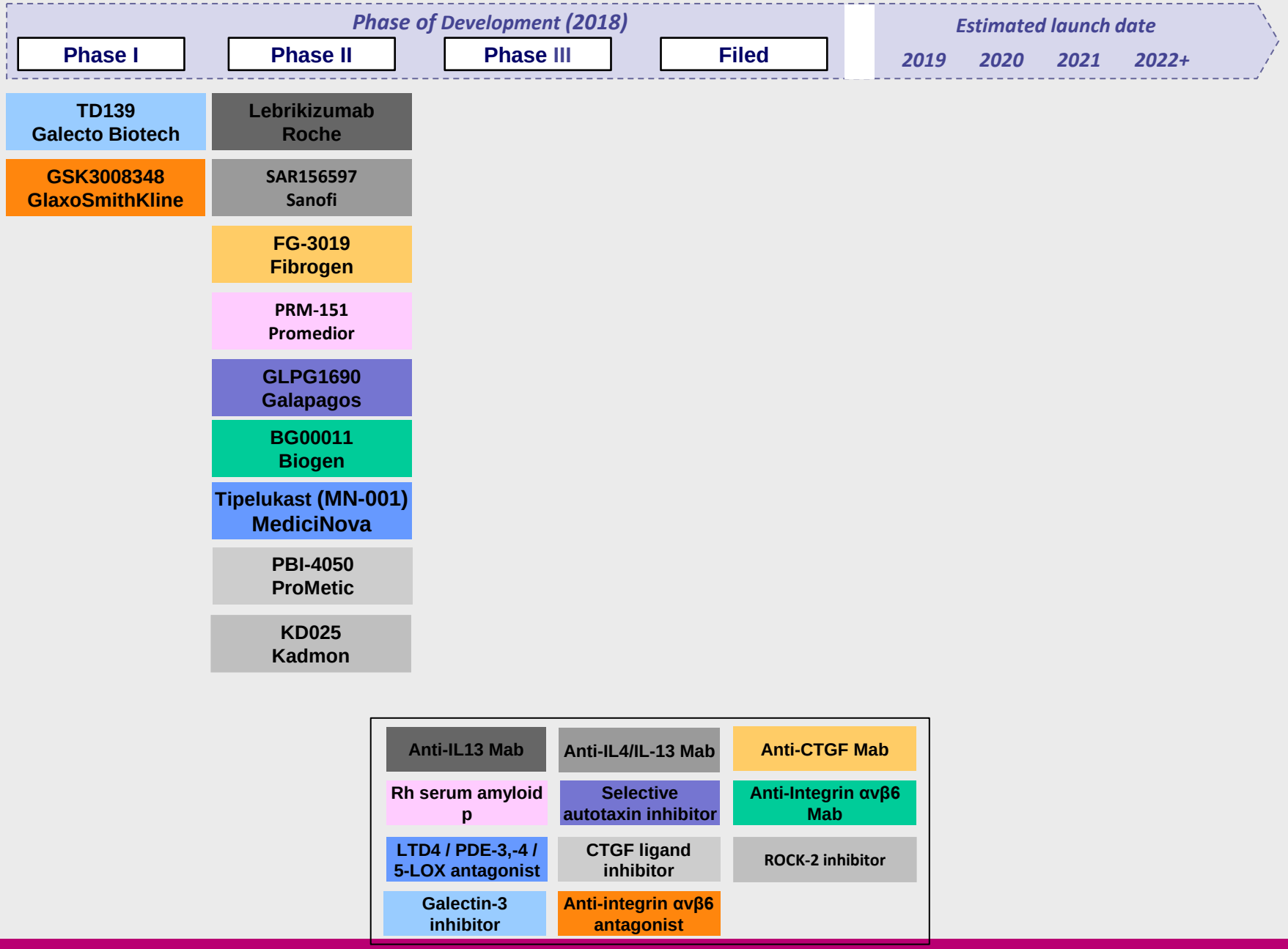
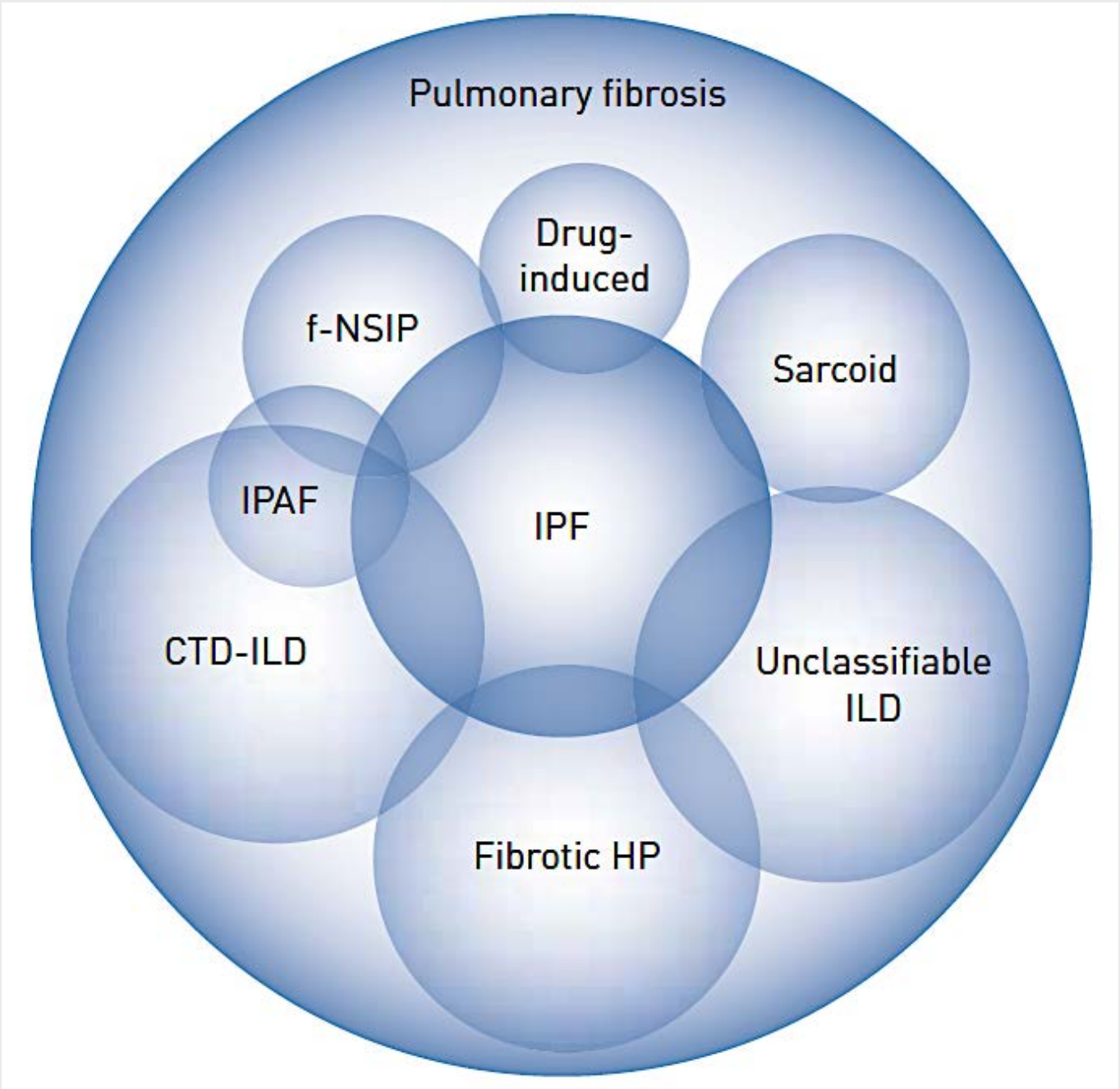
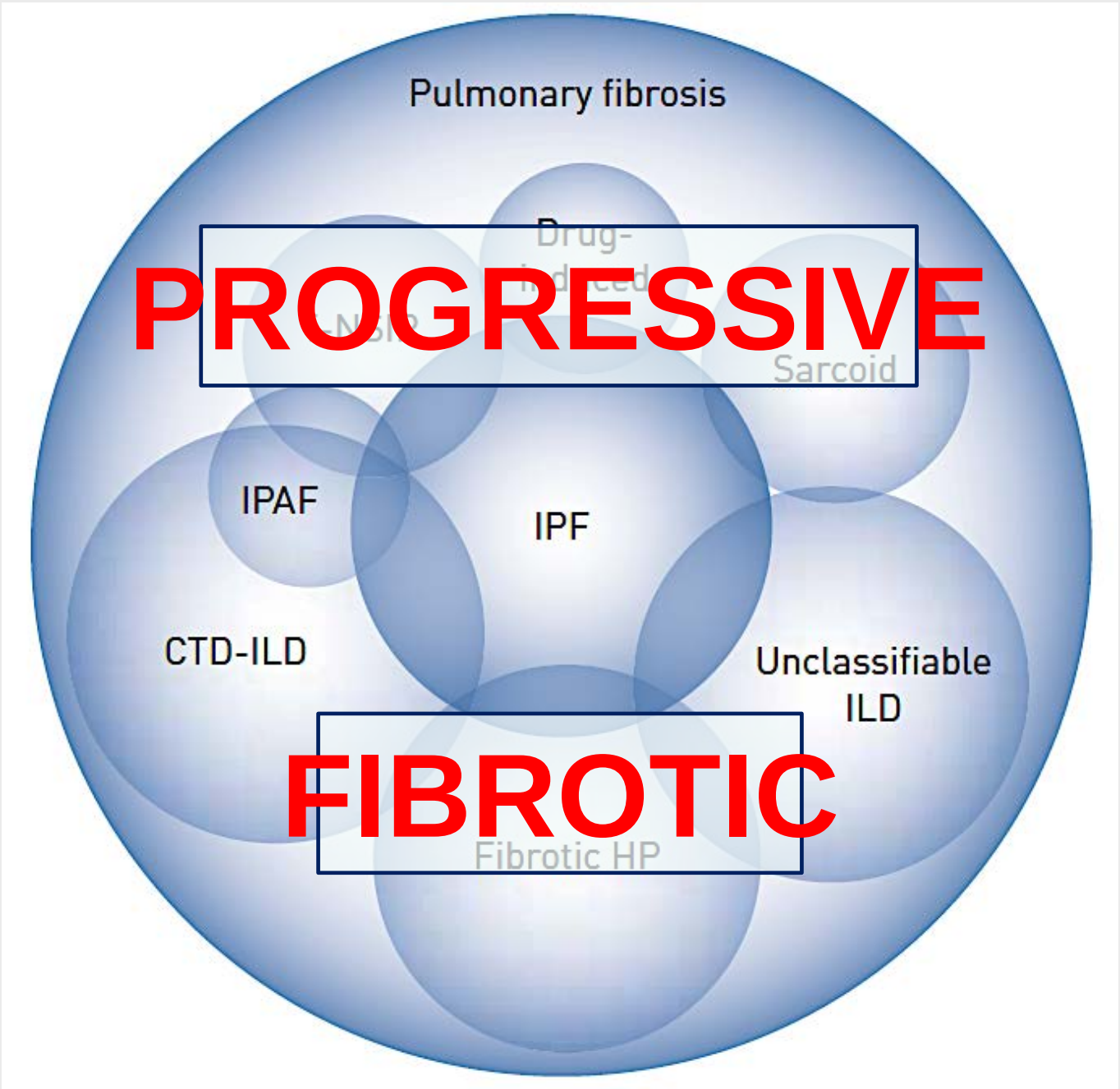

Phase of Development (2018)
Phase I
Phase II
Phase III
Filed
Estimated launch date
2019
2020
2021
2022+
TD139
Galecto Biotech
GSK3008348
GlaxoSmithKline
Lebrikizumab
Roche
SAR156597
Sanofi
FG-3019
Fibrogen
PRM-151
Promedior
GLPG1690
Galapagos
BG00011
Biogen
Tipelukast (MN-001)
MediciNova
PBI-4050
ProMetic
KD025
Kadmon
Anti-IL13 Mab
Anti-IL4/IL-13 Mab
Anti-CTGF Mab
Rh serum amyloid p
Selective autotaxin inhibitor
Anti-Integrin αvβ6 Mab
LTD4 / PDE-3,-4 / 5-LOX antagonist
CTGF ligand inhibitor
ROCK-2 inhibitor
Galectin-3 inhibitor
Anti-integrin αvβ6 antagonist

The concept of PF-ILD



Wells AU et al, *Eur Respir J* 2018; 51: 1800692



Wells AU et al, *Eur Respir J* 2018; 51: 1800692

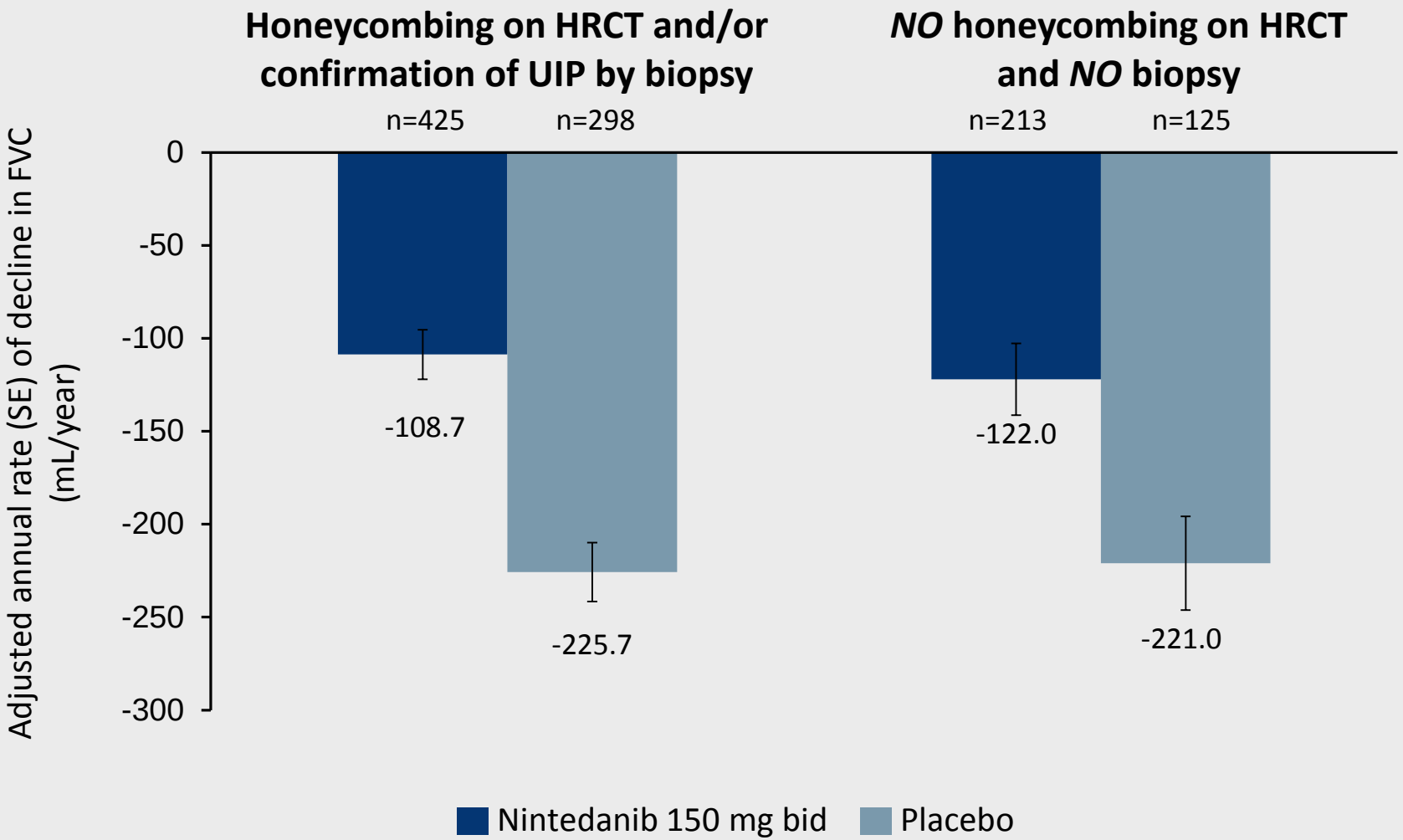
PROGRESSIVE

- Signs of progressive fibrosing lung disease:
  - Symptoms
  - Lung function
  - Imaging (HRCT)

FIBROTIC

- Signs of fibrosing lung disease at HRCT:
  - Honeycombing
  - Traction bronchiectasis
  - Volume loss
  - Reticulation

ANNUAL RATE OF DECLINE IN FVC BY HRCT CRITERIA



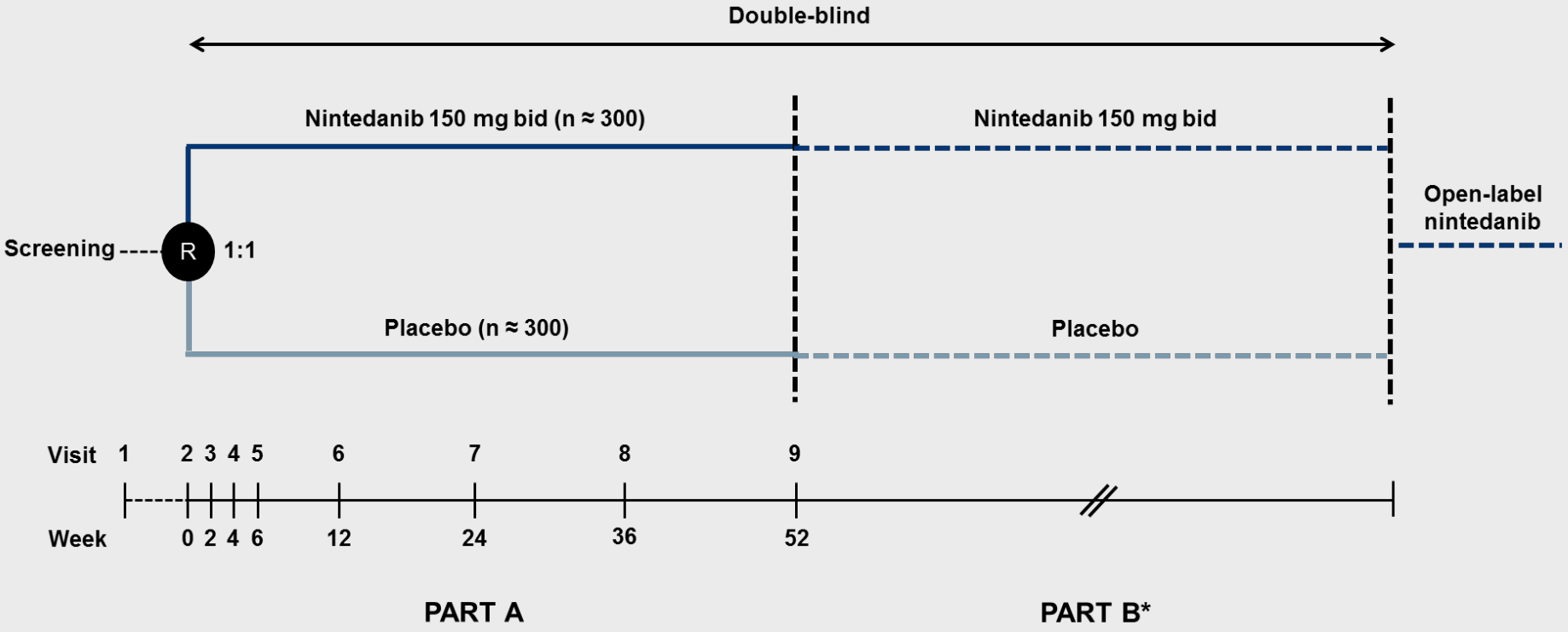
Raghu G et al, *AJRCCM* 2017; 195: 78-85

INBUILD®

(NCT02999178)

Efficacy and safety of nintedanib in patients with progressive fibrosing ILDs

INBUILD TRIAL: DESIGN



Flaherty et al, *BMJ Open Respir Res* 2017;4:e000212

POSSIBLE ILD DIAGNOSES

Trial indication:
List of possible underlying ILD diagnoses (**check only one box**)

- ☐ Idiopathic nonspecific interstitial pneumonia
- ☐ Unclassifiable IIP
- ☐ Other IIP
- ☐ Hypersensitivity pneumonitis
- ☐ Rheumatoid arthritis-associated ILD
- ☐ Mixed connective tissue disease
- ☐ Systemic sclerosis-associated ILD
- ☐ Other CTD-ILD
- ☐ Exposure-related ILD
- ☐ Sarcoidosis
- ☐ Other fibrosing ILD

Flaherty et al, *BMJ Open Respir Res* 2017;4:e000212

INCLUSION CRITERIA: PROGRESSIVE DISEASE

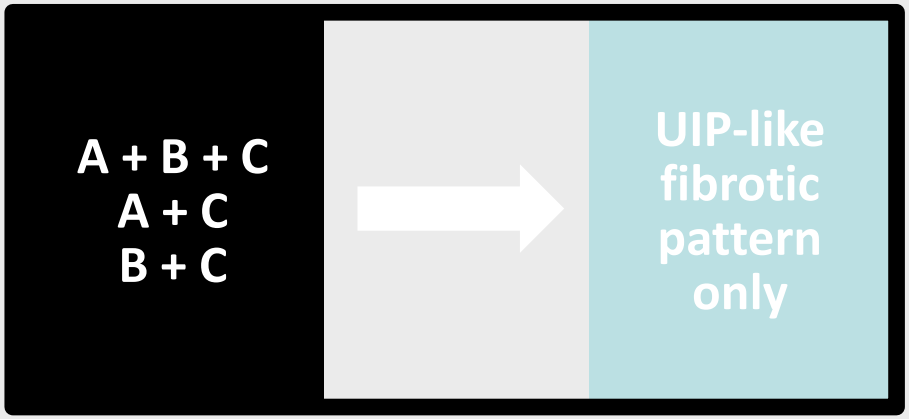
Meeting ≥1 of these criteria for disease progression in the 24 months before screening, despite treatment with (unapproved) medications used in clinical practice to treat ILDs:

- Relative decline in FVC ≥10% predicted
- Relative decline in FVC ≥5–<10% predicted and worsened respiratory symptoms
- Relative decline in FVC ≥5–<10% predicted and increased extent of fibrotic changes on chest imaging
- Worsened respiratory symptoms and increased extent of fibrotic changes on chest imaging

Flaherty et al, *BMJ Open Respir Res* 2017;4:e000212

INBUILD TRIAL: PATTERNS ON HRCT

A. Definite honeycomb lung destruction with basal and peripheral predominance
B. Presence of reticular abnormality and traction bronchiectasis consistent with fibrosis with basal and peripheral predominance
C. Atypical features are absent, specifically nodules and consolidation. Ground glass opacity, if present, is less extensive than reticular opacity pattern



Flaherty et al, *BMJ Open Respir Res* 2017;4:e000212

INBUILD TRIAL: EFFICACY ENDPOINTS

Primary endpoint

- Annual rate of decline in **FVC** (mL/year) assessed over 52 weeks
- There will be two co-primary analysis populations:
  - *all patients*
  - *patients with a UIP-like fibrotic pattern only on HRCT*

  The patient population will be enriched such that 2/3 of patients will have a UIP-like fibrotic pattern on HRCT (at least 400 patients)

Flaherty et al, *BMJ Open Respir Res* 2017;4:e000212

Time for a change: is idiopathic pulmonary fibrosis still idiopathic and only fibrotic?

Paul J Wolters, Timothy S Blackwell, Oliver Eickelberg, James E Loyd, Naftali Kaminski, Gisli Jenkins, Toby M Maher, Maria Molina-Molina, Paul W Noble, Ganesh Raghu, Luca Richeldi, Marvin I Schwarz, Moises Selman, Wim A Wuyts, David A Schwartz

Lancet Resp Med 2018; 6: 154-160

What's in a name? That which we call IPF, by any other name would act the same

Athol U. Wells[1], Kevin K. Brown[2], Kevin R. Flaherty[3], Martin Kolb[4] and Victor J. Thannickal[5], on behalf of the IPF Consensus Working Group[6]

Take-Home Message

- New diagnostic criteria for IPF are emerging (ATS/ERS/JRS/ALAT guidelines soon to be released).
- Therapy of IPF is a fast moving field.
- The concept of PF-ILD is being created and possibly validated in clinical trials.

List of References

1. *Lancet Resp Med,* 2018; 6: e7
2. *Lancet* 2017; 389: 1941-1952
3. *JAMA* doi:10.1001/jama.2018.6129
   (Published online May 20, 2018)
4. *AJRCCM* 2017; 195: 78-85
5. *N Engl J Med* 2018; 378: 1811-23