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July 2018

Forward Looking Statements

This presentation (together with any other statements or information that we may furnish to you) contains forward-looking statements within the meaning of the Private Securities Litigation Reform Act of 1995 with respect to Kiniksa Pharmaceuticals, Ltd. (and its consolidated subsidiary, together, unless context otherwise requires, “Kiniksa” or “we”). All statements contained in this presentation that do not relate to matters of historical fact should be considered forward-looking statements, including without limitation statements regarding potential product applications, potential acquisitions or collaborations, product development activities, clinical studies, regulatory submissions and approvals, and similar operating matters.

Many factors may cause actual results to differ from these forward-looking statements, including the important factors discussed under the caption “Risk Factors” in our final prospectus filed with the Securities and Exchange Commission (“SEC”) on May 24, 2018 relating to our Registration Statement on Form S-1 and our other reports filed with the SEC. These forward-looking statements reflect various assumptions of Kiniksa's management that may or may not prove to be correct. No forward-looking statement is a guarantee of future results or events, and one should avoid placing undue reliance on such statements. Except as otherwise indicated, this presentation speaks as of the date of this presentation. Kiniksa undertakes no obligation to update any forward-looking statements, whether as a result of new information, future events or otherwise.

This presentation also contains estimates, projections, and other information concerning Kiniksa's industry, Kiniksa's business and the markets for certain of Kiniksa's product candidates, including data regarding the estimated size of those markets, and the incidence and prevalence of certain medical conditions. Information that is based on estimates, forecasts, projections, market research, or similar methodologies is inherently subject to uncertainties and actual events or circumstances may differ materially from events and circumstances reflected in this information. Unless otherwise expressly stated, Kiniksa obtained this industry, business, market and other data from reports, research surveys, clinical trials, studies and similar data prepared by market research firms and other third parties, from industry, medical and general publications, and from government data and similar sources.

Use of Proforma Measure - In addition, this presentation and oral statements made in connection with this presentation reference post IPO proforma cash and cash equivalents as of March 31, 2018, which is not prepared in accordance with generally accepted accounting principles (“GAAP”). Management believes that this proforma measure provides useful information to our investors regarding our cash and cash equivalents had we closed our initial public offering and our IPO underwriters’ purchase of 1,006,425 shares of our Class A common shares under their option to purchase additional Class A common shares within the first quarter of 2018. However, the use of this proforma measure is limited as it includes certain items not included in the most directly comparable GAAP measure and should be viewed as supplemental to, and not in substitution for, cash and cash equivalents prepared in accordance with GAAP.



KINIKSA

*A company with a sequential pipeline,
FIRMLY rooted in strong biologic
rationale or validated mechanisms,
potential for multiple indications, and
designed to deliver near-, mid- and
long-term value*

The Kiniksa senior team has extensive experience in drug development & commercialization of valuable therapies

Sanj K. Patel

Chairman & Chief Executive Officer

Steve Mahoney

President & Chief Operating Officer

John Paolini

Chief Medical Officer

Brendan Classon

Head of Research

Christine Maurer

SVP Program Management

Carsten Boess

Corporate Affairs

Chris Heberlig

Chief Financial Officer

Eben Tessari

Chief Business Officer

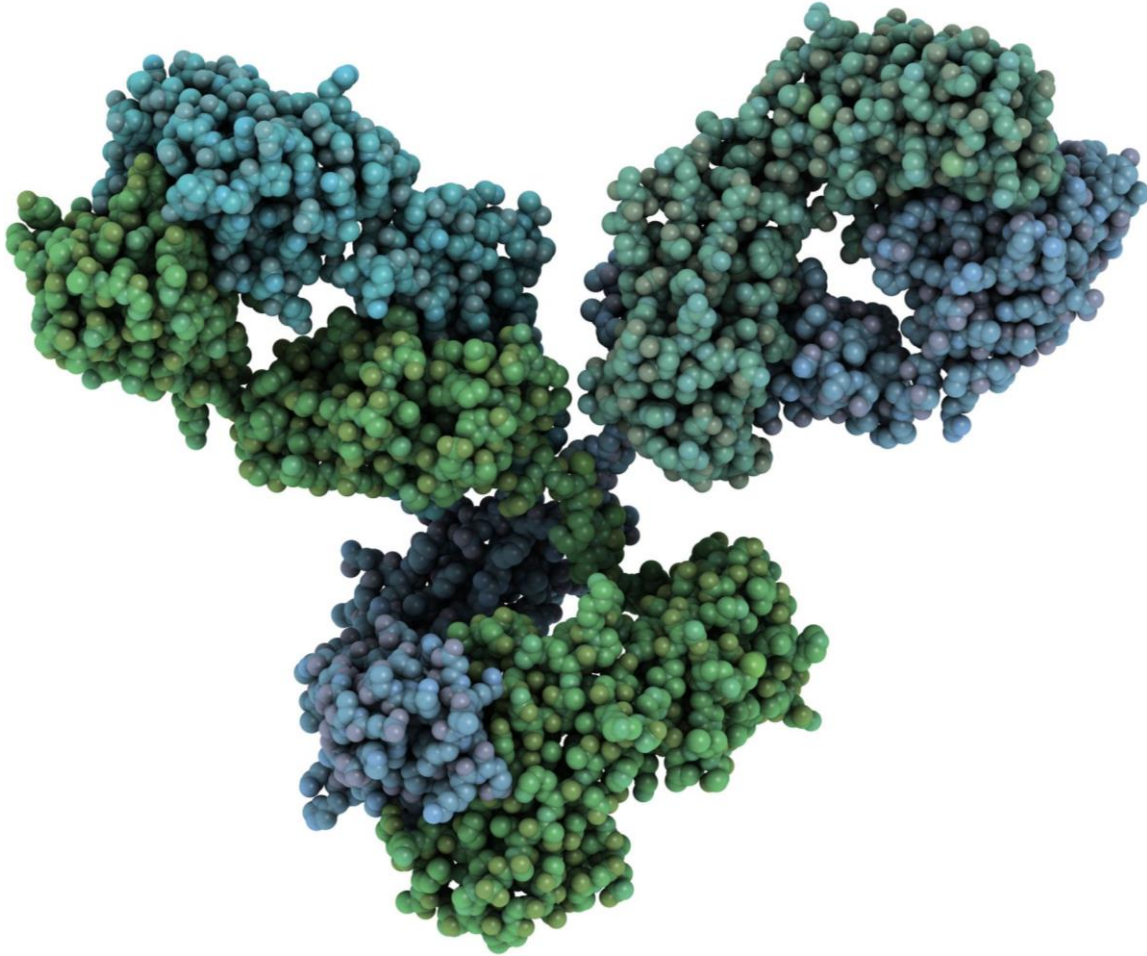


SANOFI



Our Strategy

To build a fully-integrated global biopharmaceutical company by discovering, acquiring, developing and commercializing life-changing therapies for debilitating diseases



Focus on the Patients

Rapid Product Development

Expand Existing Portfolio Across
Multiple Indications

Build Core Capability in Autoimmune and
Autoinflammatory Diseases

Identify New Products

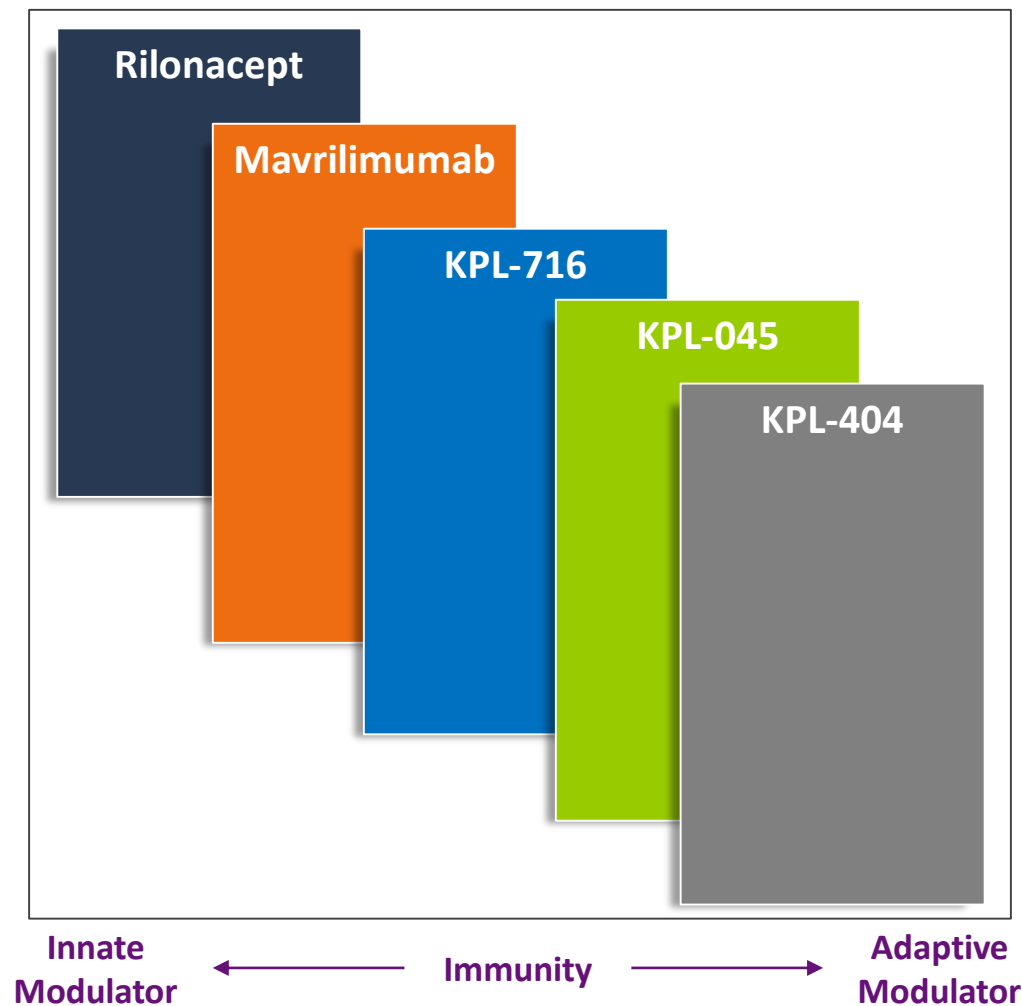
Strategic approach to building a portfolio

- Deep literature and KOL reviews to map diseases with a specialty call point on two main axes:
 - Clear Biology
 - Unmet need
 - Differentiation
- **Strict criteria / filter for:**
 - Market size / prevalence
 - Level of competition
- **Highly selective filter → 5 assets acquired out of >1k reviewed**

Autoinflammatory

Indications

Autoimmune



Identified pockets of unmet need ripe for innovation across a range of autoinflammatory & autoimmune diseases

Kiniksa Pipeline

Program & Target	Originator	Lead Indication	Phase				Status and Anticipated Next Milestone	Rights
			Preclin	1	2	3		
Rilonacept¹ IL-1α & IL-1β	Regeneron	Recurrent Pericarditis					- Additional interim data from ongoing open label Phase 2 pilot study expected in 2H 2018 - Plan to commence a pivotal Phase 3 trial in 2H 2018 pending an End of Phase 2 meeting with the FDA	Worldwide (excluding MENA)
Mavrilimumab GM-CSFRα	MedImmune	Giant Cell Arteritis					Plan to commence a Phase 2 proof-of-concept trial in 2H 2018	Worldwide
KPL-716 OSMRβ	Biogen	Prurigo Nodularis / Atopic Derm					- Data from single dose Phase 1a/1b trial expected in 2H 2018 - Plan to commence a repeat single dose Phase 1b trial in AtD patients in 2H 2018	Worldwide
KPL-045² CD30L	NovoNordisk	Autoimmune					IND filing planned for 2019	Worldwide
KPL-404² CD40	Primatepe	Autoimmune					IND filing planned for 2019	Exclusive Option for Worldwide

1) Rilonacept (ARCALYST®) is approved and marketed for cryopyrin-associated periodic syndrome, in the United States by Regeneron. We will assume the rights to this indication upon receiving approval for rilonacept in the recurrent pericarditis indication; 2) We are planning IND-enabling studies for both KPL-045 and KPL-404 in T-cell-dependent, B-cell-mediated diseases, such as pemphigus/pemphigoid, myasthenia gravis, or graft versus host disease.

Rilonacept

Approved for CAPs in US as ARCALYST



Phase 2

Mavri

Phase 2-ready

KPL-716

Phase 1a/1b

KPL-045

Pre-IND

KPL-404

Pre-IND

An opportunity in an inflammatory cardiovascular disease with established proof-of-concept and no currently-approved therapies

Lead Indication

- Recurrent Pericarditis

Patient Population¹

- ~90k patients in the US experience an acute incident of pericarditis per year
- ~3k refractory patients and ~9k who are not well-managed on existing therapies in the US
- Incremental opportunity as a steroid-sparing agent and to treat Post Pericardiotomy and Dressler Syndromes

Mechanism of Action²

- IL-1 α and IL-1 β cytokine trap
- Inhibition of IL-1 has been shown to be instrumental in the resolution of recurrent pericarditis

Competition³

- No currently-approved therapies
- Differentiated from both existing marketed IL-1 agents

Clinical Development

- Additional interim data from ongoing open label Phase 2 pilot study expected in 2H 2018
- Plan to commence a pivotal Phase 3 trial in 2H 2018 pending an End of Phase 2 meeting with the FDA

1) UptoDate, Trinity Partners, Mayo Clin Proc. 2010 ;85 (6): 572-593; New Diagnostic Criteria for Acute Pericarditis: A Cardiac MRI Perspective, 2015 American College of Cardiology; 2) Brucato et al. JAMA. 2016, 316 (18): 1906-1912; Arcalyst Prescribing Information; 3) Drugs@FDA: Arcalyst Prescribing Information, Ilaris Prescribing Information, Kineret Prescribing Information; Kaiser et al. Rheumatol Int (2012) 32:295–299; Theodoropoulou et al. Pediatric Rheumatology 2015, 13(Suppl 1):P155; Fleischmann et al, 2017 ACR/ARHP Abstract 1196; Kosloski et al, J of Clin Pharm 2016, 56 (12) 1582-1590; Cohen et al. Arthritis Research & Therapy 2011, 13:R125; Cardiel et al. Arthritis Research & Therapy 2010, 12:R192; Hong et al. Lancet Oncol 2014, 15: 656-666

Recurrent pericarditis is a debilitating disease with no currently approved therapies

Pericarditis is chest pain caused by pericardial inflammation

Acute Pericarditis is diagnosed in patients with two of the following:

- (1) Retrosternal, pleuritic chest pain (85-90% of cases), (2) Abnormal ECG (ST elevation and PR depression); (3) Pericardial effusion

Often Idiopathic Etiology:

- Absent a clear sign of infection, it is assumed that most cases are post-viral, but are termed “idiopathic”

Recurrent Pericarditis:

- Diagnosed if there is recurrence after initial episode of acute pericarditis, with a symptom-free interval of > 4-6 weeks →. First recurrence is followed by more recurrences between 20% - 30% of the time

Involvement of IL-1 in Recurrent Idiopathic Pericarditis:

- IL-1 has been implicated by several case reports and the AIRTRIP Study to be critical in idiopathic pericarditis

Recurrent pericarditis causes significant impairment of quality of life

Acute Episodes Have Favorable Prognosis:

- For most patients, acute pericarditis episodes last less than a few weeks and resolve on their own

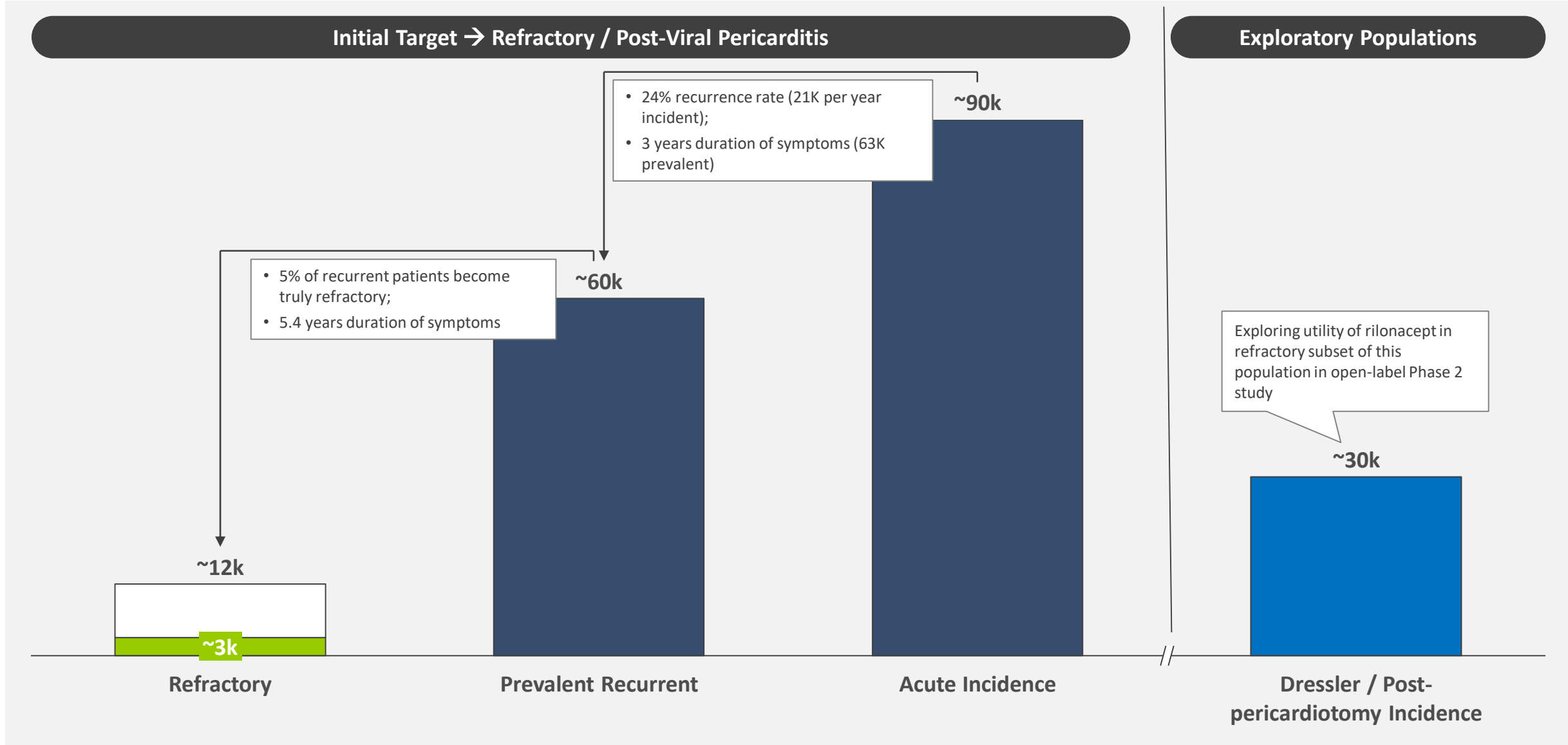
Recurrent Disease Creates Burden on QOL:

- Although pericarditis is rarely life-threatening, patients may have significant impairment on quality of life due to chest pain:
 - Interference with sleep, as chest pain worsens while reclining
 - Lower productivity at work or school
 - Some patients may be on disability or close to it
 - Standard of care treatments have significant AEs

Complications Are Rare But Severe:

- Complications of pericarditis are rare (i.e., effusion, tamponade, constrictive pericarditis) but, when they occur, they can be life threatening and often require surgery

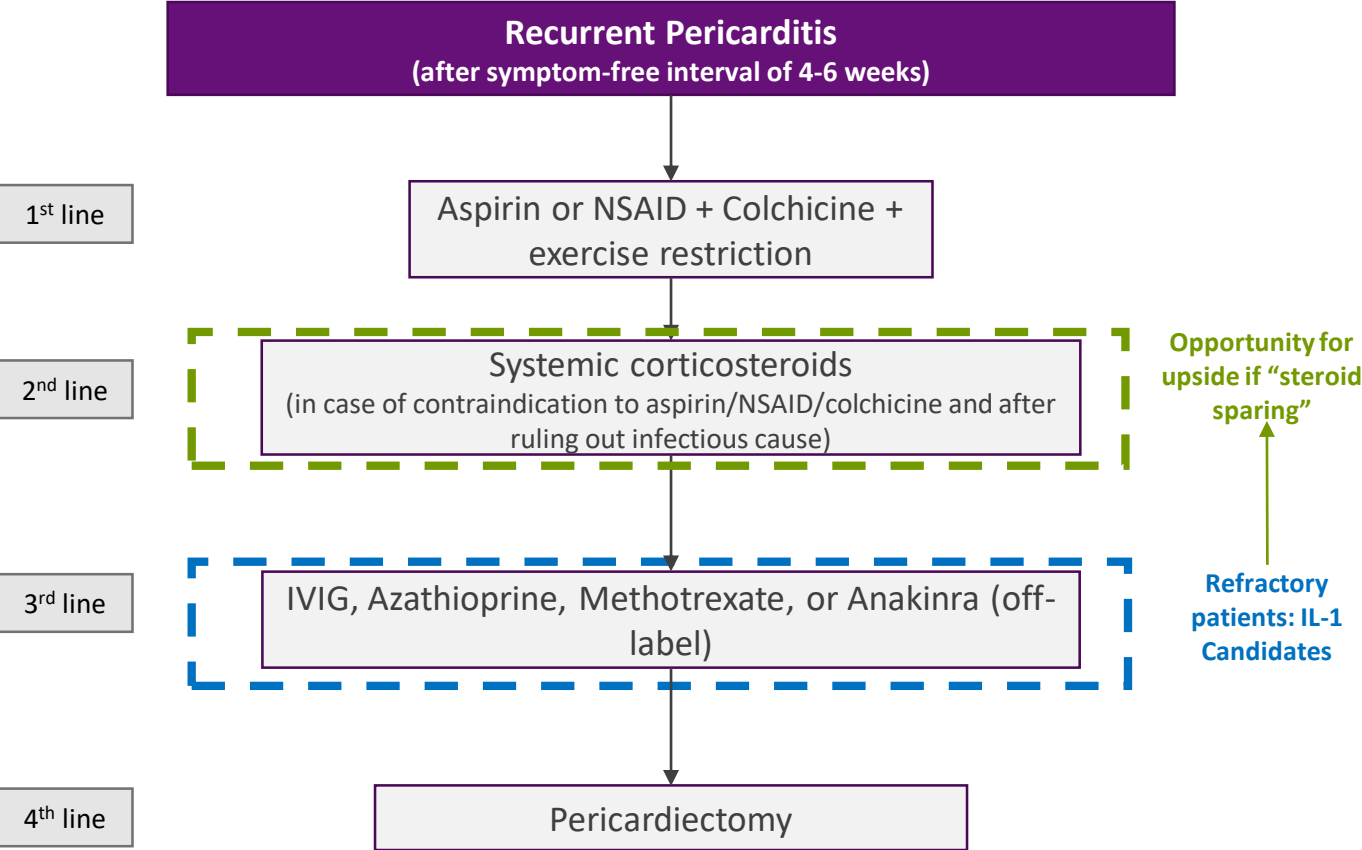
Refractory Pericarditis affects an estimated ~ 3k patients in the U.S. with an additional 9k who are not well controlled



Sources: UptoDate, Trinity Partners, Mayo Clin Proc. 2010 ;85 (6): 572-593; New Diagnostic Criteria for Acute Pericarditis: A Cardiac MRI Perspective, 2015 American College of Cardiology

Refractory patients are left with few treatment options, and rilonacept could mitigate the dangers of long-term steroid use

Recurrent Pericarditis Treatment Paradigm

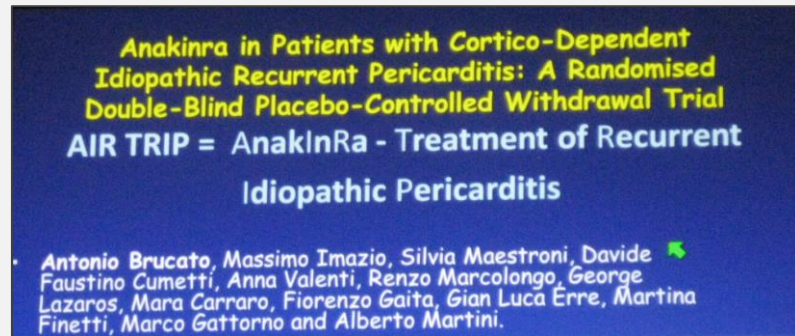


Notes

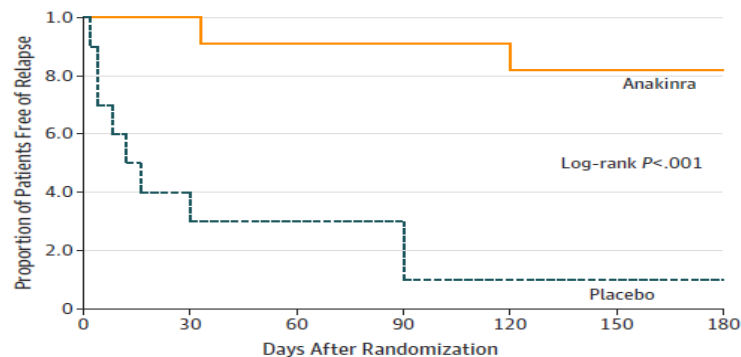
- No known current therapies have an FDA label for recurrent pericarditis
- Treatment is based on historical, empirical data
- Some patients are adequately treated on 1st line therapy
- Colchicine use leads to GI intolerance / diarrhea and discontinuation in ~10% of patients
- Data suggest steroid use may actually make recurrences more common
- Rilonacept could mitigate the dangers of long-term chronic steroid use

IL-1 α signaling is important in the pathology of recurrent pericarditis & is an important cytokine involved in multiple diseases involving sterile, serosal inflammation

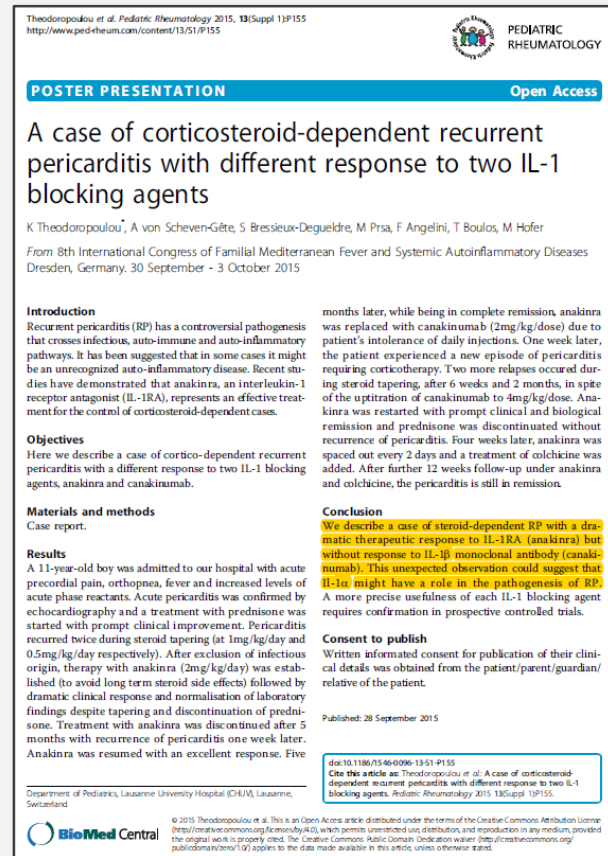
Proof-of-concept in RIP shown by anakinra (daily sc-administered IL-1 α and IL-1 β blocker) in AIRTRIP study



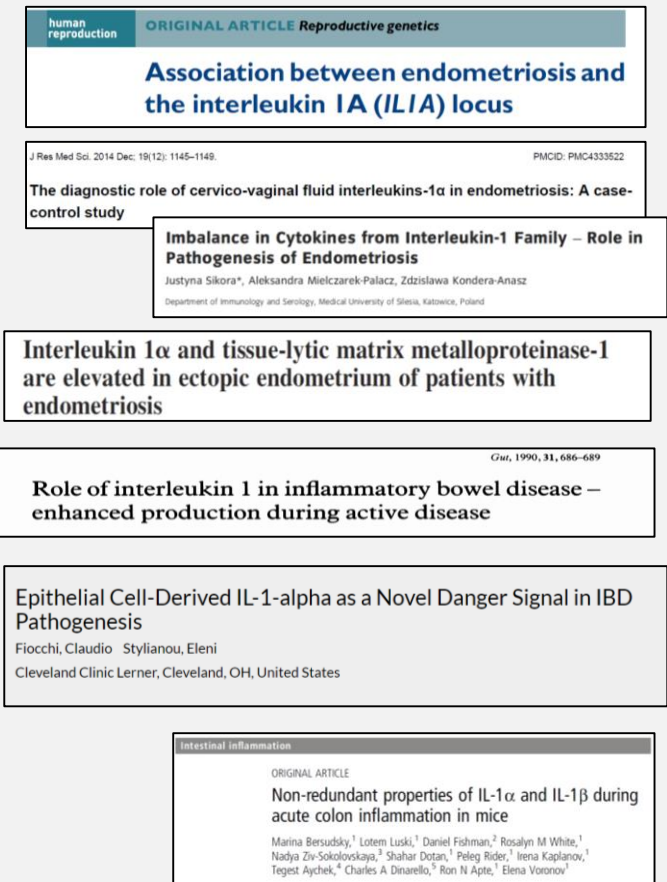
Patients With Recurrent Pericarditis Free of Relapse in the Double-Blind Withdrawal Phase, From Day 0 to Day 180 After Randomization (Intention-to-Treat Analysis)



Kiniksa key insight #1 → Ilaris® (IL-1 β -only inhibitor) does not work in this patient population



Kiniksa key insight #2 → IL-1 α signature across multiple diseases of sterile serosal inflammation

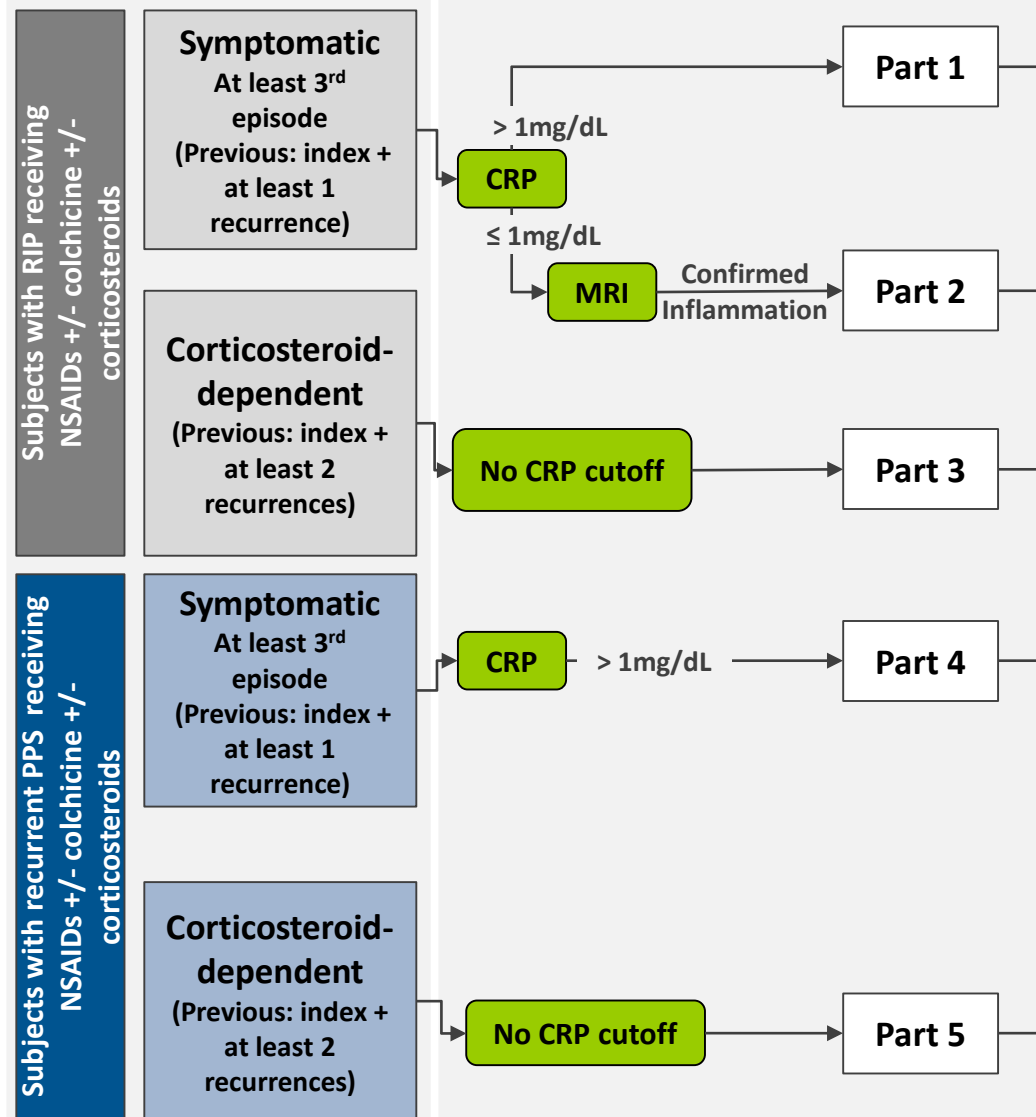


Sources: Brucato et al. JAMA. 2016, 316 (18): 1906-1912; Theodoropoulou et al. Pediatric Rheumatology 2015, 13 (Suppl 1): 155; Sapkota et al. Human Reproduction, 2015, 30 (1) 239-248; Mardanian et al. J Res Med Sci 2014, 19 (12), 1145-1149; Sikora et al. Am J of Reproductive Immunology, 2012, 68, 138-145; Hudelist et al. Human Reproduction 2005, 20 (6), 1695-1701; Ligumsky et al, Gut 1990, 31 (6) 686-689; Fiocchi et al. CTSC Project, Bersudsky et al. Gut 2014, 63, 598-609

Phase 2 study of rilonacept in pericarditis populations

Subjects 6 to 75 yrs old

Distinct Trial Populations



All three patients dosed to date in Part 1 have demonstrated reductions in NRS and CRP values with a safety and tolerability profile consistent with the label for rilonacept

Screening Period

Collect baseline data
No Wash-off

Open-Label Treatment Period

Dose: Adults: Loading Dose 320 mg (2x160mg) then 160mg/week for adults (≥ 18 years of age); Pediatric subjects (6 to <18 years of age): Loading Dose 4.4 mg/kg (2x2.2mg/kg) then 2.2mg/kg/week

Extension Period

Weaning of background therapy over 6 weeks (or at investigator discretion)

Current Tx.
(≤ 72 hrs)

Rilonacept
(6 doses)

Up to 18 weeks

↑
Loading
Dose

↑
2nd
Dose

↑
3rd
Dose

↑
4th
Dose

↑
5th
Dose

↑
6th
Dose

**Mavrilimumab
(KPL-301)**



Phase 2-ready

KPL-716

KPL-045

KPL-404

Phase 2 safety and efficacy data in over 550 rheumatoid arthritis subjects in the EU and mechanistic rationale for focusing on high unmet need vasculitides & inflammatory cardiomyopathies

Lead Indication

- Giant Cell Arteritis (GCA)

Patient Population¹

- ~75k - 150k prevalent GCA patients in the US typically treated with steroid; similar prevalence in EU

Mechanism of Action²

- Monoclonal antibody inhibitor targeting GM-CSFR α
- Blocks GM-CSF signaling, a key mediator of inflammation and autoimmunity

Competition³

- Potential first-in-class molecule
- One FDA approved therapy for GCA but there is still a persistent unmet need

Clinical Development

- Plan to file a new IND and CTA in GCA in 2H 2018
- Plan to commence a Phase 2 proof-of-concept trial in 2H 2018

1) Chandran et al., Scand J Rheumatol, 2015; Trinity Consulting – HCUP/Medicare Data, Quantitative Survey (n=102 rheumatologists); 2) Sources: Wicks, Roberts, Nature Review Immunology, 2015; Hamilton, Expert Review of Clinical Immunology, 11:4, 457-465; 3) Cortellis, UpToDate; Correspondence, Trial of Tocilizumab in Giant-Cell Arteritis, NEJM, 2017

GCA is the most common form of primary systemic vasculitis

The disease is characterized by inflammation of medium-large blood vessels with predilection for the cranial branches of the carotid artery

- **Giant Cell Arteritis (GCA) is a disease that targets older adults**

- The mean age at diagnosis is 72 years, and the disease rarely occurs in individuals younger than 50
- Prevalence in the US is estimated to be ~75k-150k

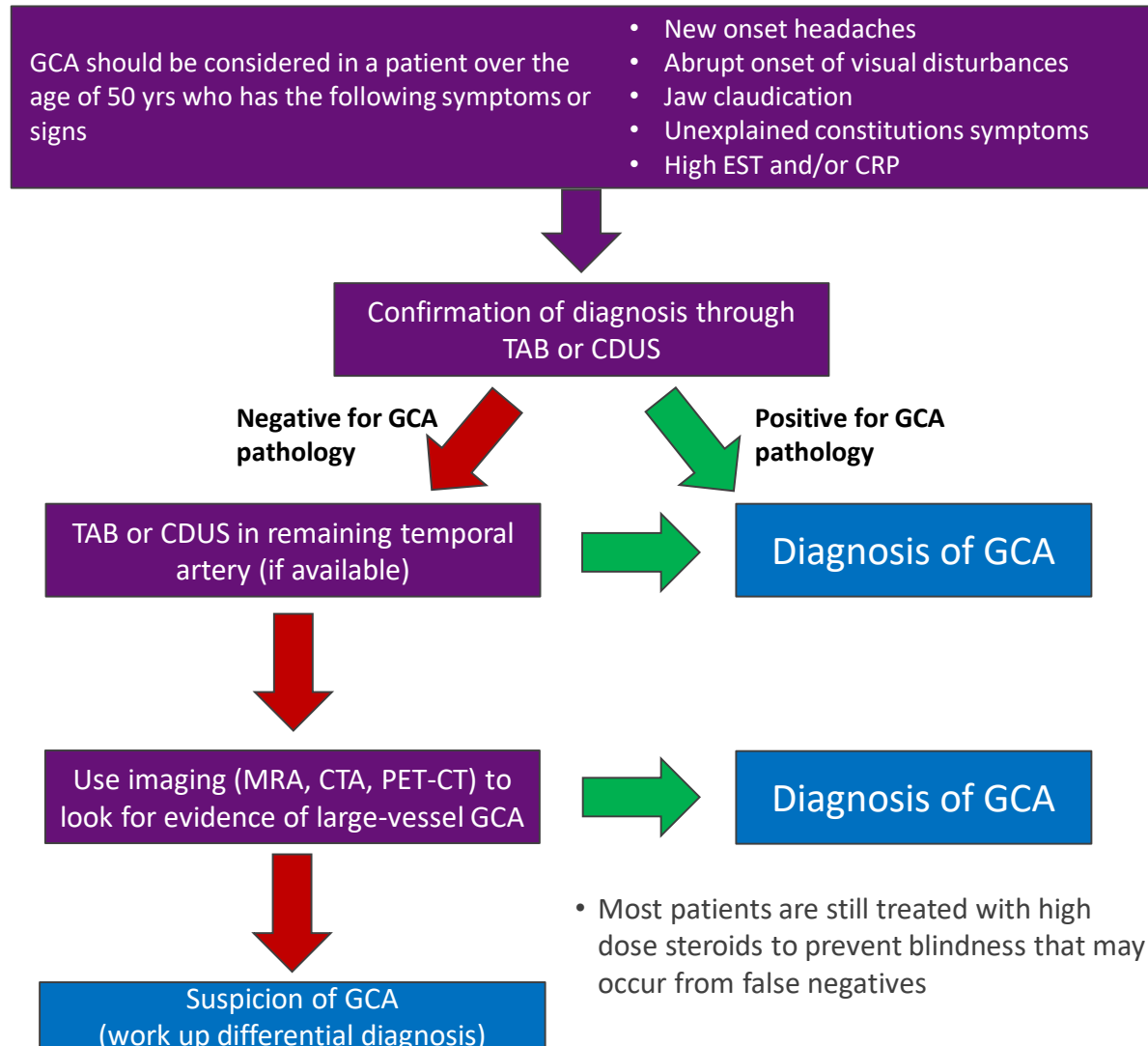
- **Symptoms include fever, fatigue, weight loss, headache, and jaw claudication; transient monocular visual loss (TMVL) and anterior ischemic optic neuropathy (AION); diagnosis is confirmed by biopsy**

- Inflammation of tiny muscular arteries from cranial branches of the aortic arch gives rise to most characteristic symptoms
- If left untreated, bilateral blindness will develop in 25-50% of patients who present with loss of vision in one eye

- **A significant proportion (~50%) of patients have disease relapses or more chronic disease and require high doses of prednisone for a number of years to control symptoms**

- Data show that a subset of patients continue to develop visual symptoms despite long-term, high-dose steroid therapy, suggesting that steroids do not affect underlying disease process in all patients

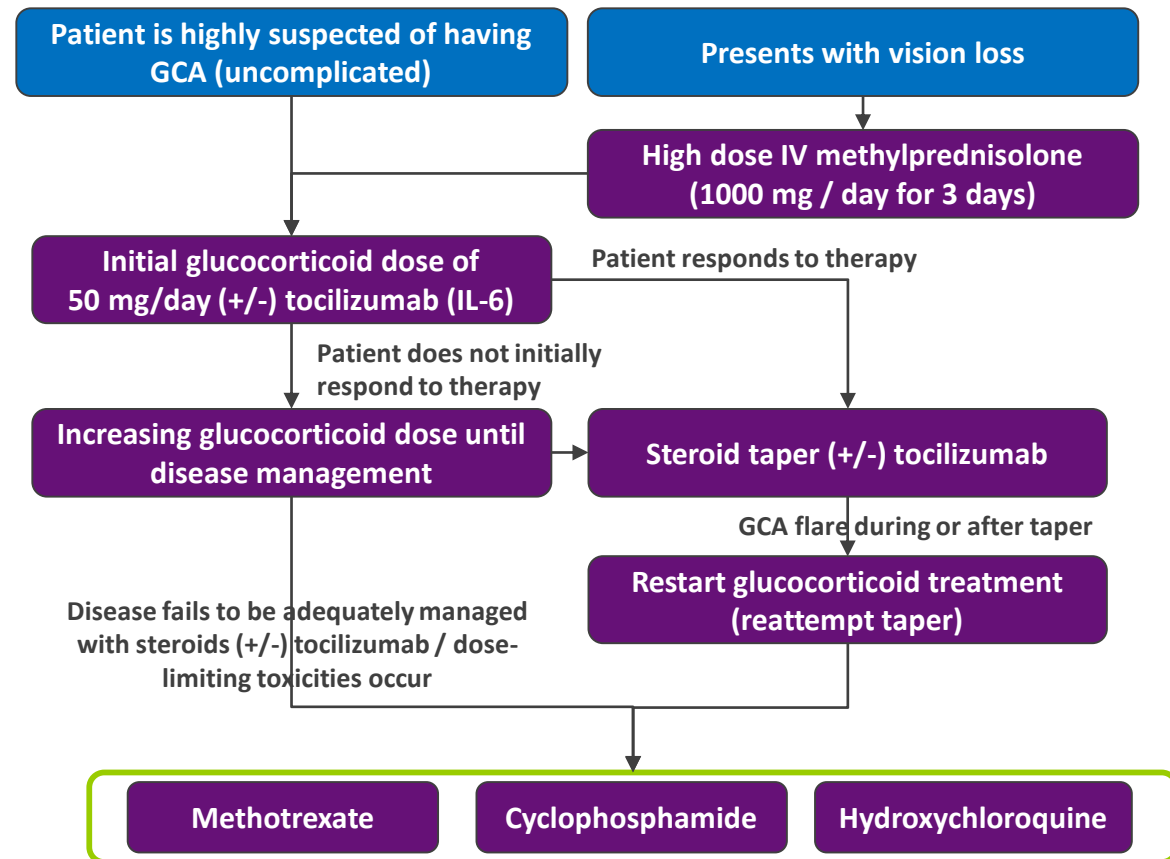
Diagnosis Algorithm for GCA



Incidence of GCA is ~18 per 100k in pts >50yrs old (US prev. of ~75k-150k)

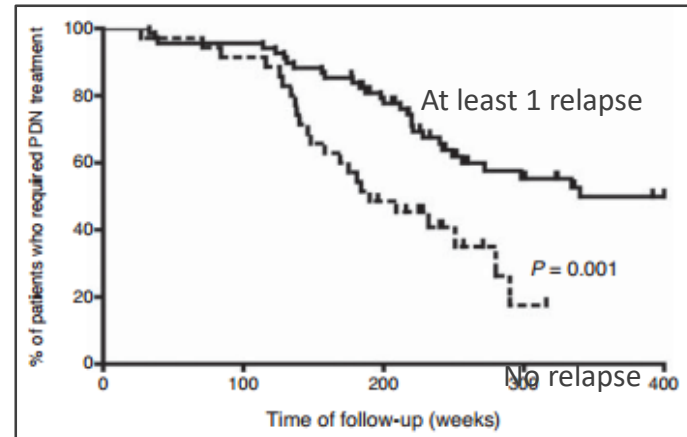
The majority of patients will relapse causing them to become dependent on chronic steroids

GCA Treatment Algorithm

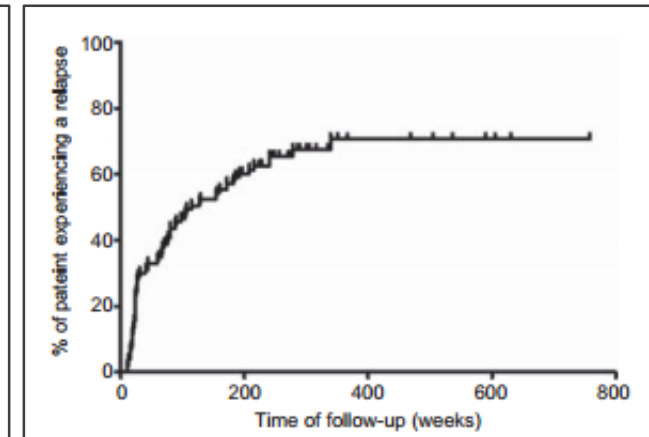


Steroid Treatment is Insufficient

% of GCA Patients on Steroids



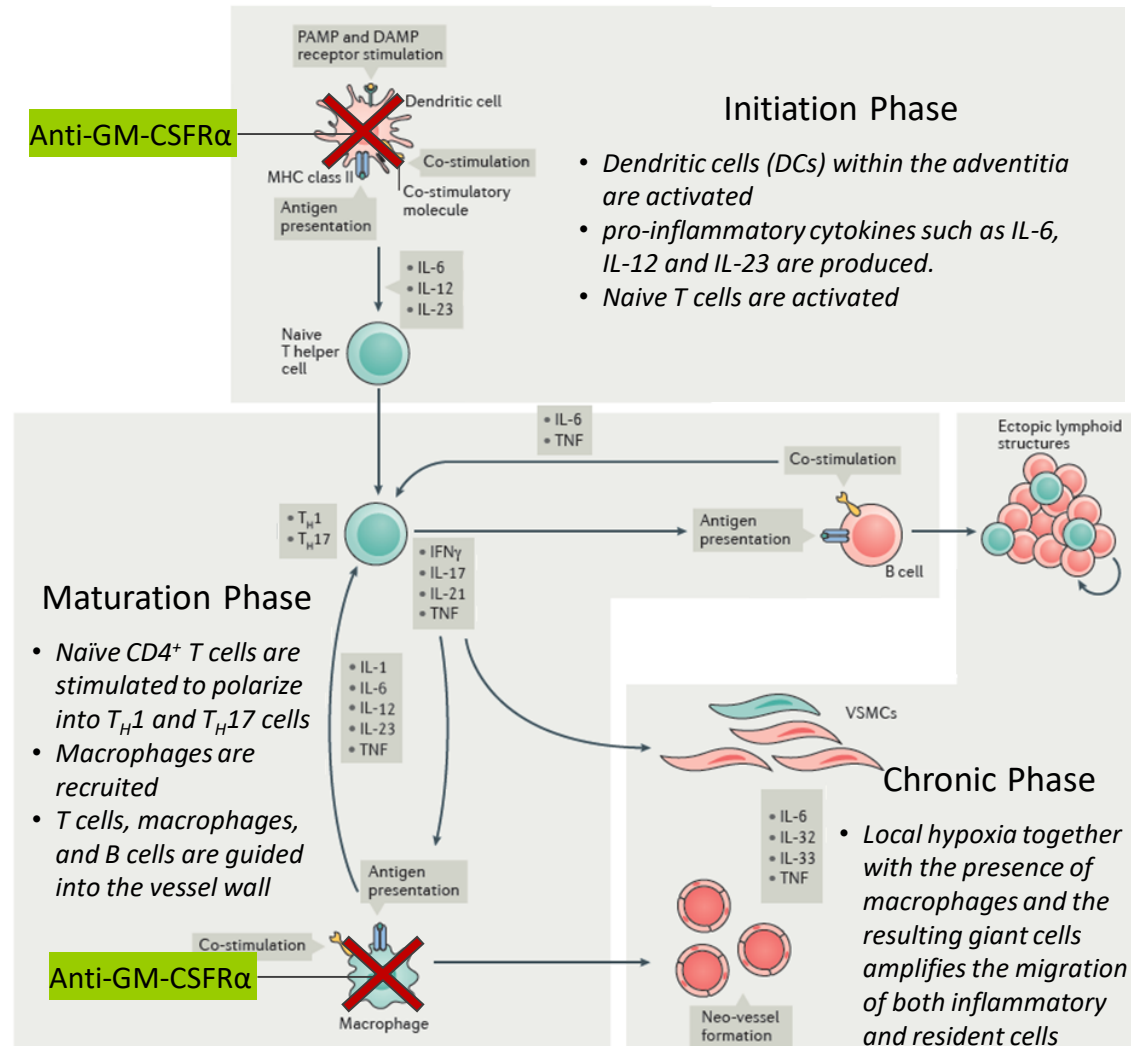
% of GCA Patients who Relapse Over Time



- According to a cohort study that followed 106 patients over 4.5-10.1 yrs, sixty-eight patients (64%) experienced at least 1 relapse, and 38 (36%) experienced 2 or more
 - Cumulative prednisone dose during the first year was significantly higher in relapsing patients (6.2 ± 1.7 g vs 5.4 ± 0.78 g, $p=0.015$).
- The prevalence of GCA is expected to increase due to aging of the population
 - The projected worldwide disease burden of GCA by 2050 is >3 M, and a projected ~500 K people will be visually impaired due to GCA by 2050

GM-CSF implicated in the pathology of giant cell formation by promoting activation, differentiation, survival and proliferation of key cell types

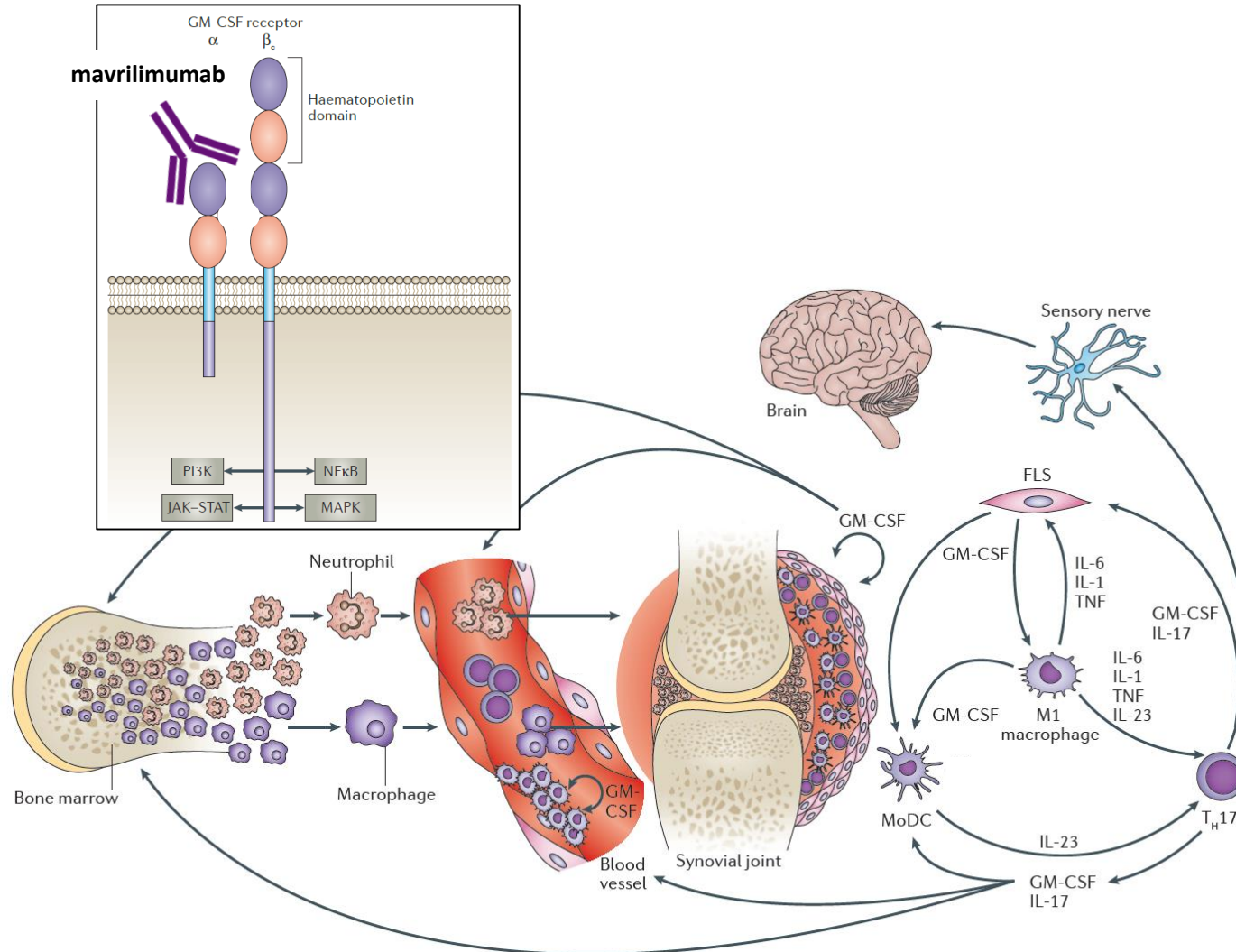
Pathology of GCA



- GM-CSF is critical for the stimulation of hematopoietic progenitor cells in the bone marrow to generate differentiated myeloid-cell progeny (dendritic cells [DCs], neutrophils, eosinophils and monocyte/macrophages)
- GM-CSF enhances trafficking of these cell types through activated endothelium blood vessels, directly contributing to myeloid cell accumulation in blood vessels
- GM-CSF promotes activation, differentiation, survival and proliferation of trafficking monocytes and macrophages as well as resident tissue macrophages in inflamed tissues
- As shown in the diagram, DCs and macrophages are critical in all three stages of GCA pathology; therefore, blockade of GM-CSF with mavrilimumab has significant potential to attenuate both new-onset and relapsing disease
 - In the chronic phase GM-CSF promotes giant cell formation, the hallmark pathologic finding of GCA, from activated macrophages
- Further evidence is provided by data showing elevation of GM-CSF in the lesions of GCA patients

Sources: Weyand, Ann Int Med 121:484, 1994; Watanabe Joint Bone Spine, in press; Wicks, Roberts, Nature Reviews Rheumatology, 2016; Dejaco et al., Nature Reviews Rheumatology, 2017

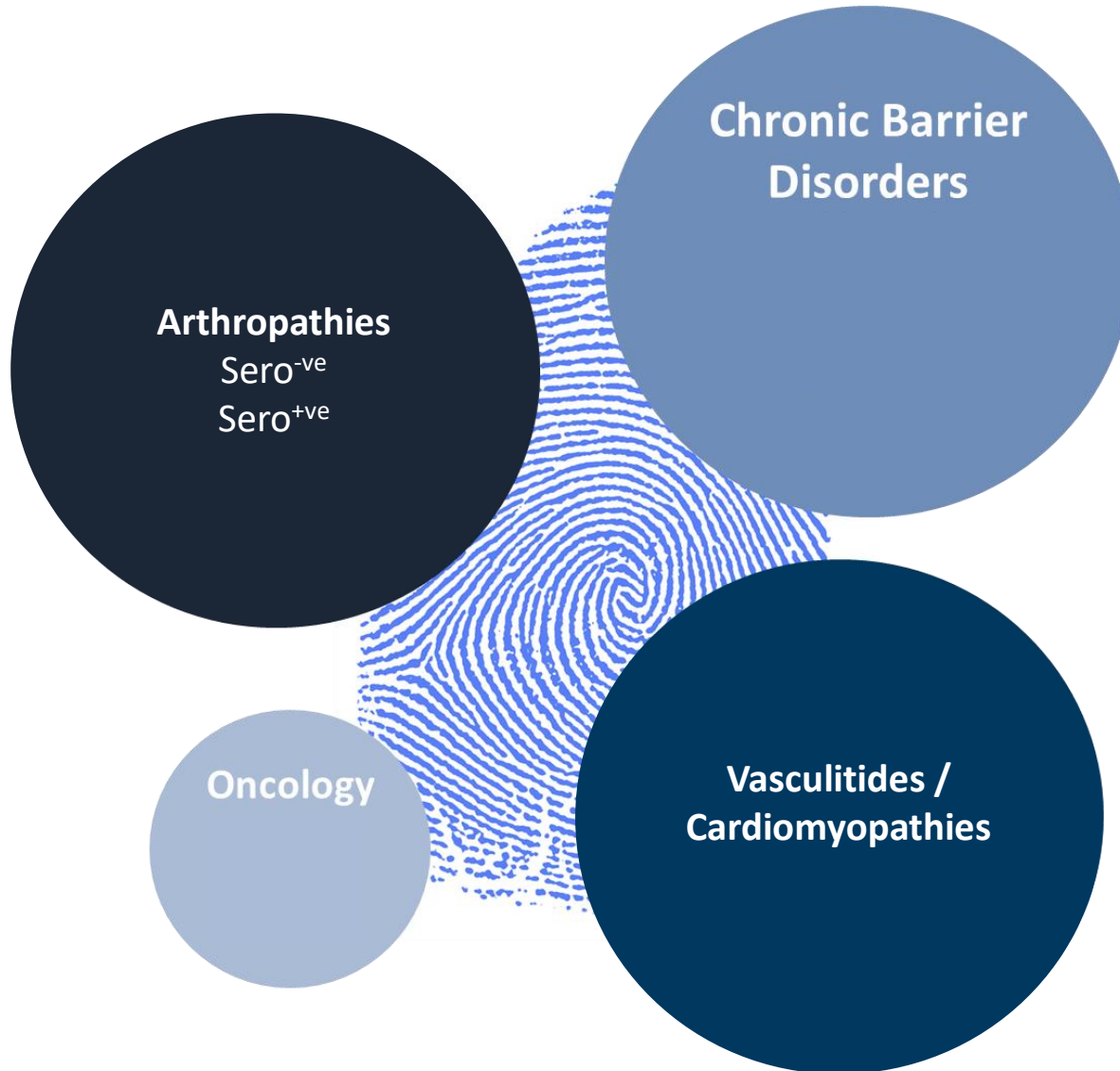
Mavrilimumab, a GM-CSFR α antagonist, blocks GM-CSF signaling – a key mediator of inflammation and autoimmunity



- **Granulocyte-macrophage colony stimulating factor (GM-CSF)** is a growth factor first identified as an inducer of differentiation and proliferation of myeloid cells (neutrophils, eosinophils, and monocytes/macrophages)
 - Activated macrophages produce proinflammatory cytokines such as TNF, IL-6, IL1, lipid-derived mediators and chemokines
 - Downstream signaling is mediated by STAT5, JAK2, NF- κ B, PI3K
- **Data suggest GM-CSF signaling plays a role in several additional cell types including, antigen-presenting cells, T-cells, and B-cells**
 - GM-CSF has a range of functions on mature eosinophils including dose-dependent eosinophil priming, migration, and degranulation
- **GM-CSF is involved in a wide range of biological processes in both innate and adaptive immunity**

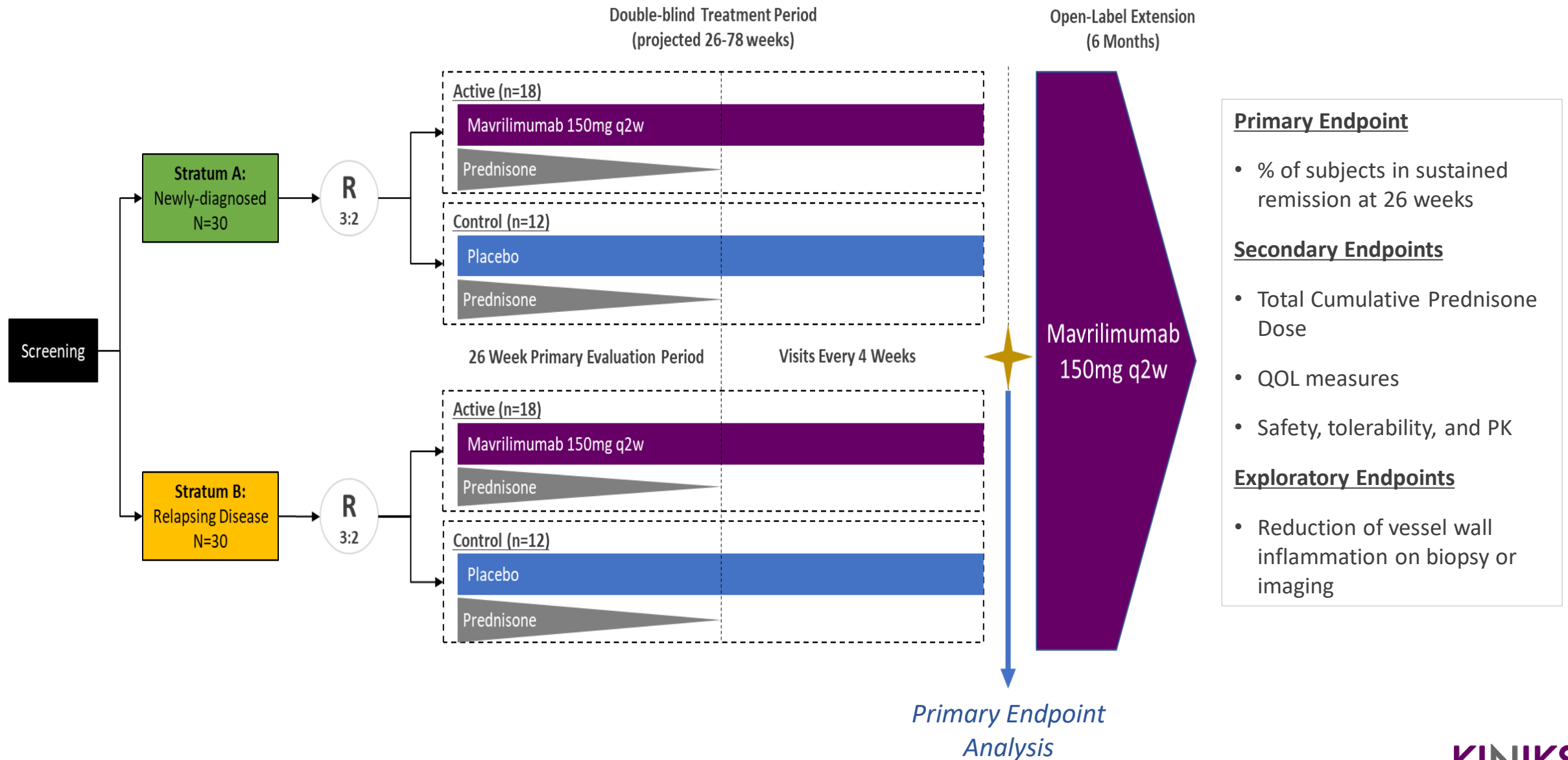
Potential for multiple indications

Following GM-CSF fingerprint across multiple potential additional indications



- **Scientific rationale for studying GM-CSF antagonism across a host of immune mediated diseases**
 - Unmet medical need exists in majority of potentially applicable indications
- **Phase 2 drug supply available to begin studies**
- **To-date, mavrilimumab has been studied in over 550 patients; has achieved primary and secondary endpoints in multiple P2 RA trials**
 - Proof of biologic activity and target engagement provides rationale for additional clinical development

Preliminary Phase 2 GCA study design



Rilonacept
Approved for CAPs in US

Mavri

KPL-716



Phase 1a/1b

KPL-045

KPL-404

A potentially differentiated molecule for a variety of pruritic and fibrotic indications

Lead Indications

- Prurigo Nodularis (PN)
- Atopic Dermatitis (AtD)

Patient Population¹

- PN: Estimated ~300k patients in the US
- AtD: Estimated ~1 M moderate-to-severe patients in the US; ~300k of which are eligible for systemic biologics

Mechanism of Action²

- Monoclonal antibody inhibitor of signaling through OSMR β
- OSMR β is a key receptor subunit used by two inflammatory cytokines, IL-31 and Oncostatin M

Competition³

- Potential for differentiated efficacy and safety
- Competitors block either IL-31 or OSM alone

Clinical Development

- Completed enrollment of single dose Phase 1a/1b in NHV/AtD as proxy for IL-31 driven pruritus; data expect in 2H 2018
- Plan to commence a repeat single dose of Phase 1b AtD patients in 2H 2018

1)Trinity Consulting - HCUP/Medicare Data 2012/2013; Quantitative Survey (n=100 dermatologists); Dantas, 2015, "Prevalence of dermatoses in dermatologic evaluation requests from patients admitted to a tertiary hospital for 10 years"; Mortz et al., Britis Journal of Dermatology, 2001; 2) Trinity Qualitative Interviews; 3) Simpson et al., N Engl J Med, 2016; Ruzicka et al., N Engl J Med, 2017; Reid et al., 2016 ACR Abstract # 1881; Cortellis

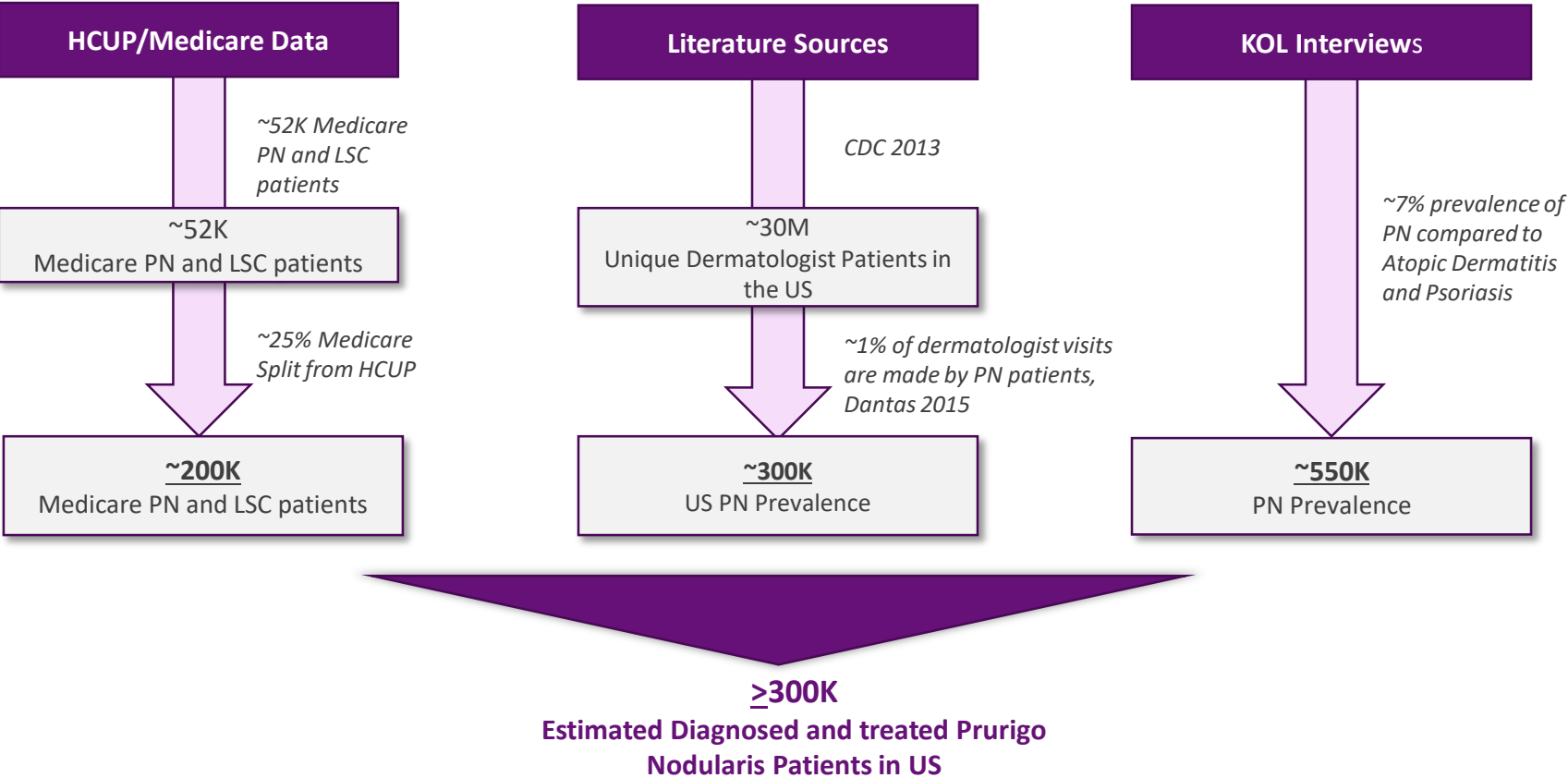
Prurigo Nodularis (PN) is a chronic, serious and debilitating condition with no currently approved FDA treatments

- IL-31 and OSM are markedly increased in PN skin and lesions
- PN rated by KOLs as **more debilitating and serious than psoriasis and atopic dermatitis**
- Interferes with sleep, causes exhaustion and social embarrassment
- Many PN patients are not in the healthcare system due to lack of adequate treatment options, so the prevalence could be significantly higher

Long-lasting, severe, unremitting pruritus; mechanical trauma to the skin via itching then causes nodules

Exact PN epidemiology is uncertain, but estimated prevalence is $\geq 300k$ in the US

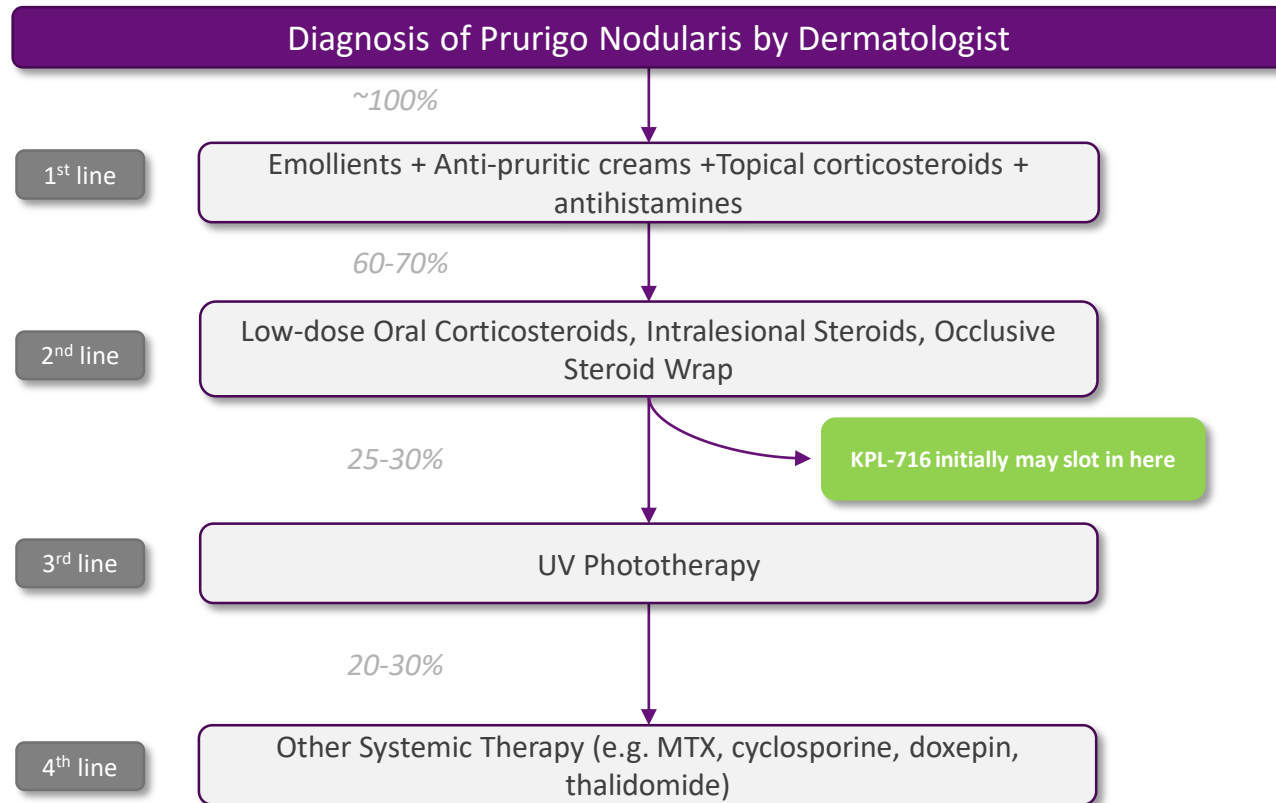
Prevalence of prurigo nodularis was assessed using literature sources; KOL interviews, HCUP and Medicare were used to triangulate literature estimates



KOLs estimated that there are **at minimum 300K** PN patients in the total US population; **the prevalence is probably higher**, given that **many PN patients are not in the healthcare system** and are not seeking treatment, and are thus not accounted for; additionally, KOLs confirmed that 100% of PN patients who go to a dermatologist are treated with at minimum first line therapies

Triangulation of epidemiology sources results in an estimated prevalence of $\geq 300K$ prurigo nodularis (PN) patients in the US

PN Therapeutic Algorithm



Notes

- Inexpensive
- Not effective for all patients
- Inexpensive
- Not effective for all patients
- Antihistamines have potential to cause drowsiness
- Not effective for severe patients
- Inconvenient, requires 3 trips to derm. office per week
- Not effective for all patients
- Potentially dangerous side effects
- Typically only used short-term

Note: none of the above therapies are approved specifically for prurigo nodularis

Prurigo Nodularis is currently treated by dermatologists, who use a combination of several medications and behavioral therapies; treatment is usually unsuccessful

Literature support for IL-31 and OSM signaling through OSMR β receptor driving distinct and overlapping inflammatory pathologies

IL-31 is a robust pruritogen

- IL-31 delivery (independent of mode of administration) leads to pruritus in multiple animal species (mouse, dog, cyno, human)^{1,2,3,4,7}
- Serum and skin IL-31 levels correlate with pruritus intensity in multiple animal species^{2,4,5}
- IL-31R α is located on peripheral nerve endings, as well on the cell body within the dorsal root ganglia (DRG)^{4,7}
- IL-31 induces increases cutaneous nerve fiber density, as well as nerve elongation and branching both in vitro and in vivo⁶
- IL-31 levels are elevated in skin samples from atopic dermatitis and Prurigo Nodularis patients⁷
- Nemolizumab, an anti-IL-31R α antagonist, demonstrated in Ph2 atopic dermatitis trials reduction of NRS scores by up to 90% over 52 weeks⁸

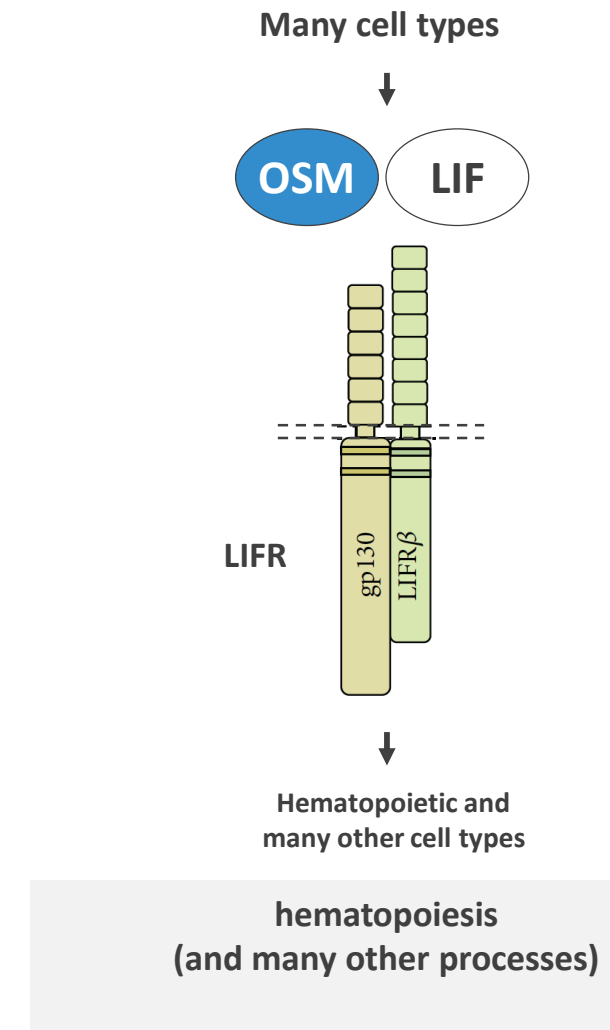
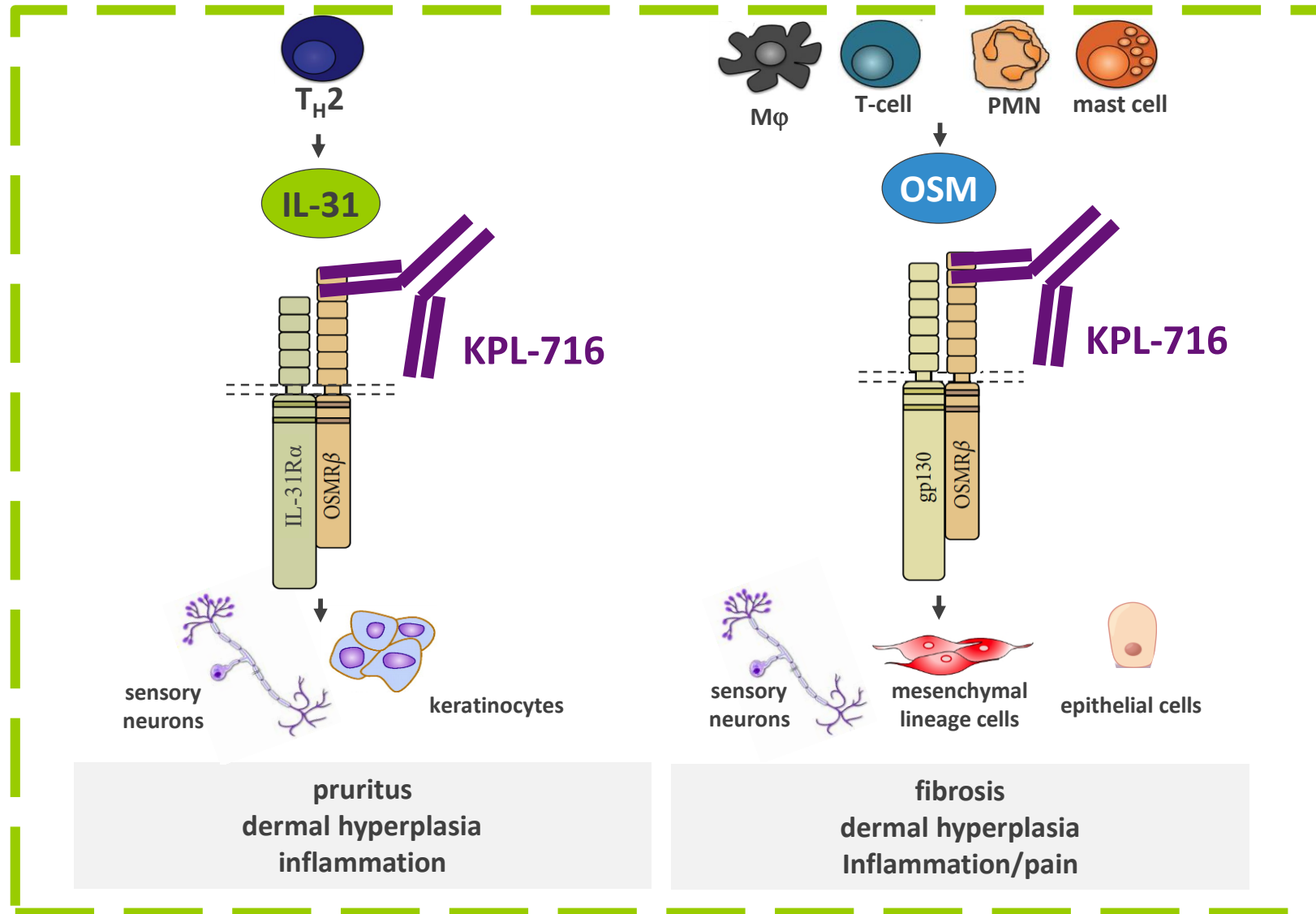
OSM acts upstream of T_H2 and drives barrier dysfunction and TGF β -independent fibrosis

- OSM and OSMR expression levels are elevated in systemic sclerosis skin^{9,10}
- OSM is a key driver of keratinocyte dependent pro-fibrotic activity^{11,13}
- OSM contributes to dermal hyperplasia and inflammation^{10,11,12,13}
- OSM acts to upregulate IL-4R α / Th2 response and is present in the lesions of AtD skin^{13,14,15}
- Interstitial lung disease in humans is associated with elevated levels of OSM¹⁶
- Intranasal OSM injection leads to lung inflammation and fibrosis¹⁶
- OSM promotes TGF β -independent extracellular matrix (ECM) over-production in lung fibroblasts^{16,17}
- OSM promotes mucosal epithelial barrier dysfunction¹⁷

1. Dillon et al., Interleukin 31, a cytokine produced by activated T cells, induces dermatitis in mice, Nat Immunol. 2004
2. Oyama et al., Cynomolgus monkey model of interleukin-31-induced scratching depicts blockade of human interleukin-31 receptor A by a humanized monoclonal antibody, Exp Dermatol. 2016
3. Gonzales et al., Interleukin-31: its role in canine pruritus and naturally occurring canine atopic dermatitis, Vet Dermatol. 2013
4. Cevikbas et al., A sensory neuron-expressed interleukin-31 receptor mediates T helper cell-dependent itch: involvement of TRPV1 and TRPA1, J Allergy Clin Immunol. 2014
5. Marsella et al., Investigation of the correlation of serum IL-31 with severity of dermatitis in an experimental model of canine atopic dermatitis using beagle dogs, Vet Dermatol. 2017
6. Feld et al., The pruritus- and Th2-associated cytokine IL-31 promotes growth of sensory nerves, J Allergy Clin Immunol. 2016
7. Sonkoly et al., IL-31: a new link between T cells and pruritus in atopic skin inflammation, J Allergy Clin Immunol. 2006
8. Kabashima, 5268 - Efficacy and safety of nemolizumab over 64 weeks in patients with moderate-to-severe atopic dermatitis: Results from the long-term extension of the Phase 2 study, AAD 2017 Abstract

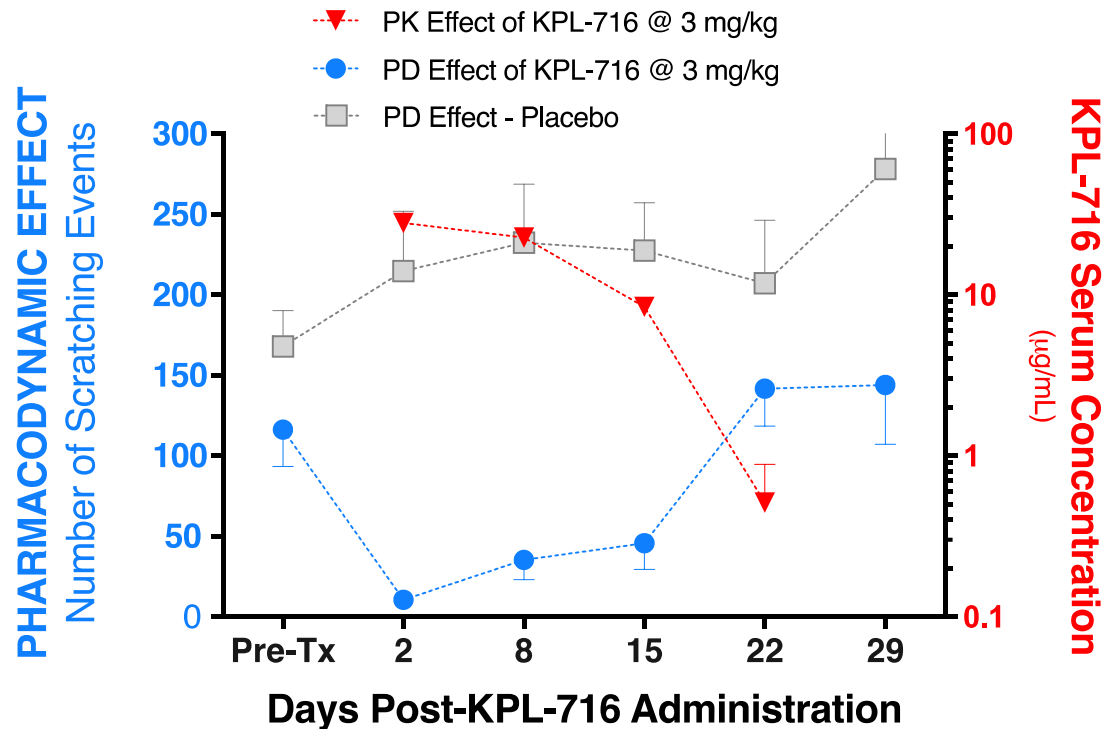
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KPL-716 inhibits IL-31 & OSM signaling through OSMR β but avoids inhibiting signaling critical to hematopoiesis through OSM/LIFR



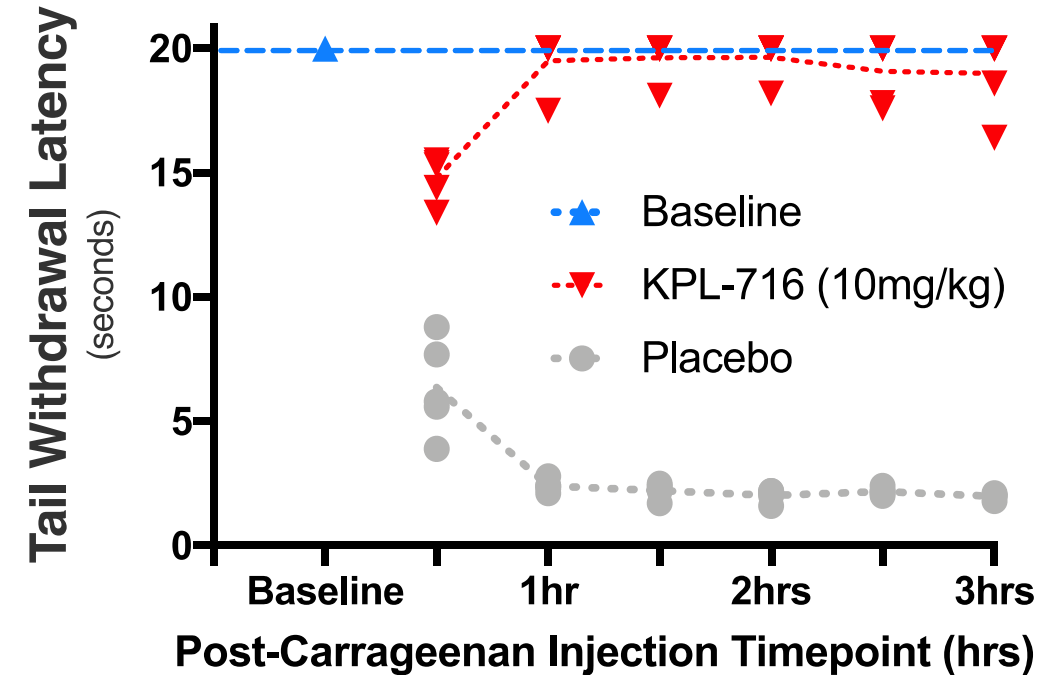
KPL-716 demonstrated potential efficacy in two validated primate models of pruritus and inflammation after a single dose

NHP Model of Pruritus¹



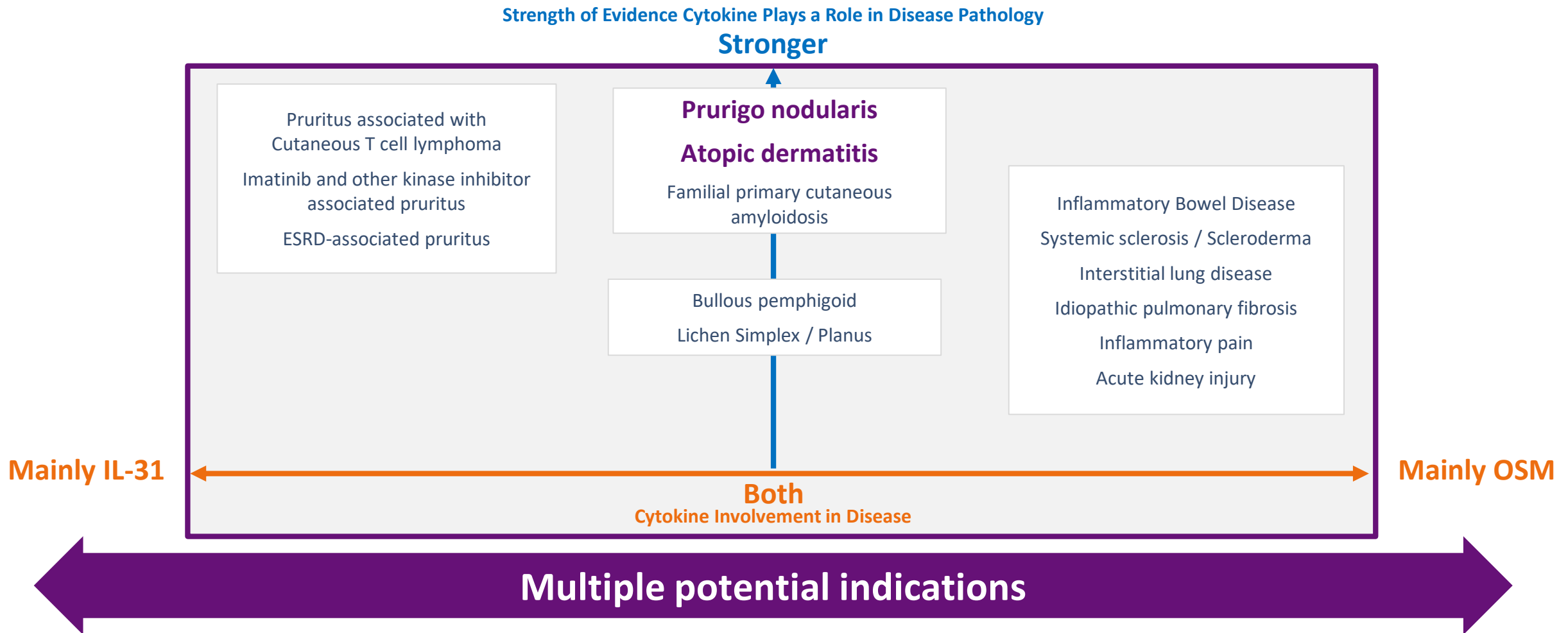
A single dose of KPL-716 inhibited pruritic response driven by supraphysiologic levels of IL-31 for over 2 weeks at 3mg/kg

NHP Model of Inflammation¹



Single dose of KPL-716 increased tail withdrawal latency → implicates OSMR β in the inflammatory response

Inhibiting OSMR β signaling could impact a variety of diseases characterized by pruritus and/or barrier dysfunction and fibrosis

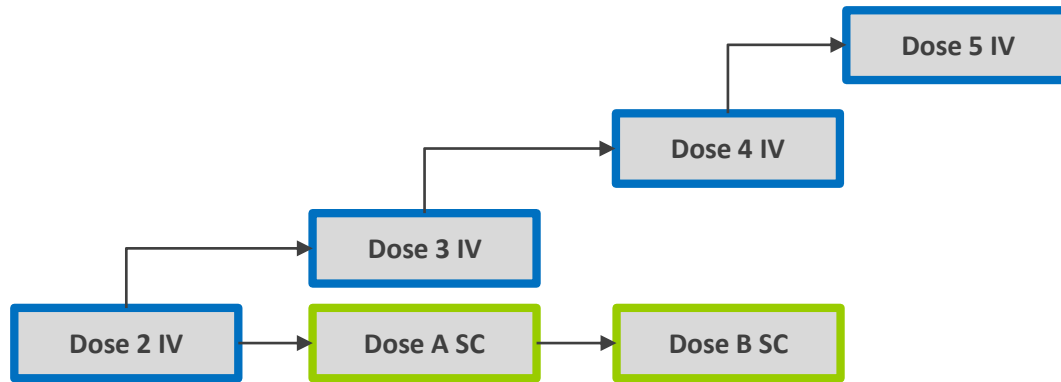


Note: Indicates 2018 clinical activity

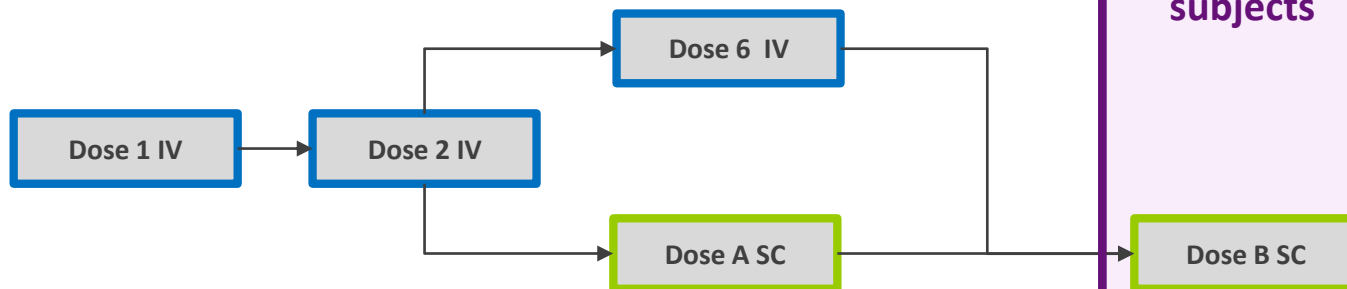
KPL-716 P1 Design & Anticipated Readouts

2H 2018 Single Dose Readouts

Phase 1a: Normal Healthy Volunteer Single Dose Portion (Data 2H2018)



Phase 1b: Subjects with Atopic Dermatitis Single Dose Portion (Data 2H2018)



Repeat Single Dose in AtD subjects

- **Safety in normal healthy volunteers (NHV) and atopic dermatitis (AtD) patients**
- **PK information to inform target mediated drug disposition and PK/PD effect**
 - IV and SC PK in NHV
 - IV and SC PK in AtD patients
- **Clinical efficacy read-outs in atopic subjects in double blind, placebo controlled setting**
 - Numerical Rating Score (NRS) reductions for pruritus → anti-IL-31 agent nemolizumab demonstrated reductions in P1b in atopic patients
 - Other secondary endpoints

Note: Dose # refers to different amounts of KPL-716; dose numbers not associated with dose level

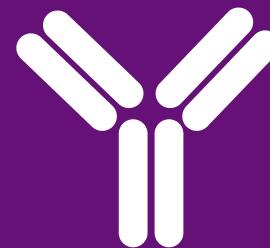
Rilonacept

Approved for CAPs in US

Mavri

KPL-716

KPL-045



Pre-IND

KPL-404

Signal inhibitor of the CD30/CD30L interaction – a T-cell co-stimulatory receptor involved in activated T-memory function

- Biology important for activated T-memory cell function
- Comprehensive non-clinical package and POM established in models
- Favorable pharmacokinetic profile supports further development
- Preparing for IND enabling studies

Rilonacept

Approved for CAPs in US

Mavri

KPL-716

KPL-045

KPL-404



Pre-IND

A central control node of T-cell-dependent, B-cell-mediated humoral adaptive immunity – designed to inhibit CD40/CD40L interaction

- External PoC of CD40 antagonism established by Novartis in autoimmune disease
- Favorable pharmacokinetic profile supports further development
- Preparing for IND enabling studies

Transaction / Terms Summary

Program	Outlicensing Process	Deal Description	Revenue Sharing	Other Remaining Disclosed Payment Terms
Rilonacept	Kiniksa proactively contacted REGN with idea / rationale	- In-license 50/50 profit split - Kiniksa assumes development costs - REGN manufactures product - CAPS in US & Japan transfers to Kiniksa upon BLA approval; REGN retains rights in oncology and local administration to the eye and ear	50/50 Profit Split	Up to \$27.5m in approval milestones in the aggregate
Mavrilimumab	AZ/MedImmune divesting much of its non-respiratory immunology portfolio to concentrate on immuno-oncology	In-license	Tiered royalties of low double digit percentages up to twenty percent on annual net sales of licensed products	\$10m on specified regulatory milestones or by 12/31/2018 - Up to \$72.5m in aggregate for first two indication - Up to \$85.0 million upon the achievement of annual net sales thresholds up to, but excluding, \$1.0 billion in annual net sales - Up to \$1.1 billion upon the achievement of additional annual net sales thresholds starting at \$1.0 billion and higher
KPL-716	Biogen divested its tissue injury and fibrosis portfolio of assets (n=5)	- Asset acquisition - Biogen retains a time-limited right-of-first negotiation if the asset is to be sold	Tiered high-single digit percentages to below the teens royalties on net sales	\$175.0 million in the aggregate upon achievement of specified clinical and regulatory milestone for multiple indications - Up to \$150.0 million in the aggregate upon the achievement of specified annual net sales thresholds.
KPL-045	Novo Nordisk outlicensed all non-core (e.g., metabolic / hormone) therapeutic areas	In-license agreement	Undisclosed royalty	\$1m upon the earlier to occur of a specified regulatory milestone and January 2020 - Undisclosed future milestone payments upon the achievement of specified clinical, regulatory and sales milestones
KPL-404	Virtual company sought team to bring program forward	Option to purchase all outstanding shares	N/A	- Undisclosed option extension fees through January of 2019 - Option exercise fee of \$10m payable in a combination of cash and Class A common shares - Milestone payments of up to \$10m payable in a combination of cash and Class A common shares

Ongoing Trials & Near-term Inflection Points

Program	Activity	Indication	Description	Timing
Rilonacept® IL-1α & IL-1β	Ongoing open-label Phase 2 pilot study	Recurrent Pericarditis	Explore clinical and biochemical endpoints of pericarditis symptomatology and collect inter- and intra-subject variability data on both at-baseline and on-treatment parameters	Additional interim data expected in 2H 2018
	Phase 3	Recurrent Pericarditis	Plan to commence a pivotal Phase 3 clinical trial pending end of Phase 2 meeting with the FDA	2H 2018
Mavrilimumab GM-CSFRα	Phase 2	Giant Cell Arteritis	Plan to commence a Phase 2 proof-of-concept clinical trial	2H 2018
KPL-716 OSMRβ	Phase 1a & 1b single dose	Normal Healthy Volunteers & Atopic Dermatitis subjects as a proxy for IL-31 driven pruritus	Evaluates safety, target engagement and potential for pruritus efficacy signal	Enrollment complete; Data expected in 2H 2018
	Phase 1b repeat single dose	Atopic Dermatitis	Plan to commence a repeat single dose of Phase 1b in Atopic Dermatitis patients to assess multiple dose safety, pruritus and inflammation	2H 2018
KPL-045 CD30L	IND-enabling studies	Autoimmune	Manufacturing scale-up and preparing for IND-enabling studies	IND filing expected in 2019
KPL-404 CD40	IND-enabling studies	Autoimmune	Manufacturing scale-up and preparing for IND-enabling studies	IND filing expected in 2019

Kiniksa at a glance

Every Second Counts!

5 pipeline programs

169 issued patents

~\$490m gross proceeds raised to date

~\$375m¹ post IPO proforma cash and cash equivalents as of 3/31/18 (no debt)

Bermuda based corporate structure

1) Our post IPO proforma cash and cash equivalents includes ~\$155 million from our IPO net of underwriting discounts, fees and estimated offering expenses as though we had closed the IPO of our Class A common shares in Q1 2018 and as such is not prepared in accordance with GAAP.



Every Second Counts!™