A New Path to Medicine

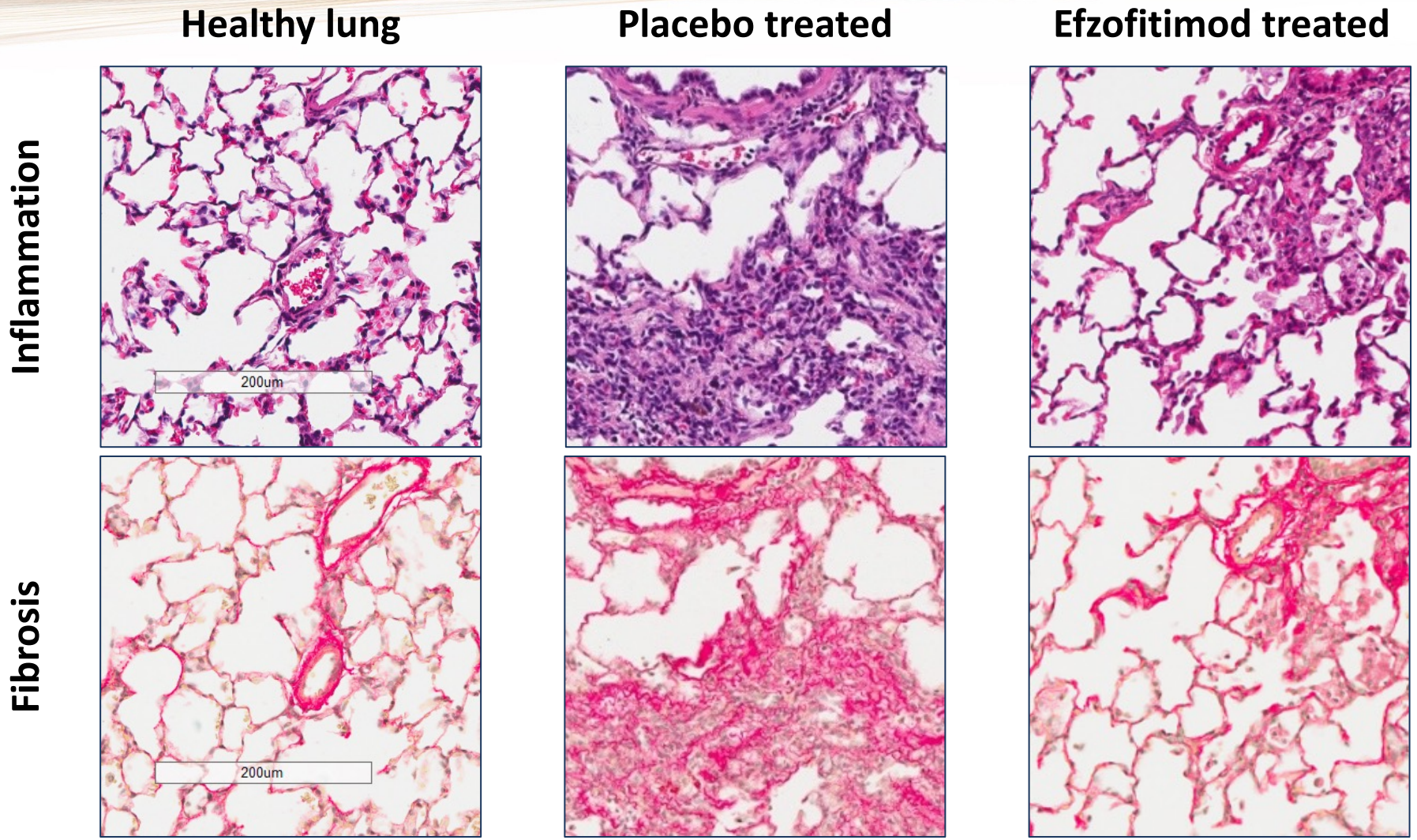
Company Reception

May 16, 2022

Forward Looking Statements

The following slides and any accompanying oral presentation contain forward-looking statements within the meaning of the Private Securities Litigation Reform Act of 1995 and other federal securities laws. The use of words such as “may,” “might,” “will,” “should,” “expect,” “plan,” “anticipate,” “believe,” “estimate,” “project,” “intend,” “future,” “potential,” “opportunity,” or “continue,” and other similar expressions are intended to identify forward-looking statements. For example, all statements regarding: the potential therapeutic benefits of proteins derived from tRNA synthetase genes and our product candidates, including efzofitimod and ATYR2810, and development programs, including our NRP2 antibody program and our tRNA synthetase program; the ability to successfully advance our product candidates and undertake certain development activities (such as the initiation of clinical trials, clinical trial enrollment, the conduct of clinical trials and announcement of clinical results) and accomplish certain development goals, and the timing of such events; the potential market opportunity for our product candidates; our ability to receive regulatory approvals for, and commercialize, our product candidates; our ability to identify and discover additional product candidates; potential activities and payments under collaboration agreements; and the ability of our intellectual property portfolio to provide protection are forward-looking statements. All forward-looking statements are based on estimates and assumptions by our management that, although we believe to be reasonable, are inherently uncertain. All forward-looking statements are subject to risks and uncertainties that may cause actual results to differ materially from those that we expected. These risks, uncertainties and other factors are more fully described in our filings with the U.S. Securities and Exchange Commission, including our Annual Report on Form 10-K, our Quarterly Reports on Form 10-Q, and in our other filings. The forward-looking statements in this presentation speak only as of the date of this presentation and neither we nor any other person assume responsibility for the accuracy and completeness of any forward-looking statement. We undertake no obligation to publicly update or revise any forward-looking statement, whether as a result of new information, future events or otherwise, except as required by law.

Efzofitimod: A Novel Mechanism to Treat Lung Inflammation and Fibrosis

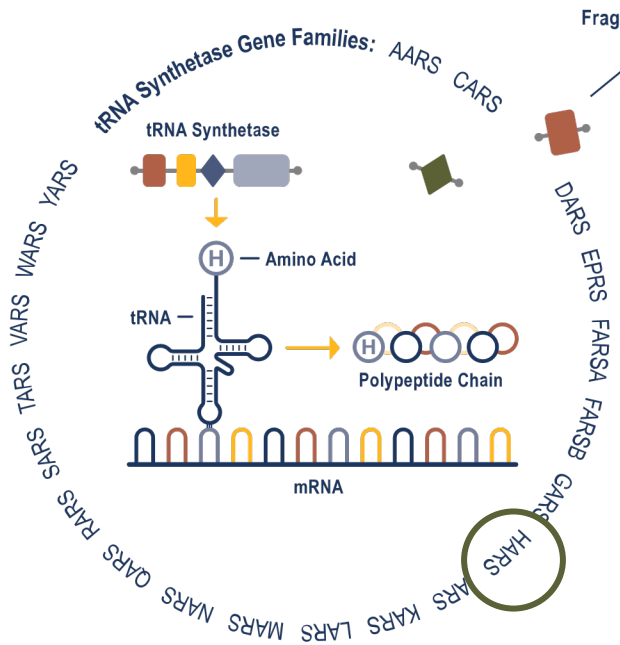


Representative histology showing immune cell infiltrate and collagen content in a rat model of bleomycin induced lung fibrosis presented at the ATS annual meeting 2018.

Foundational Science: Novel Functions of Extracellular tRNA Synthetases

INTRACELLULAR

Catalyze Protein Synthesis

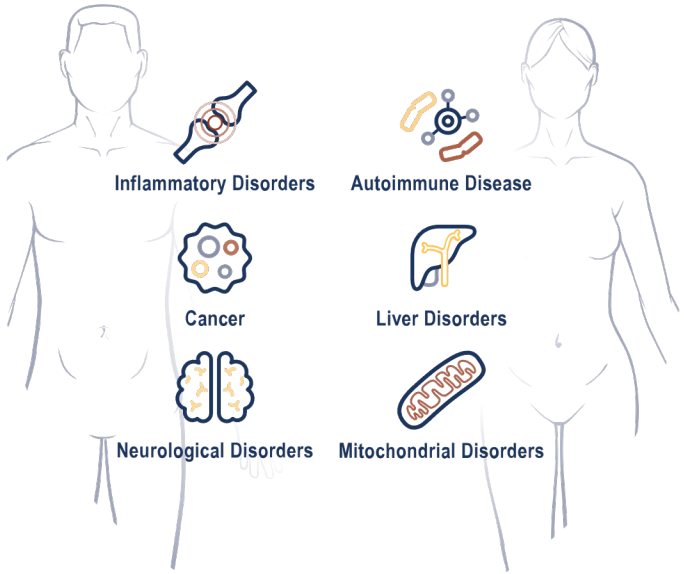


EXTRACELLULAR

Secreted in Circulation and Tissue to Regulate Diverse Pathways

PHYSIOLOGICAL

Pathway Disruption Associated with Disease



tRNA synthetases are secreted extracellularly and occur in novel forms which lose their canonical function

Extracellular tRNA synthetase disruption (genetic/ autoimmune) is associated with disease in humans

Validated synthetase platform is an engine for new protein therapeutics (e.g. efzofitimod) and new target ID (e.g. NRP2)

aTyr

Efzofitimod (ATYR1923, KRP-R120)

A Novel Immunomodulator for Fibrotic Lung Disease

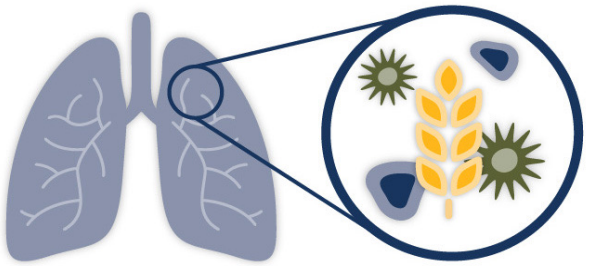
Efzofitimod: First-in-Class Therapy for Fibrotic Lung Disease

MOA	• Targets innate and adaptive immune cells during active inflammation to restore immune balance via selective modulation of NRP2
Pre-Clinical Evidence	• Anti-inflammatory and anti-fibrotic effects demonstrated in translational models of multiple ILDs • Reduces inflammatory cytokines and pro-fibrotic chemokines *in vitro* and *in vivo* • No toxicity observed in GLP toxicology rodents and non-human primates out to 6 months
Clinical Experience	• Safe and well-tolerated in clinical trials to date with exposure to 24 weeks • No immunogenicity observed • Controls inflammation by downregulating cytokines implicated in sarcoidosis and other ILD • Dose-dependent disease control demonstrated in pulmonary sarcoidosis patients

MOA = mechanism of action

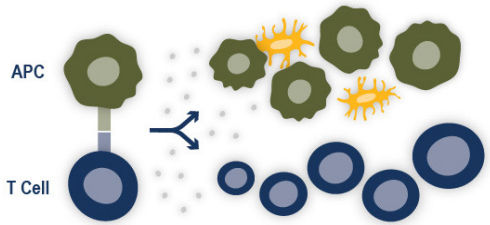
Efzofitimod Therapeutic Goal: Restore Immune Balance and Prevent Fibrosis

Disease Trigger



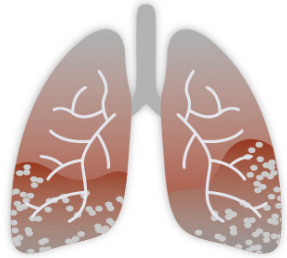
Organic; inorganic; infectious; autoimmune

Aberrant Immune Responses



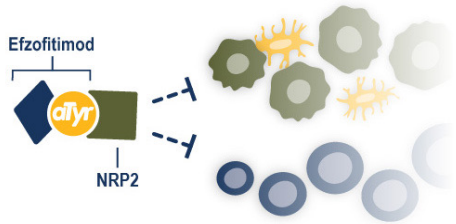
T-cell activation; Pro-inflammatory cytokine/chemokines triggering fibrotic pathways; NRP2 upregulation on immune cells

Lung Inflammation & Fibrosis



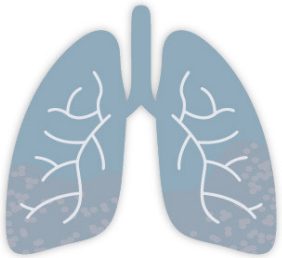
Persistent, unresolved inflammation in the lung can lead to fibrosis; patients experience chronic cough, dyspnea, mortality

Efzofitimod Dampens Immune Responses



Efzofitimod binds to NRP2 and downregulates cytokine and chemokine production and T-cell activation

Stabilized Lung



Reduced inflammation and fibrotic deposition; symptom relief, stabilized lung function*

*aTyr hypothesis

First Efzofitimod Indication: Pulmonary Sarcoidosis

- Multisystem inflammatory disorder of unknown etiology, characterized by the formation of granulomas (clumps of immune cells)
- Lung affected in ~90% of all patients
- ~5x increased mortality in advanced disease
- SOC: corticosteroids; cytotoxic immunosuppressants; anti-TNFs – limited development pipeline
- High unmet need for treatments with improved safety and clinically established efficacy

Large orphan population

50-75% require treatment

Persistent or progressive disease in **30-50%**

10-30% develop fibrosis with **5-10%** mortality

SOC – standard of care; Culver e2019; Baughman 2016; Baughman 2006, Mana 1994; Viskum 1993; Nagai 1999; Judson 1993; Neville 1983; Israel 1986; Wijsenbeek 2019

Efzofitimod (ATYR1923, KRP-R120)

Results from Phase 1b/2a Study in Pulmonary Sarcoidosis

Peter H. S. Sporn, M.D.
Professor of Medicine (Pulmonary and Critical Care); Cell and Developmental Biology; and Medical Education
Northwestern University Feinberg School of Medicine

Phase 1b/2a Trial Design

Design	• Randomized (2:1), double-blind, placebo-controlled, multiple ascending dose • 24 week study: 6 monthly IV doses of efzofitimod tested at 1, 3, and 5 mg/kg • Forced steroid taper to 5 mg by week 8; amendment to allow taper to 0 mg at week 16 in responders
Population	• 37 histologically confirmed pulmonary sarcoidosis patients • ≥10 mg stable oral corticosteroid treatment • Symptomatic/active disease at baseline
Primary Endpoint	• Safety and tolerability of multiple ascending IV efzofitimod doses
Secondary Endpoints	• Steroid-sparing effect • Immunogenicity • Pharmacokinetics (PK) • Exploratory: Lung function (FVC and other PFTs); sarcoidosis symptom scores and quality of life scales; inflammatory and sarcoidosis biomarkers; FDG-PET/CT imaging

FVC = Forced Vital Capacity; PFTs = Pulmonary Function Tests

Phase 1b/2a Inclusion / Exclusion Criteria

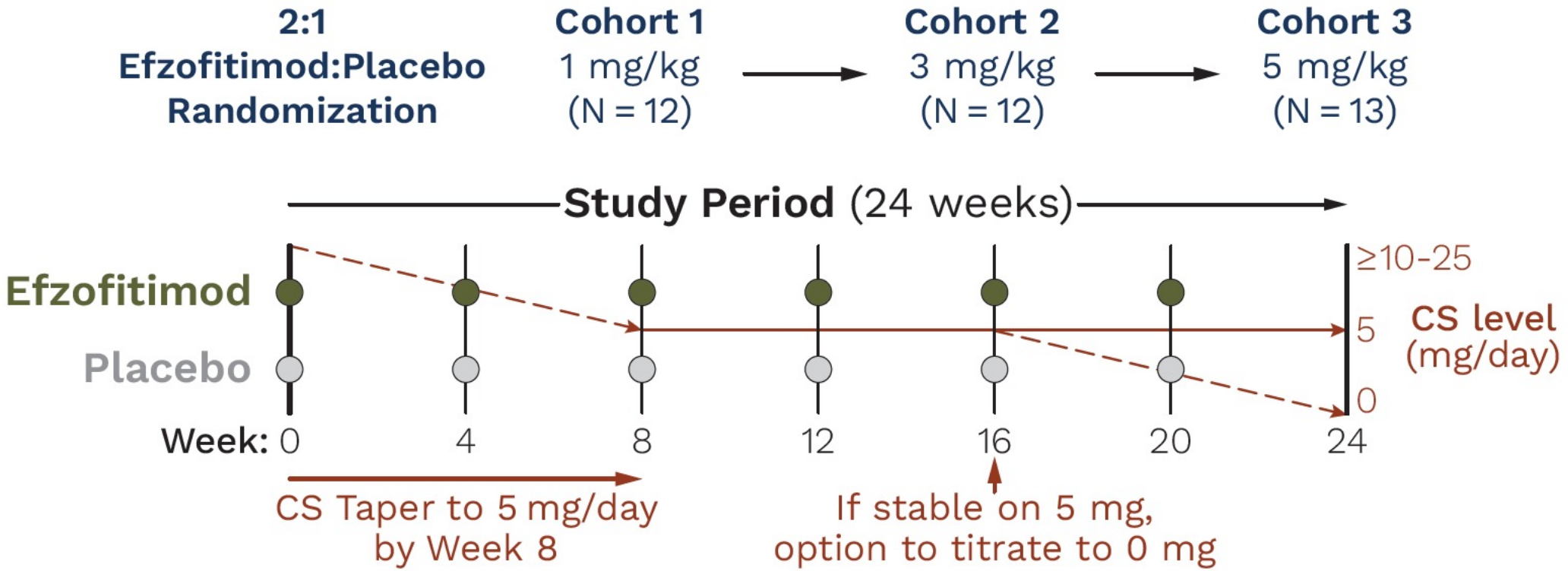
Inclusion

- Histologically proven diagnosis of pulmonary sarcoidosis
- Stable treatment with 10 -25 mg/day oral corticosteroid
  - oral immunomodulator allowed
- Symptomatic/active disease at baseline
  - FVC ≥ 50% percent predicted
  - MRC Dyspnea Scale score (>= 1)

Exclusion

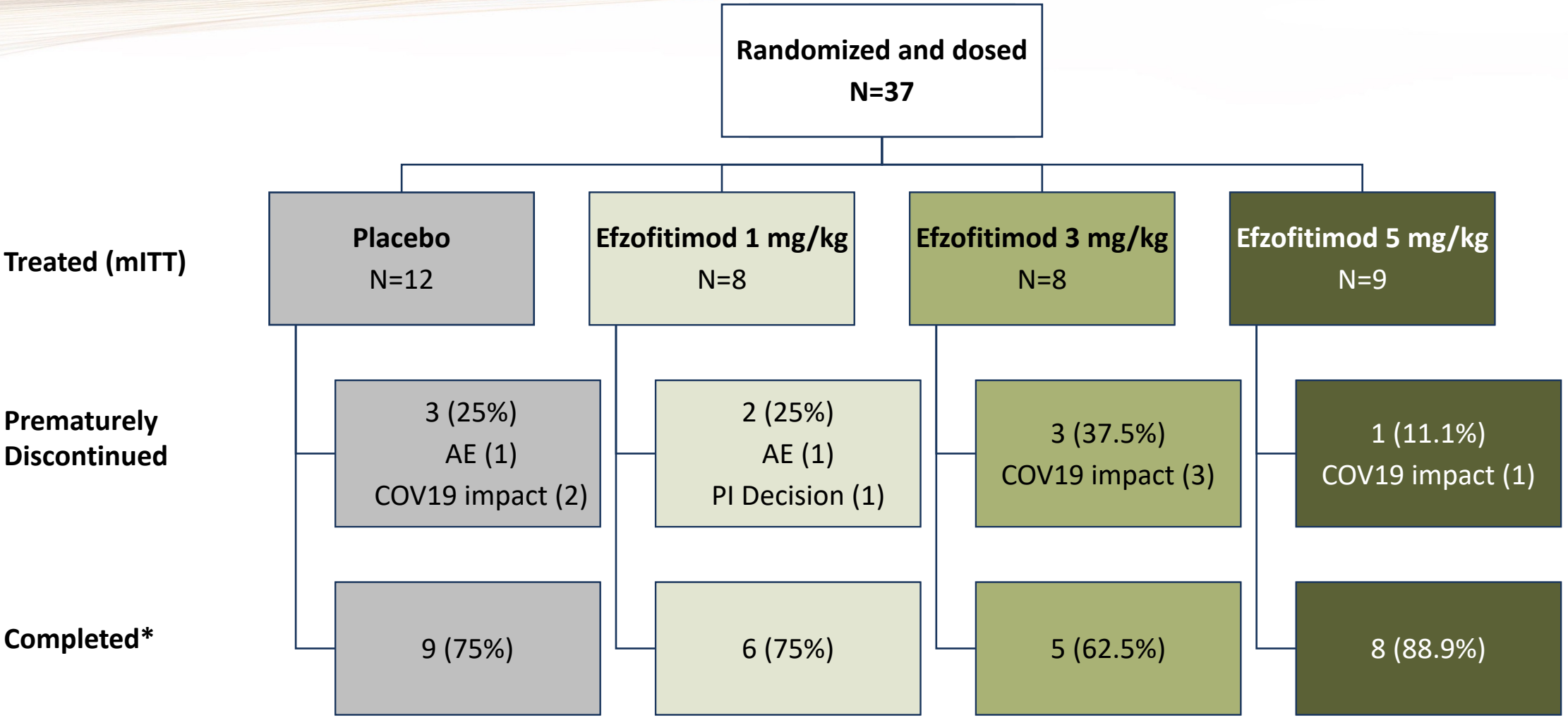
- Disease consistent with Lofgren's syndrome
- Treatment with biological immunomodulator such as tumor necrosis factor-alpha inhibitors
- Clinically significant cardiac, neurological, gastrointestinal, and/or renal sarcoidosis
- PH requiring vasodilator treatment

Phase 1b/2a Study Schema



(1) subtherapeutic dose

Phase 1b/2a Disposition



mITT = randomized and dosed; *Defined as receiving at least 5 doses and having an end of study assessment

Baseline Demographics and Disease Characteristics Generally Balanced

Demographics	Placebo N=12	Efzofitimod 1 mg/kg N=8	Efzofitimod 3 mg/kg N=8	Efzofitimod 5 mg/kg N=9
Age (Years); mean (SD), >=65	52.5 (10.2), 0	54.5 (11.3), 1	51.8 (11.4), 2	50.8 (9.2), 0
Sex (Male); n (%)	5 (42)	4 (50)	4 (50)	4 (44)
Race; White / African American	9 / 3	5 / 3	6 / 2	3 / 6
Disease characteristics Mean (SD)				
FVC (% predicted)	77.3 (11.5)	68.3 (9.7)	83.8 (7.3)	83.8 (16.6)
Duration of disease (Years)	4.2 (3.3)	7.4 (6.1)	5.9 (5.1)	7.7 (9.9)
Baseline Dyspnea Index Score	4.8 (2.0)	4.3 (1.8)	7.6 (3.0)	6.3 (2.5)
Background Therapy				
Steroid dose (mg/day), mean	13.3	11.3	14.4	13.9
Immunomodulator; n (%)	6 (50)	3 (38)	1 (13)	4 (44)

Monthly Dosing of Efzofitimod was Safe and Well-Tolerated

n (%)	Placebo N=12	Efzofitimod 1 mg/kg N=8	Efzofitimod 3 mg/kg N=8	Efzofitimod 5 mg/kg N=9
AEs	10 (83)	8 (100)	7 (88)	8 (89)
Drug-related AEs	4 (33)	3 (38)	1 (13)	3 (33)
Severe AEs (Grade 3 or 4)	4 (33)	2 (25)	0	2 (22)
SAEs	1 (8)	1 (13)	0	0

- No new or unexpected findings with repeat dosing
- No dose-response relationship observed
- Most frequent AEs were consistent with underlying disease: Cough, fatigue, wheezing
- No signal of immunogenicity
- No drug related SAEs
- No deaths

Safety population

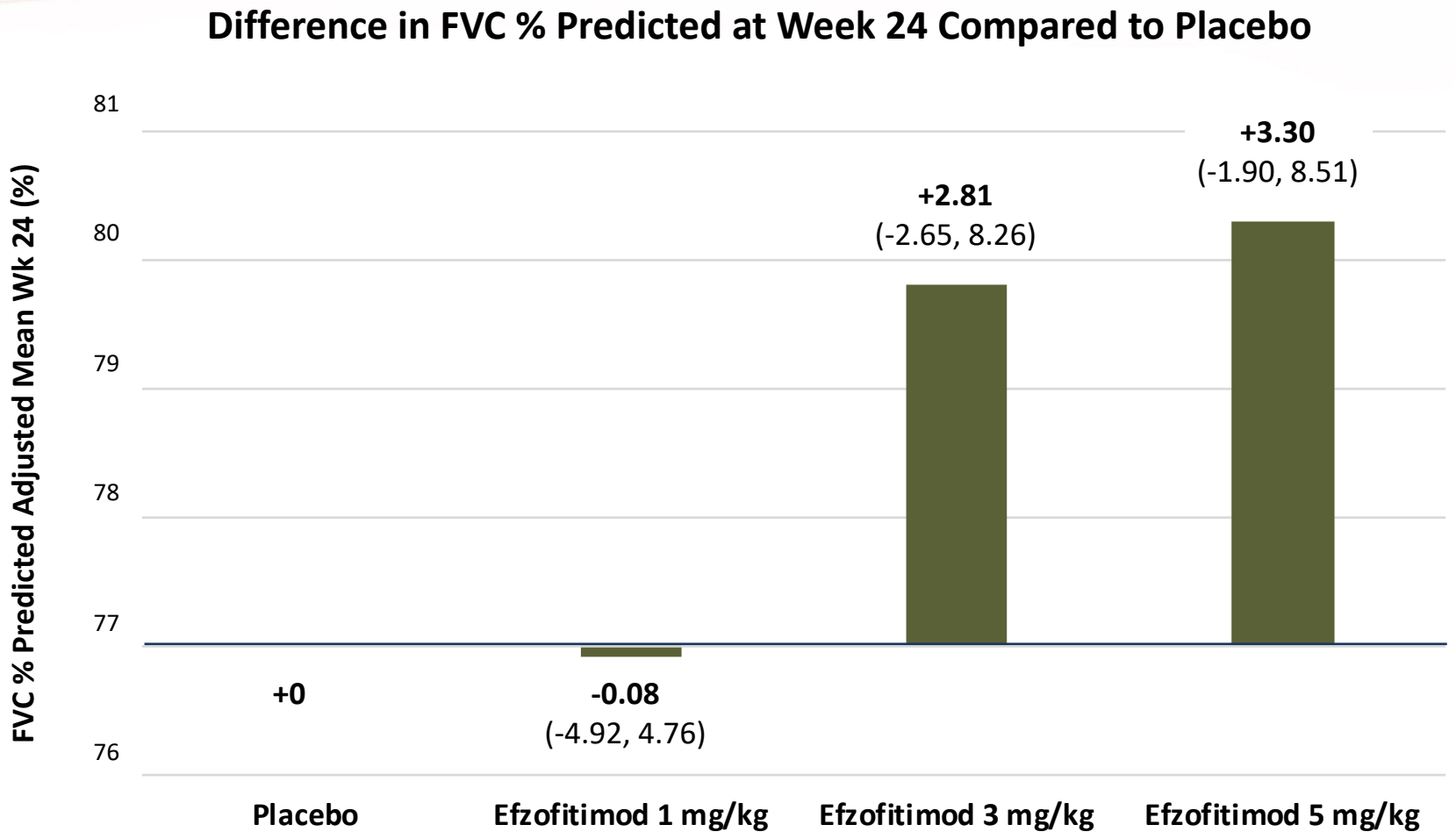
Dose-dependent Reduction in Steroid Utilization Compared with Placebo

Post-taper Period	Placebo N=12	Efzofitimod 1 mg/kg N=8	Efzofitimod 3 mg/kg N=8	Efzofitimod 5 mg/kg N=9
Average daily dose (mg), adjusted mean	7.17	6.83	6.54	5.62
- relative reduction vs placebo (%)	-	-5%	-9%	-22%
Change from baseline (%), mean	-46	-41	-49	-58
- difference in adjusted means (%), mean (95% CI)	-	1.2 (-20, 22)	-2.3 (-23, 19)	-12.3 (-33, 8)
Tapered to 0 mg and maintained taper, n (%)	0	0	0	3 (33)

- 58% overall steroid reduction from baseline and 22% relative reduction compared to placebo in steroid utilization post taper in the 5 mg/kg treatment group
- 33% of patients able to taper off of steroids completely in 5 mg/kg treatment group while controlling disease symptoms

D51-EOS; adjusted mean refers to least squares mean from ANCOVA adjusting for baseline steroid use

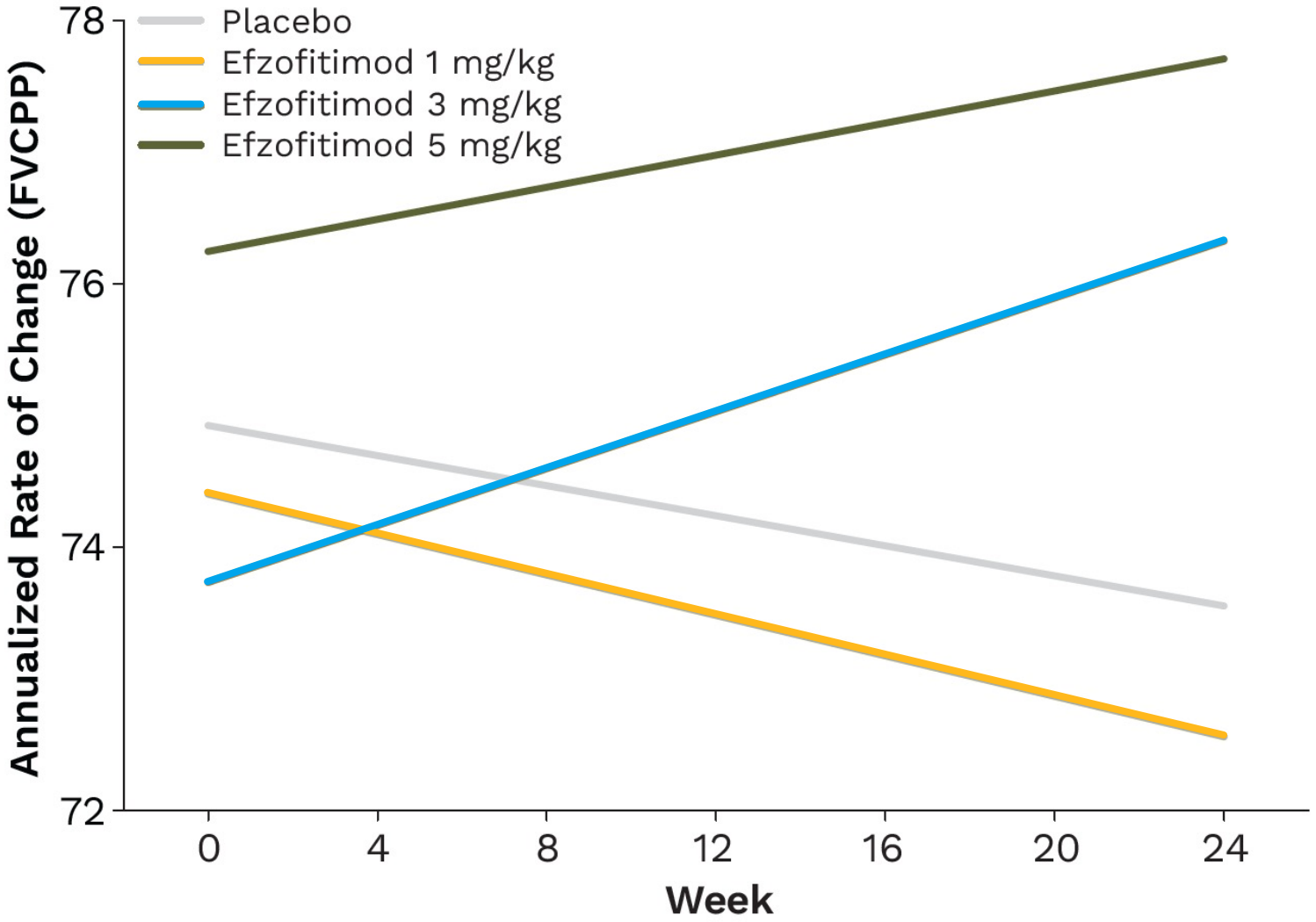
Dose-dependent Improvement in FVC % Predicted Compared to Placebo



FVC % predicted adjusted mean at week 24 in the placebo arm = 76.8%

Differences represent from MMRM adjusting for baseline FVC % Predicted, mean (95% CI)

Higher Efzofitimod Doses Show Trends of Improvement in FVC % Predicted



Dose-dependent Improvements in PFTs Over Time Compared with Placebo

FVC, mean (SD)	Placebo N=12	Efzofitimod 1 mg/kg N=8	Efzofitimod 3 mg/kg N=8	Efzofitimod 5 mg/kg N=9
Absolute CFB at Week 24 (mL)	-40 (230)	-80 (160)	120 (130)	110 (250)
FVC % predicted, mean (SD)				
Absolute CFB at Week 24 (%)	-0.9 (6.1)	-2.3 (3.9)	2.6 (2.5)	2.6 (5.6)
Dlco % predicted, mean (SD)				
Absolute CFB at Week 24 (%)	-6.2 (14.4)	-6.1 (4.0)	-1.4 (5.0)	4.4 (14.6)

PFTs = Pulmonary Function Tests; CFB = Change From Baseline

Dose-dependent Symptom Improvements Compared with Placebo

Differences in Adjusted Means vs Pbo at Week 24	Efzofitimod 1 mg/kg N=8	Efzofitimod 3 mg/kg N=8	Efzofitimod 5 mg/kg N=9
Dyspnea (TDI)	-0.76	3.33	4.49
Cough (LCQ)	-3.49*	2.98*	2.05
Fatigue (FAS)	0.76	-4.78	-7.77*
King's Sarcoidosis Score: Lung	-6.41	11.29	16.17*
King's Sarcoidosis Score: General Health	-5.1	2.13	18.33*

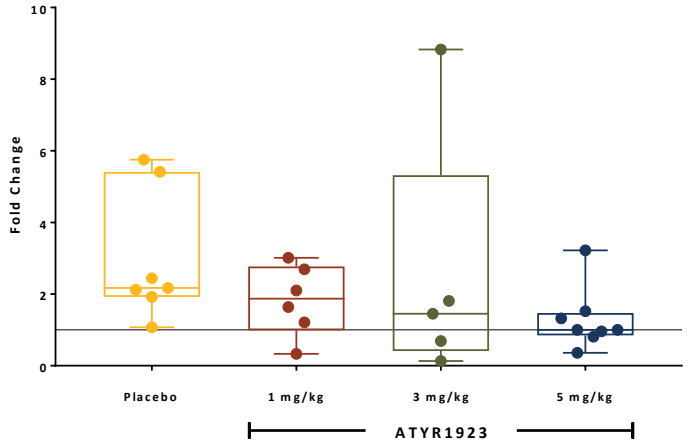
= clinically meaningful improvement based on published MCID

*p<0.05 on difference between adjusted means from MMRM; Pbo = Placebo

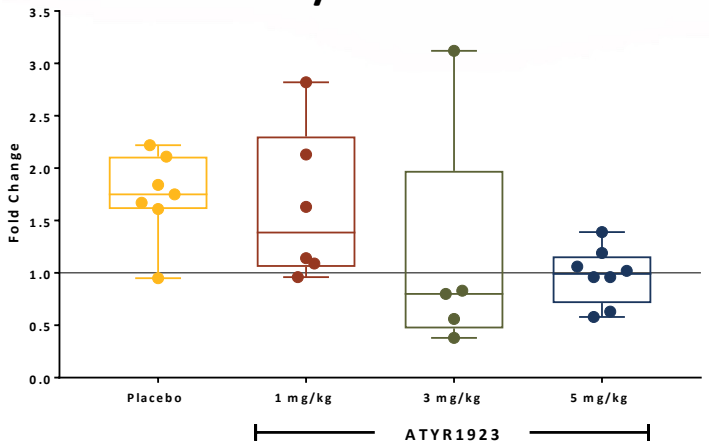
MCIDs: TDI - Witek 2003; LCQ – Raj 2009; FAS - de Kleijin 2011 (Negative score is better for Fatigue); KSQ Lung – Baughman 2021; KSQ Lung – Baughman 2021

Dose-dependent Control of Key Disease and Inflammatory Biomarkers

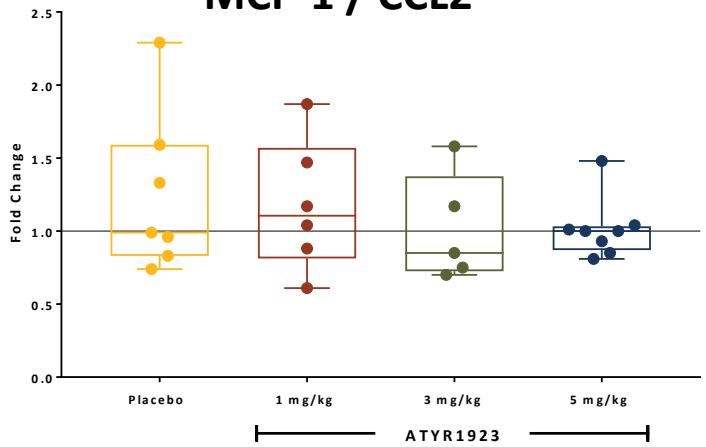
IFNγ



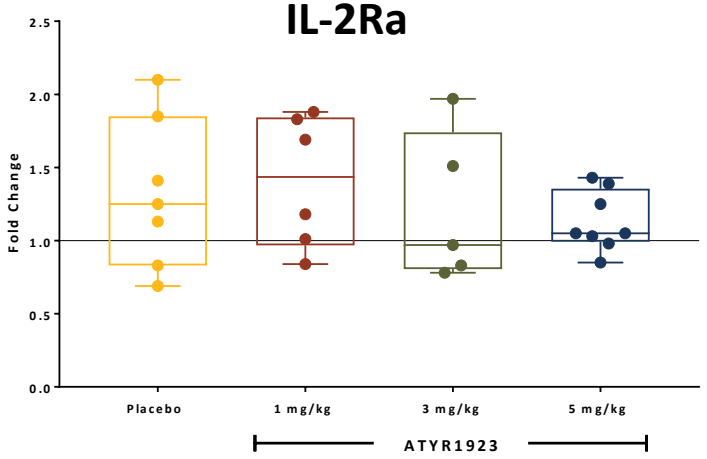
IP-10 / CXCL10



MCP-1 / CCL2



IL-2Ra



Graphs represent fold change at Week 24 compared to Baseline

Phase 1b/2a Data Supports Proof-of-Concept and Clinical Advancement

- Efzofitimod was safe and well-tolerated
- Strong suggestion of efficacy: Dose-response and consistent positive trends across key efficacy endpoints and multiple analysis populations
- Clinically meaningful symptom improvements in dyspnea, cough, fatigue and King's Sarcoidosis scores
- Dose dependent control of inflammatory biomarkers confirms anti-inflammatory effect

Efzofitimod (ATYR1923, KRP-R120)

Phase 3 EFZO-FIT™ Study

Robert P. Baughman, M.D.
Emeritus Professor of Medicine
University of Cincinnati

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EFZO-FIT™ : Phase 3 Study of Efzofitimod in Pulmonary Sarcoidosis

Clinical POC Established

- Phase 1b/2a study was first in sarcoidosis to demonstrate effects on physiologic and quality of life measures concurrent with steroid reduction

FDA Alignment

- Positive FDA End-of-Phase 2 meeting supports advancement of clinical development
- Aligned on prioritization of endpoints that are most clinically meaningful to patients and providers

Global Pivotal Trial

- Multiple sites in North America, Europe and Japan
- Expected to initiate in Q3 2022

Trial Design

Objectives	• Primary: Assess the efficacy of efzofitimod in patients with pulmonary sarcoidosis • Secondary: Assess the safety and tolerability of efzofitimod in patients with pulmonary sarcoidosis • Exploratory: Explore the utility of serum biomarkers in evaluation of interventions to treat pulmonary sarcoidosis
Design	• Phase 3, randomized, double-blind, placebo-controlled, multicenter study
Randomization	• Target enrollment of 264 patients • 1:1:1 efzofitimod 3 mg/kg, efzofitimod 5 mg/kg, or placebo, with 88 patients assigned to each arm • Randomization stratum: ○ Presence or absence of concomitant immunosuppressant therapy, and ○ OCS dose at baseline (< 10 mg/day versus ≥ 10 mg/day [prednisone or equivalent])
Duration	• Screening: up to 4 weeks • Treatment Period: 48 weeks (12 monthly doses through Week 44); Primary Assessment at Week 48 • Final Visit: Week 52 (8 weeks post-dose follow-up)

Still under discussion with Kyorin clinical team

Major Efficacy Endpoints

Primary Endpoint

- Change from baseline in mean daily OCS dose post-taper

Secondary Endpoints

- Annual rate of change in absolute value of FVC
- Percent change from baseline in mean daily OCS dose post-taper at Week 48
- Change from baseline in KSQ-Lung score at Week 48

Key Inclusion / Exclusion Criteria

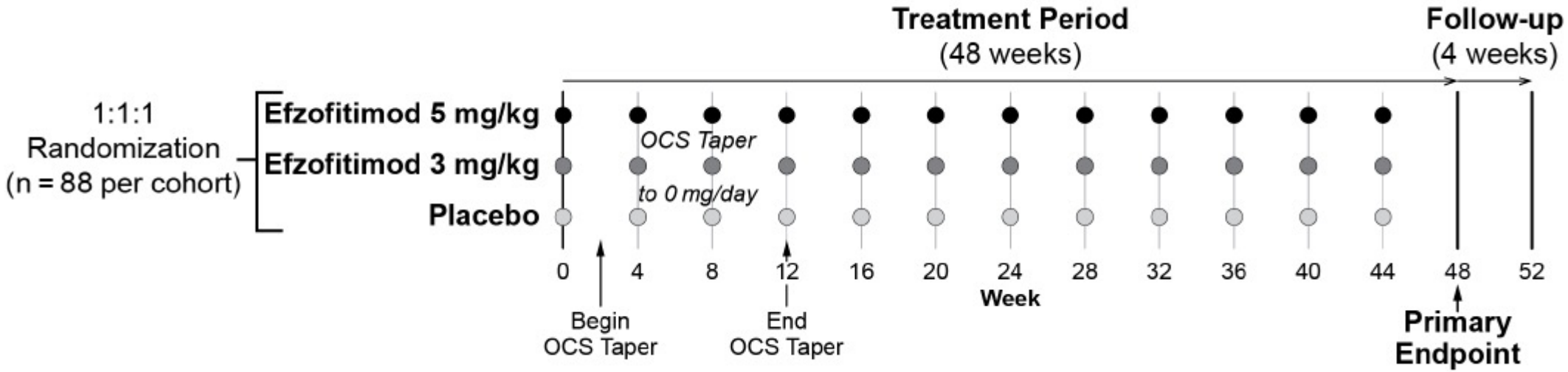
Inclusion

- Adults ages 18-75, inclusive
- Diagnosis of pulmonary sarcoidosis for ≥6 months
- Requiring stable treatment with ≥7.5 but ≤25 mg/day oral corticosteroids
- Medical Research Council (MRC) Dyspnea Scale ≥ 1
- KSQ Lung Score ≤ 70

Exclusion

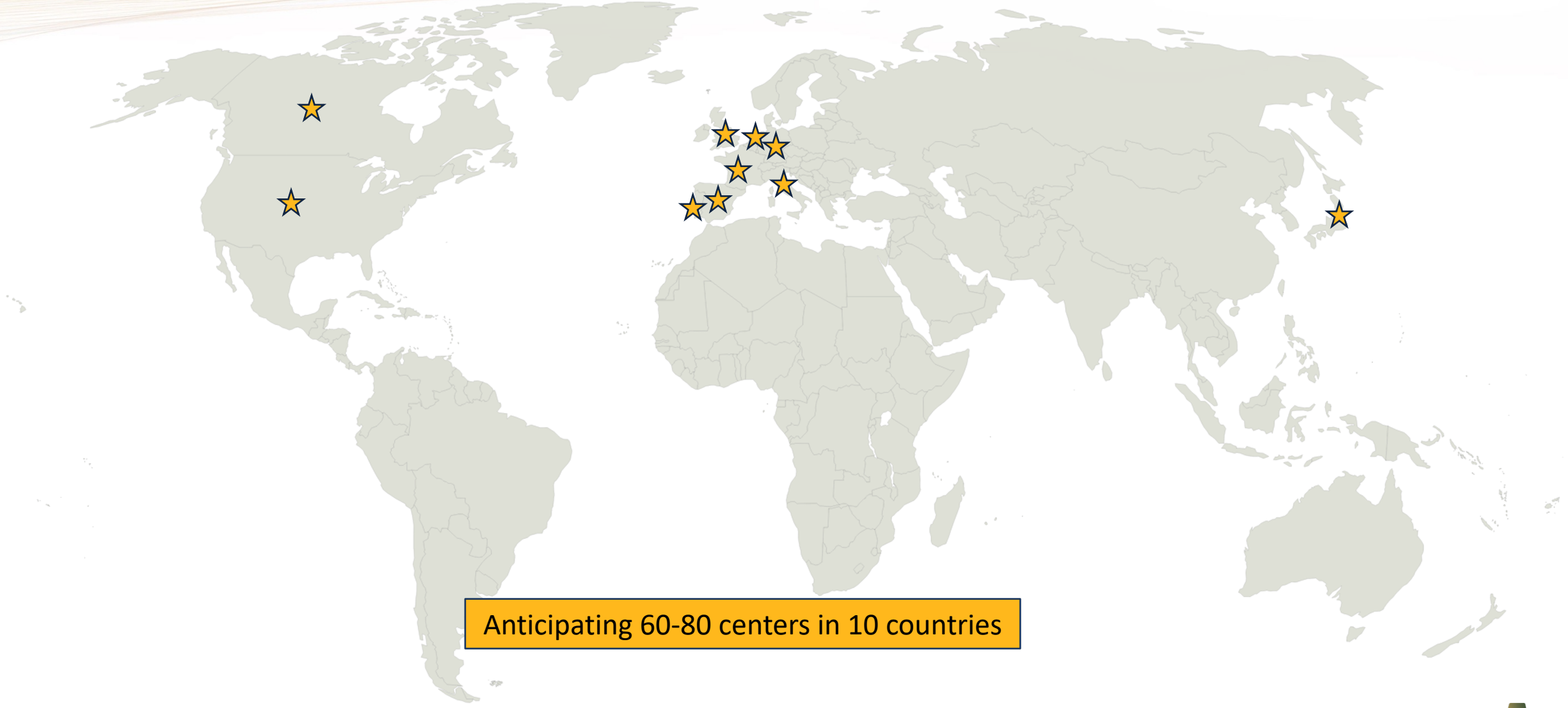
- Extent of fibrosis > 20%
- Forced Vital Capacity < 50%
- Treatment > 1 oral immunomodulator
- Clinically significant pulmonary hypertension
- Patients with cardiac/neuro/renal sarcoidosis
- Treatment with biological immunomodulators

Phase 3 Study Schema



OCS = oral corticosteroids

Multi-center Trial with Sites in North America, Europe and Japan



Anticipating 60-80 centers in 10 countries

Thank You