Investor Presentation

September 2019

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Treating Orphan Diseases Caused by Complement & Leukotriene Dysregulation

Unique MOA

- **Nomacopan (Coversin):** Bifunctional inhibitor of C5 & LTB4
  Focused on orphan inflammatory & autoimmune diseases where treatment options limited and C5 and LTB4 implicated

Diversified Targets

- **Four orphan conditions currently in Phase II and Phase III studies**
  - Subcutaneous (TMA, BP, & PNH) and topical delivery (AKC)

Clinical Data

- **Positive initial clinical readouts across clinical studies**
- Excellent safety profile; no drug-related SAEs over 20 cumulative patient-years of cumulative patient treatment data

Market Potential

- **$500 million peak sales targets, including follow-on indications**
- Platform molecule with a large and active pipeline

Pathway to Approval

- Potential upcoming value inflection points via multiple mid-stage clinical study readouts in 2019-2020 from TMA, BP, and AKC

TMA: Thrombotic Microangiopathy; BP: Bullous Pemphigoid; AKC: Atopic Keratoconjunctivitis; PNH: Paroxysmal Nocturnal Hemoglobinuria

Dual C5 & LTB4 MOA Provides Unique Option for Multiple Poorly Treated Inflammatory Diseases

Background

- **C5 & LTB4 act synergistically**:
  C5 activates granulocytes, LTB4 promotes granulocyte migration
- **Each pathway proven as treatment option:**
  Eculizumab (Soliris®) – inhibits C5, and Zileuton® inhibits LTB4

Evidence for Treatment

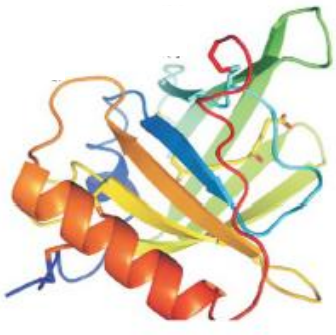
- **Additive effect** demonstrated in preclinical RA, lung and BP models with Nomacopan: significantly more effective than LTB4 or C5 alone
- **Elevated levels of C5a and LTB4** seen in human and animal tissue - blister fluid (BP), eye (uveitis), GVHD (HSCT-TMA)
- **Clinical response seen** across the three target conditions (BP, AKC, HSCT-TMA) where C5 and LTB4 implicated

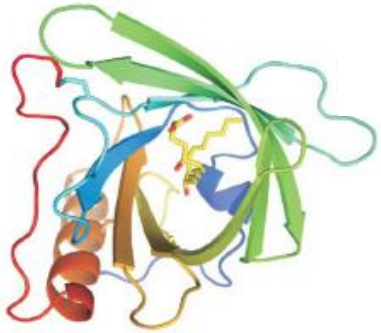
Opportunity

- **Nomacopan: good safety profile in clinical studies to date**
- **Potential to address unmet need across a wide range of conditions**

Nomacopan: Well-Designed by Nature Tightly Binding Both C5 & LTB4

Independent, Highly Specific C5 & LTB4 Inhibitory Activity



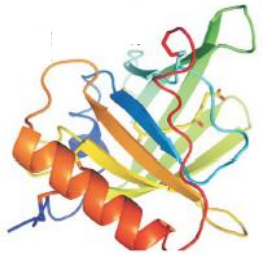


- Nomacopan: biological with high specificity for C5 & LTB4
- Avoids off-target effects by binding ligands not receptors
- No immunogenicity to date in human studies
- Multiple modes of administration (sub Q, topical, nebulized)
- Delivers high degree of patient comfort
  - Sub Q: Highly soluble & low viscosity
  - Topical drops: pH neutral / comfortable

Nomacopan: Extensive and Growing Positive Efficacy Data Across Four Indications

2015-2018		2019	Projected Next Step
9 patients sub Q **~20 cumulative patient-years*** of treatment data **PNH Phase II** **Achieved primary Phase II end point** Six Phase II patients transfusion dependant; two-thirds now transfusion independent	**TMA**	2 patients sub Q – chronic Named patient program	Initiate **pivotal study** Q4 2019
	BP	3 patients sub Q – chronic ongoing Phase II	Completion of 9 patient **Phase II** Q4 2019
	AKC	3 patients topical – chronic ongoing Phase II	Completion of 16 patient **Phase I/II** Q4 2019
	PNH	Phase III study naïve patients	Objective to introduce potential pen injector

** as of September 2019*



Subcutaneous Nomacopan:

Clinically Testing C5 & LTB4 Binding in

HSCT – Thrombotic Microangiopathy

HSCT-TMA
High Mortality Rate – and No Approved Treatments

- Arteriolar thrombi associated with intimal swelling, fibrinoid necrosis, thickening of arterioles and capillaries, and subendothelial accumulation of proteins & cell debris
- Cause of TMA after bone marrow transplant not well understood
  - Can occur in up to 30% patients
  - Typical diagnostic markers include elevated sC5b9 and proteinuria
- Diagnosis & treatment of pediatric TMA-HSCT refined by Dr. Jodele at Cincinnati Children's Hospital
- No approved therapy; current treatment by immunosuppressants
- In severe pediatric cases, mortality ≥80%
- Orphan condition with a prevalence of circa 5,000 treatable patients

Upcoming Pediatric HSCT-TMA Pivotal Trial Potential Expansion to Related Indications

- Complement's role highlighted by elevated terminal complement components (sC5b9) in many patients.
- LTB4 believed to have a role in TMA-HSCT in terms of graft vs. host disease (GVHD) progression
- Nomacopan used on a named patient basis for pediatric HSCT-TMA
- Positive FDA meeting, trial designed around multiple responder endpoints
- Nomacopan has received pediatric Fast Track and Orphan status for adult and pediatric HSCT-TMA
- **Pivotal trial anticipated to commence Q4 2019; primary endpoint TMA response**

Potential expedited approval with a single pivotal study
Gateway condition into other TMAs
Potential first approval for nomacopan

Rosenthal (2016) *Journal of Blood Medicine 7:* 181-186

Activity Demonstrated in HSCT–TMA Across Multiple Responder End Points (First 2 Named Patients)

TMA Marker	Patient	Baseline	Day 7	Day 14	Day 28	Day 60
Hemolytic anemia	1	Yes				Resolved
	2	Mild				
Red blood cell fragments	1	Yes				Resolved
	2	Yes			Resolved	
Thrombocytopenia	1	Yes				Resolved
	2	Yes		Resolved		
Increased LDH	1	Yes			Resolved	
	2	Yes		Resolved		
Proteinuria and/or increased creatinine	1	Yes			Resolved	
	2	Yes				N/A
Hypertension	1	Yes				Resolved
	2	Yes		Resolved		
Neurology	1	Yes		Resolved		
	2	No				
GI bleed	1	No				
	2	Yes			Resolved	

Patient 1 : treated at GOSH made a complete recovery and nomacopan was discontinued after seven weeks
Patient 2 : despite resolution of the TMA markers, patient died at day 63 of lung damage considered unrelated to treatment with nomacopan. Note - data for patient 2 is up to day 28

Nomacopan in HSCT-TMA
Differentiation Against Competitors

Clear Positioning

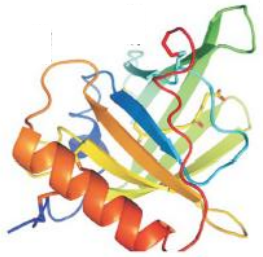
- Patient group to be treated identified via histologic TMA diagnosis in tissue biopsy or laboratory & clinical markers

Differentiated MOA

- Daily dosing facilitates more complete complement suppression
- LTB4 inhibition may further reduce epithelial activation

Next Steps

- **Pivotal pediatric study scheduled to commence Q4 2019**
- Responder-based study based on achieving end points related to disease resolution
- Working with FDA on optimizing dosing through the Model-Informed Drug Development (MIDD) Pilot Program



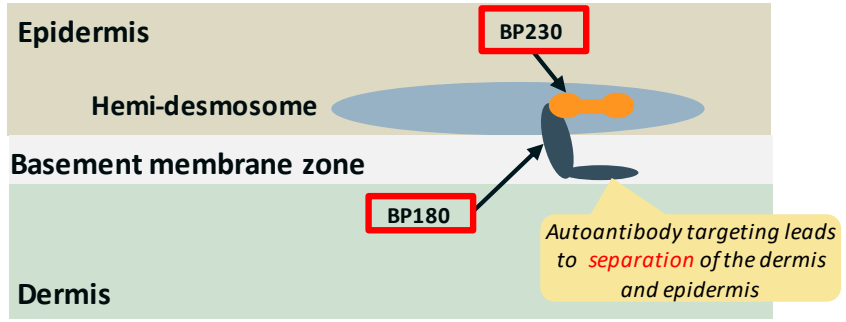
Subcutaneous Nomacopan:

Clinically Testing C5 & LTB4 Binding in

Bullous Pemphigoid

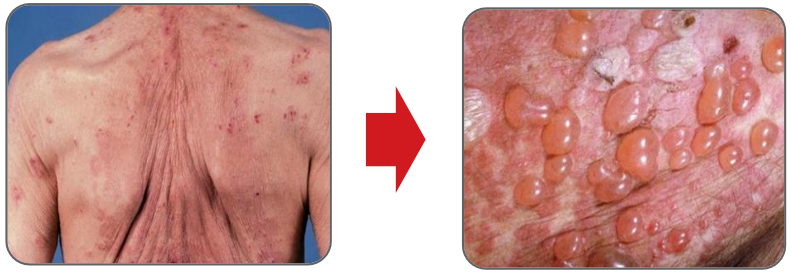
Bullous Pemphigoid
Significant Unmet Need

Schematic of dermal-epidermal junction



Key molecular targets that lead to BP pathophysiology

Presentation of bullous pemphigoid



Urticarial-like skin rash

Fluid-filled blisters

- Severe autoimmune blistering skin disorder primarily of the elderly
- Caused by immune complex mediated inflammation via autoantibodies
- No approved therapy. Current treatment with chronic corticosteroids poses substantial health concerns
- KOLs: essentially no change in standard of care in decades
- Understanding of BP has grown, but specific therapies still lacking
- Orphan condition, with a prevalence of circa 120K patients in US and EU

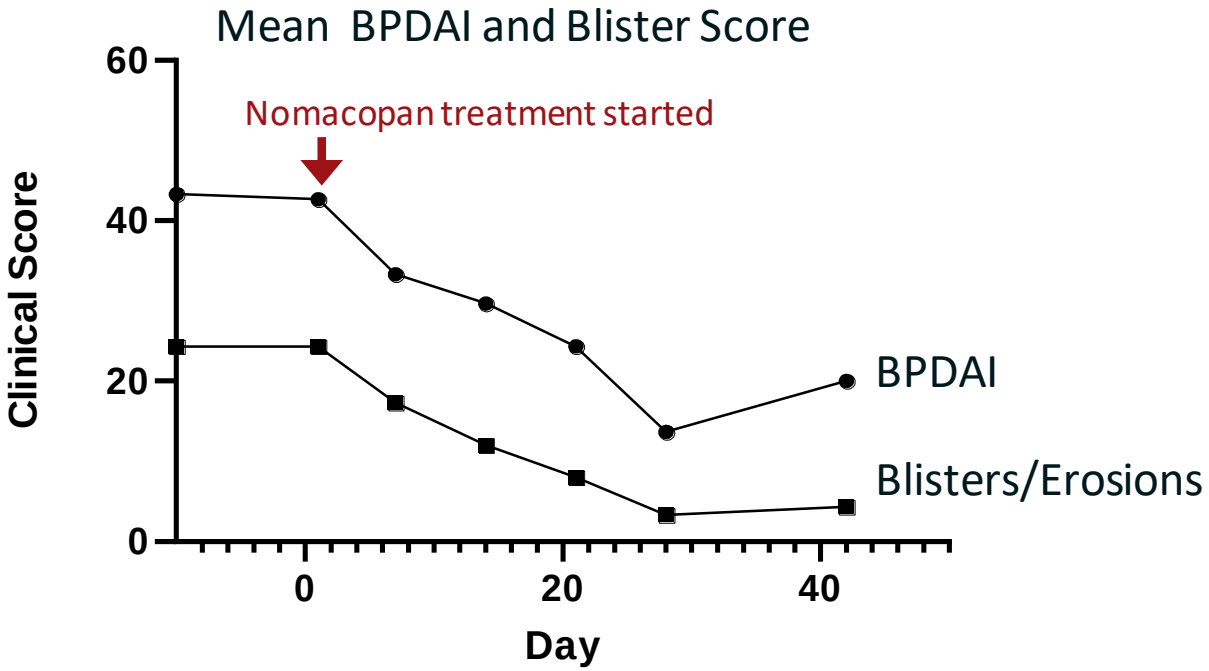
Bullous Pemphigoid
Evidence of Combined C5 & LTB4 Involvement

- Orphan condition in which there is evidence that both terminal complement activation (C5) and LTB4 have a role in driving disease
  - Preclinical studies with nomacopan showed a dose response and combined C5 & LTB4 more effective than LTB4 alone (1)
  - Elevated C5/LTB4 in *ex-vivo* study of BP patients
- Clinical objective is steroid sparing and rapid reduction symptoms
- Ongoing Phase II study : 42 days – mild to moderate patients

Significant market with expansion potential into related conditions

1. Sezin (2019) *JCI insight 4: e*128239

Bullous Pemphigoid Initial Study Data*
First 3 Patients: Global and Blisters BPDAI



- By Day 7, 21 and day 42 of treatment, the BPDAI global score fell by a mean of 31%, 45% and 52%
- By Day 7, 21 and day 42 of treatment, blisters/erosions dropped by a mean of 45%, 75% and 87%

Daily SQ nomacopan well tolerated, no drug-related adverse events

Planned amendment to extend into severe patients

* Prior to treatment, two of three patients were on topical corticosteroids (mometasone), while the third patient was naïve to steroid treatment & received no steroid while on nomacopan. There was no planned tapering of topical steroid dose, but patients did use less than the 30g permitted to day 21. After day 21, any use of mometasone was regarded as rescue therapy, and both patients were only treated with nomacopan.

In the 7-11-day period prior to initiation on nomacopan, the two patients showed either no or minor improvement in BPDAI score (between 0 and 5%), and no improvement in blisters while being treated with topical steroids alone.

Nomacopan in Bullous Pemphigoid Differentiation Against Competitors

Clear Positioning

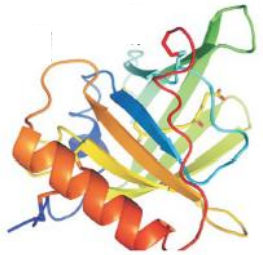
- Positioned as 2nd line treatment for patients with moderate to severe symptoms, previously treated with corticosteroids
- Better safety and side effect profile than steroids and non-specific immunosuppressants
- Used to reduce or eliminate dependency on steroids

Differentiated MOA

- Nomacopan acts upstream of competitors' targets – in particular C5 & LTB4 are part of inflammatory process leading to generation of cytokines via antibody

Next Steps

- Potential post-Phase II regulatory interaction following study completion to help define **pivotal study design**



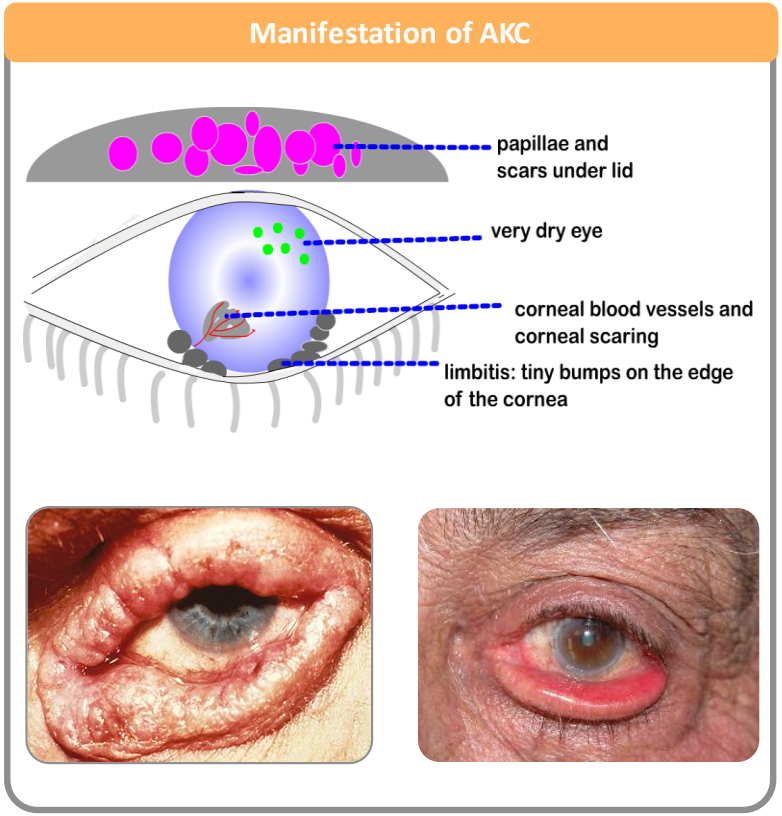
Topical Nomacopan:

Clinically Testing C5 & LTB4 Binding in

Ophthalmology / AKC

Atopic Keratoconjunctivitis (AKC): Chronic Ocular Disease Which Can Lead to Vision Loss

- Atopic Keratoconjunctivitis: severe eye surface inflammation causing infiltration of immune cells (neutrophils & T cells)



- Chronic disease can result in severe dry eye, corneal neovascularisation, & formation of sterile corneal ulcers
- If left untreated, can lead to vision loss; other complications include cataracts, keratoconus, infectious keratitis and tear dysfunction
- Topical drugs, such as steroids or cyclosporine treatments complicated by issues with comfort and side effects of chronic usage
- Orphan condition with a prevalence of 160K patients in the US and EU

Atopic Keratoconjunctivitis (AKC)

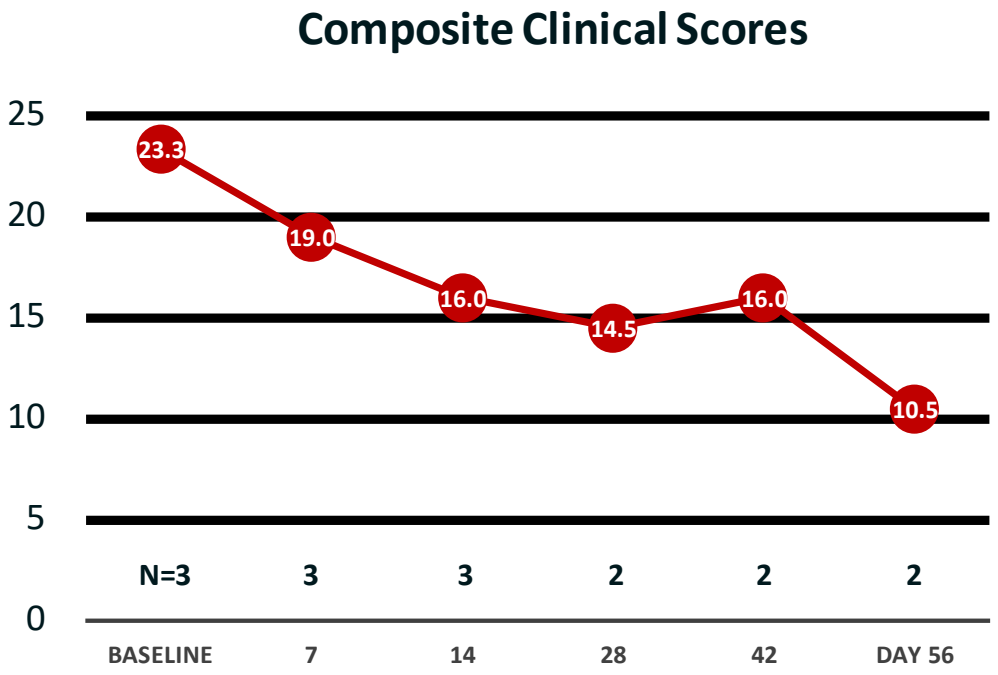
- Infiltration of neutrophils & T cells into conjunctival epithelium
- LTB4 levels – 20X in allergic conjunctivitis patients
- Complement system in tears is functionally active, and concentration of all components is greatly increased in closed-eye tears
- C5, likely generated by macrophages, may be aggressively activated by corneal disease (1)
- Preclinical model demonstrated equivalence to cyclosporine
- **Ongoing Phase I/II study.**

Potential to be first surface of eye biological treatment
Different formulation allows potential pricing & licensing flexibility
Expansion potential into other poorly-treated surface of the eye diseases

1. Bora (2008) *Semin Immunopathol 30:* 85-95

Initial AKC Clinical Data – First 2 Patients to Day 56
Rapid Improvement in Composite Clinical Scores

- Patients had deteriorating moderate – severe AKC despite ≥3 months treatment with cyclosporin t.i.d. ± steroids
- Part A of study – first in man – successfully completed – drug well tolerated
- Overall improvement across both signs and symptoms
  - Patient 1 had a disease flare at Day 42 but then recovered



- No serious drug-related AEs: drops comfortable and well tolerated

- **Next stage of trial (part B) ongoing**
  - **16 patients randomized, double masked**
  - **To enhance recruitment, trial expanded to additional clinical centers**

Nomacopan in AKC
Clear Differentiation Vs. Competitors

Clear Positioning

- Positioned as 1st line treatment for patients with moderate to severe symptoms, given **clear unmet need**

Differentiated MOA

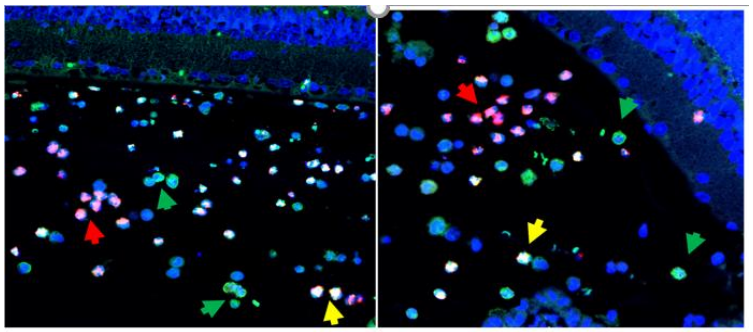
- Comfortable on application (pH neutral)
- Rapid improvement in signs & symptoms in human studies
- Upstream effects on T cells and granulocytes

Next Steps

- Potential regulatory interaction following Phase I/II study completion to help define **pivotal study design**
- Opportunity to expand into broader dry eye

C5 & LTB4 Activity in Back of the Eye Indications
Potential New Target Indication: Uveitis

LTB4 receptor
C5a receptor

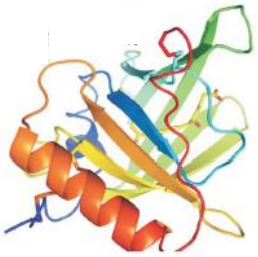


Autoimmune Uveitis (EAU) Model

- Intravitreal nomacopan reported significant improvement in uveitis model versus control
- Significant down-regulation of Th1/Th17 cells and IL-17A production

- Effect approx. equal to intravitreal dexamethasone
- C5 & LTB4 receptors both identified in retina*
- Uveitis treatment currently primarily steroids

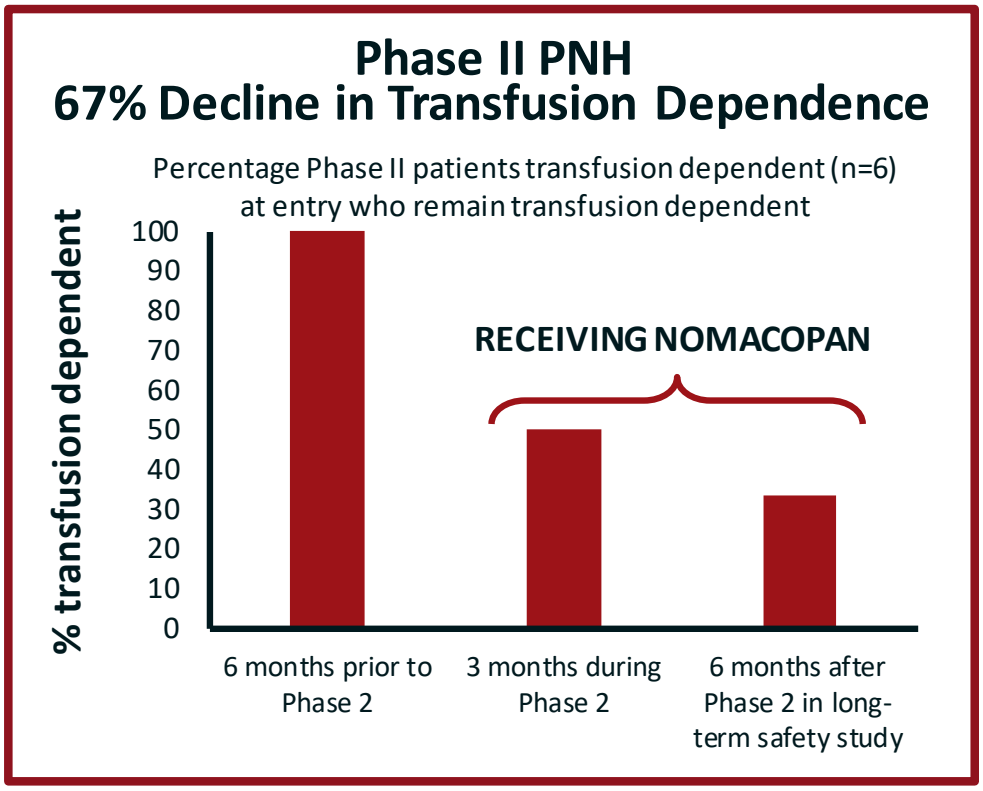
* Eskandarpour M et al. Poster 797 (B0275) ARVO 2019



Subcutaneous Nomacopan:

Paroxysmal Nocturnal Hemoglobinuria

PNH Phase II: Transfusion Reduction Validated Efficacy - Option Value For Continuing to MAA

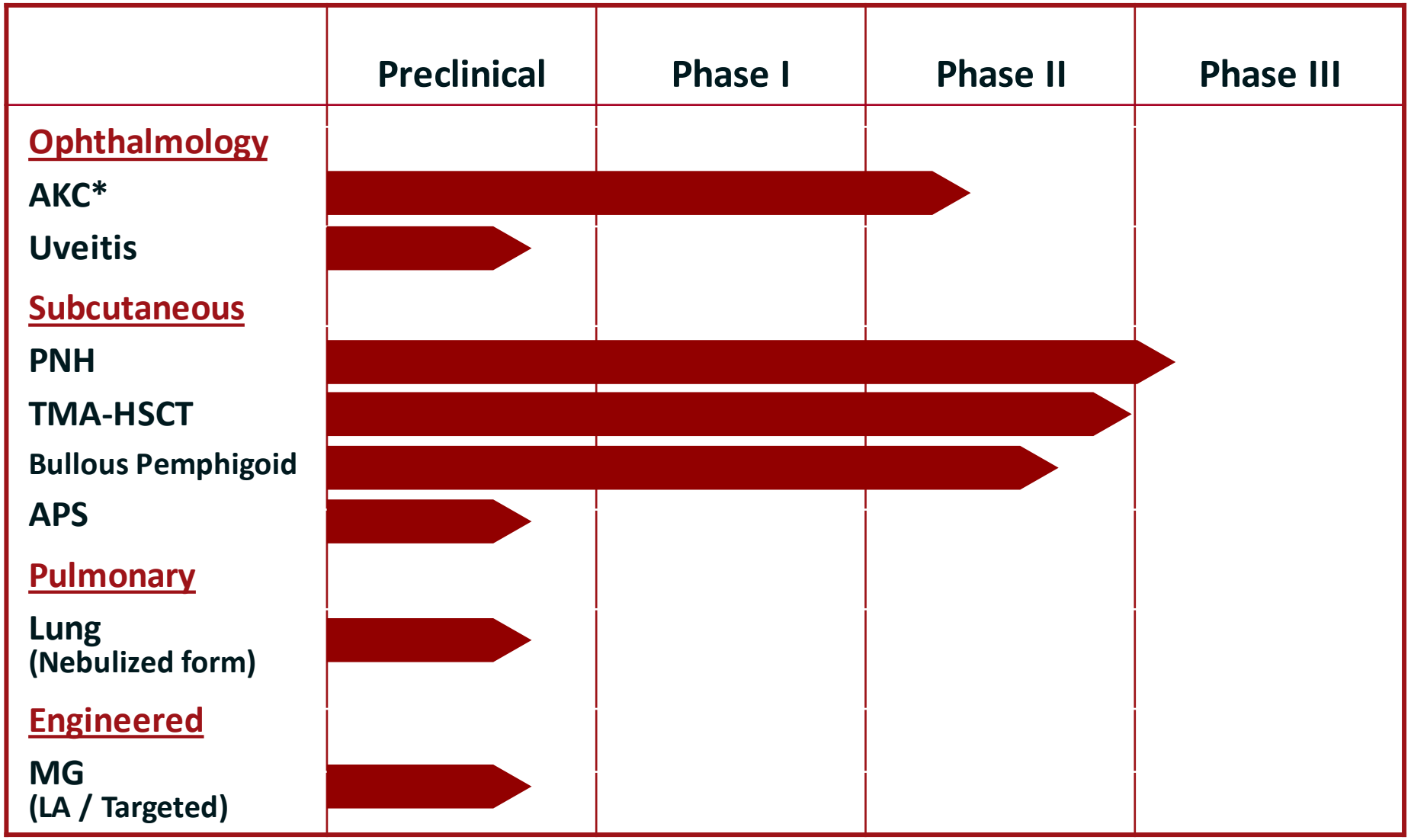


- Six patients transfused in six months prior to entering Phase II
- Three became transfusion independent during Phase II
- Four transfusion independent during follow-up long-term safety study

- Successful Phase II study achieved primary end point LDH ≤1.8X ULN
- Phase III study in naïve patients – primary end point: transfusion independence
- Potential pen injector will hold weekly dose, stable at room temperature with 0.4ml daily injection

Wrap up

Four Primary/Lead Indications in Phase II/III Plus Pipeline of Earlier-Stage Targets



* Phase I/II study

APS: Antiphospholipid Syndrome; MG: Myasthenia Gravis

Financial and Clinical Wrap up
Meaningful Upcoming Clinical Milestones

- Growing efficacy data set across four severe orphan conditions
- 20 cumulative patient-years of treatment shows positive safety data and well tolerated
- Multiple expected short- to mid-term clinical readouts:
  - **BP**: Complete mild to moderate patients Q4 2019
  - **TMA**: Q4 2019 - pivotal trial approval/open
  - **AKC:** Part B update Q4 2019
  - **PNH:** Staged readouts
- $20 million financing facility* with Aspire Capital
- ~1.6 billion ordinary shares outstanding / equivalent to 16 million ADSs

* Circa $5.5 million drawn down to date

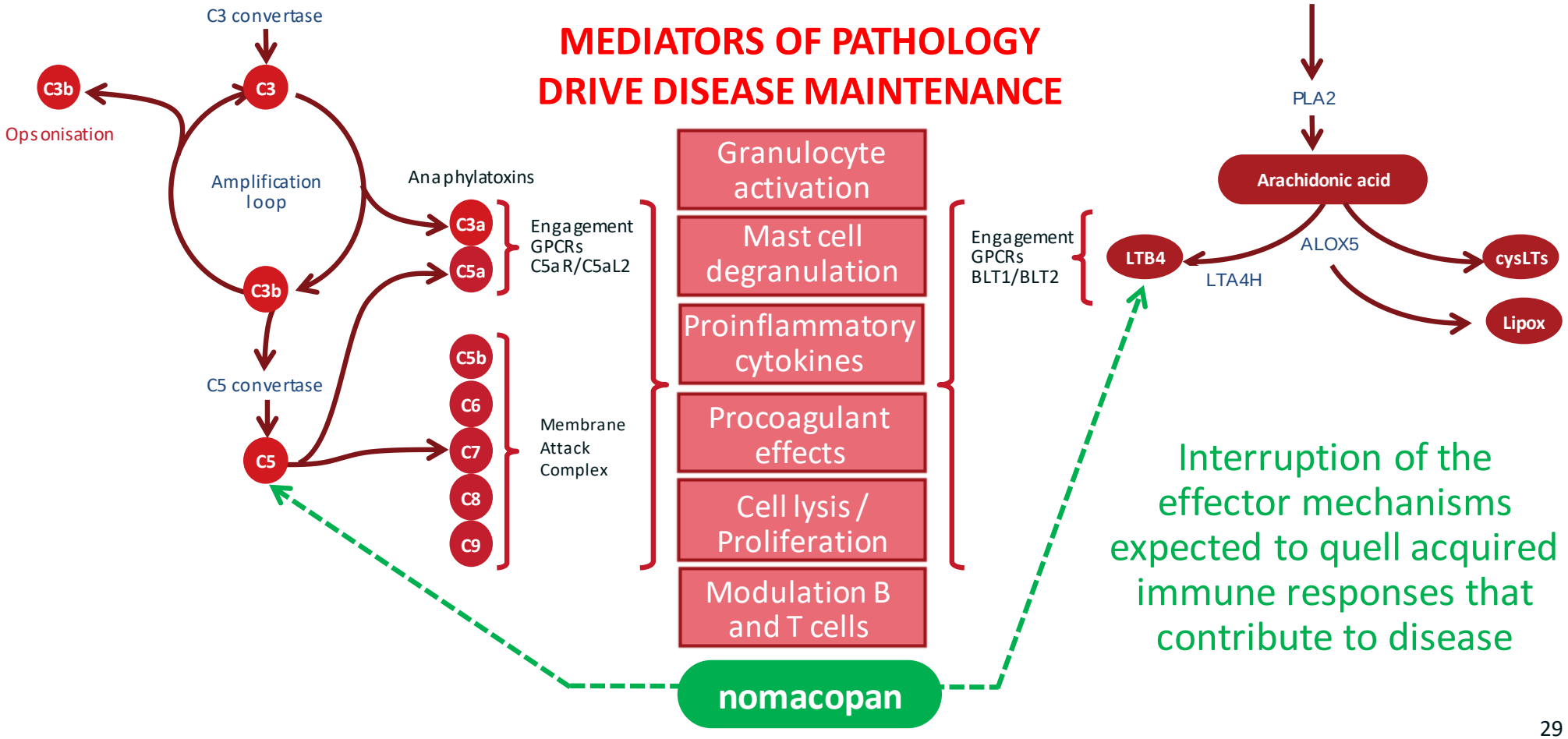
APPENDIX

Nomacopan Competitive Advantage: Synergistic C5 and LTB4 inhibition

IgG and IgM binding to C1q
Complex carbohydrates
Damage associated molecular patterns

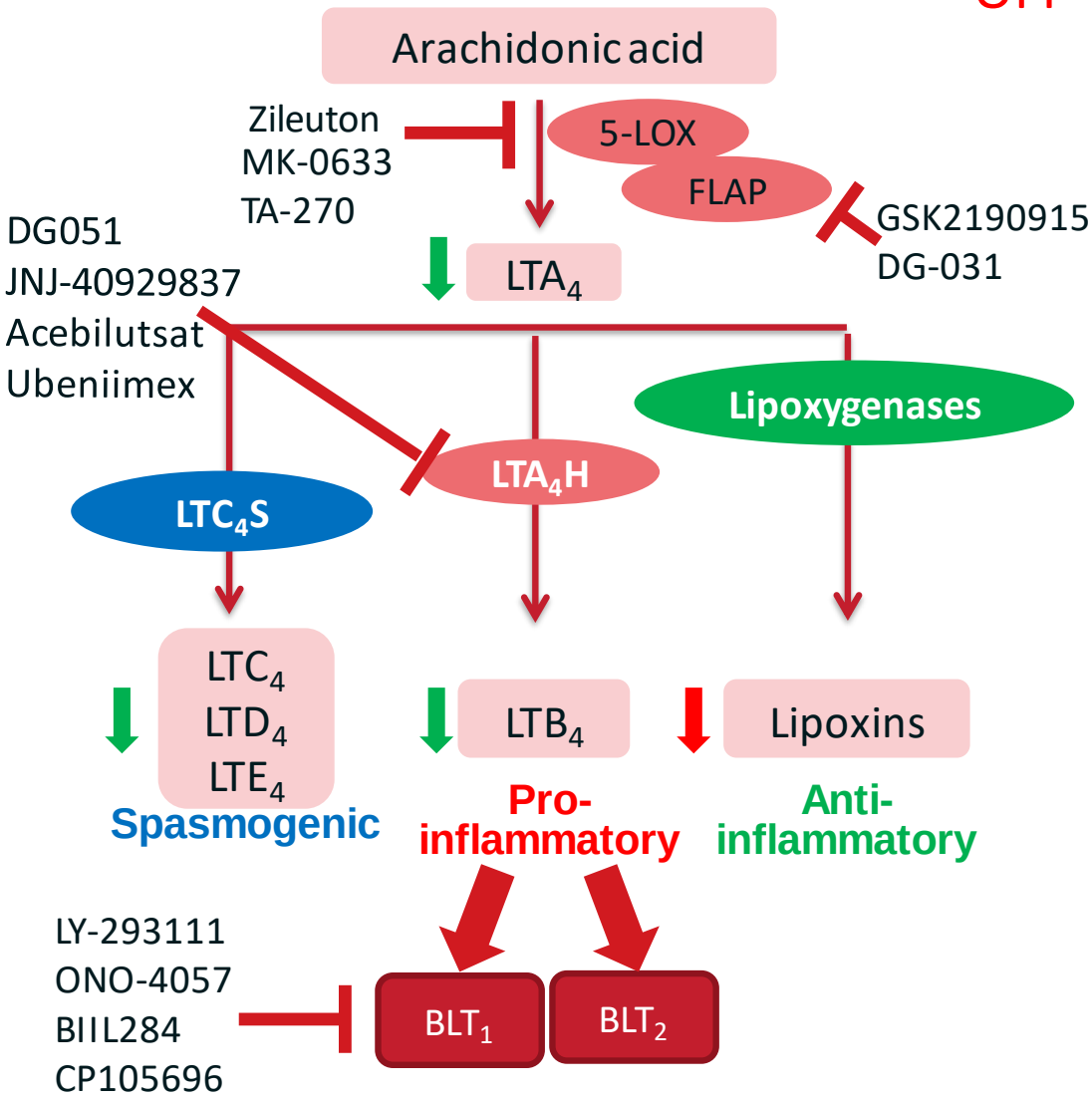
INDEPENDENT ACTIVATION PATHWAYS SIGNAL CONTINUALLY IN DEVELOPING & ESTABLISHED DISEASE

IgG binding to Fcγ receptors
Cytokines e.g. TNF and IL8
Toll-like receptors
C5a/MAC & LTB4 autoactivation



Interruption of the effector mechanisms expected to quell acquired immune responses that contribute to disease

LTB4 Ligand Capture Advantageous and Unique MOA



OFF-TARGET EFFECTS OF OTHER INHIBITORS

5-LOX / FLAP inhibitors

- Reduces anti-inflammatory lipoxins

BLT1/BLT2 antagonists

- Realization that anti-inflammatory mediators also signal through BLT1/BLT2

LTA_4H inhibitors

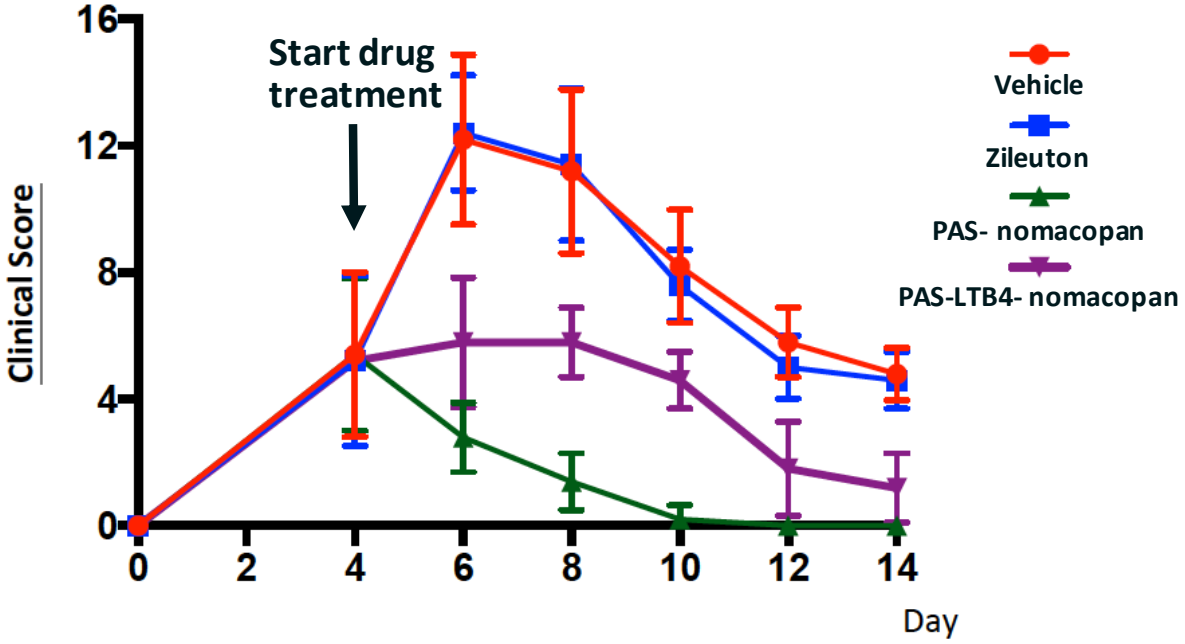
- Secondary anti-inflammatory role for LTA_4H in degrading pro-inflammatory/ remodelling mediator PGP

Expanding Pipeline Focused on Indications Where C5 and LTB4 Both Involved

Dual action C5+LTB4 (PAS-nomacopan) more effective than inhibition of LTB4 only (PAS-LTB4-nomacopan)

Dual action nomacopan more effective than C5-only nomacopan (or Zileuton®) alone

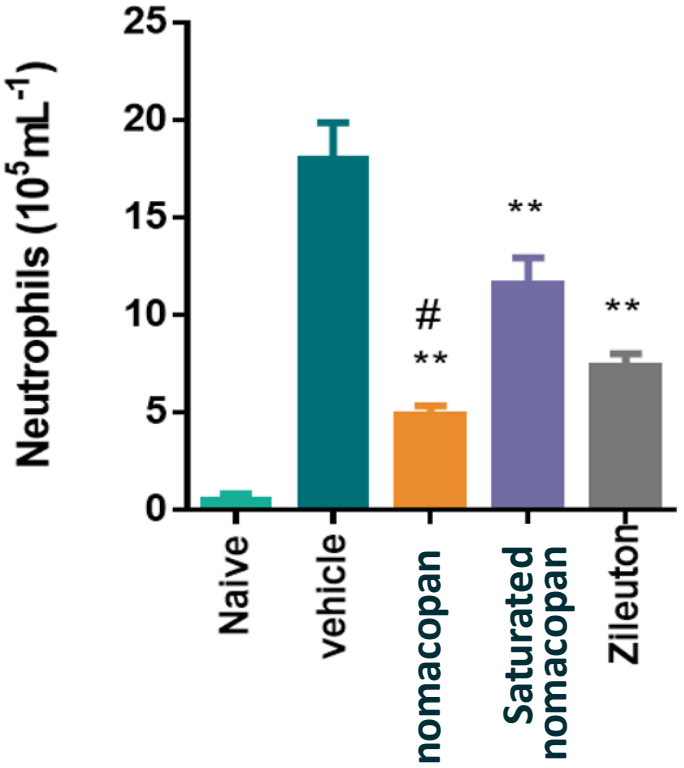
Rheumatoid Arthritis Therapeutic model



N = 5 mice in each treatment group

Work performed in group of Prof Andrew Luster, Mass Gen Hospital, Boston, USA

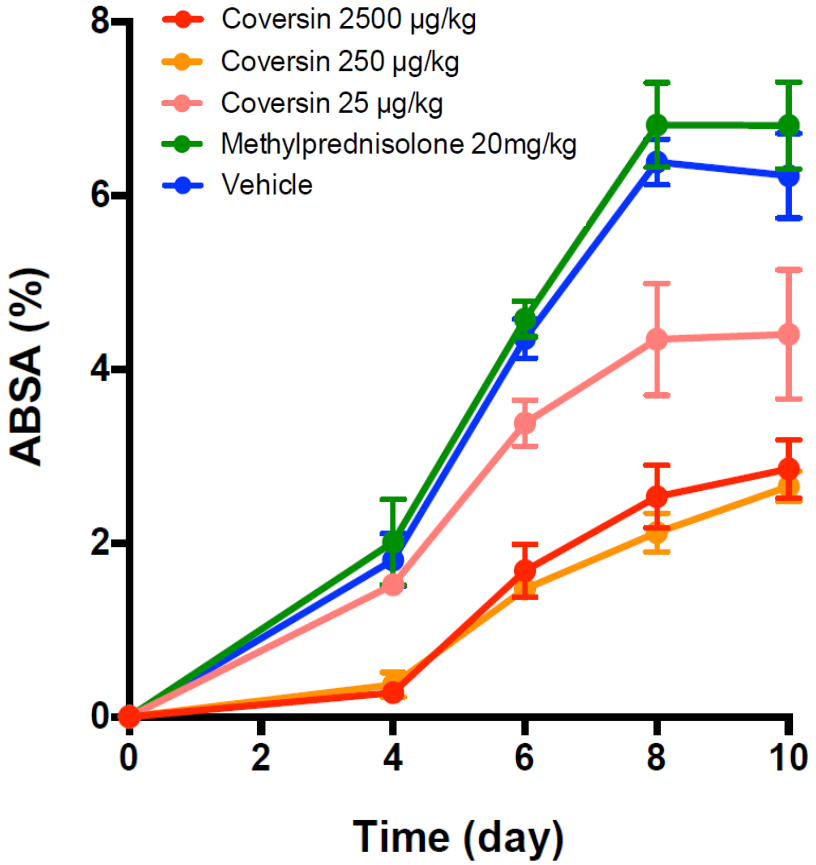
Cell recruitment to Lung Induced by LPS



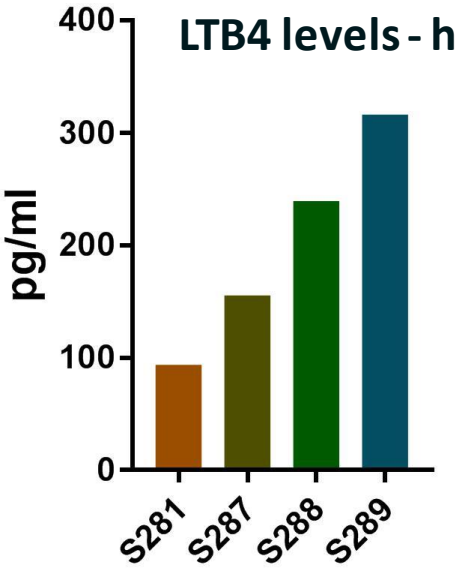
Mouse model by Pneumolabs

Epidermolysis Bullosa Acquisita Preclinical Efficacy & Elevated C5a/LTB4 in Bullous Pemphigoid patients

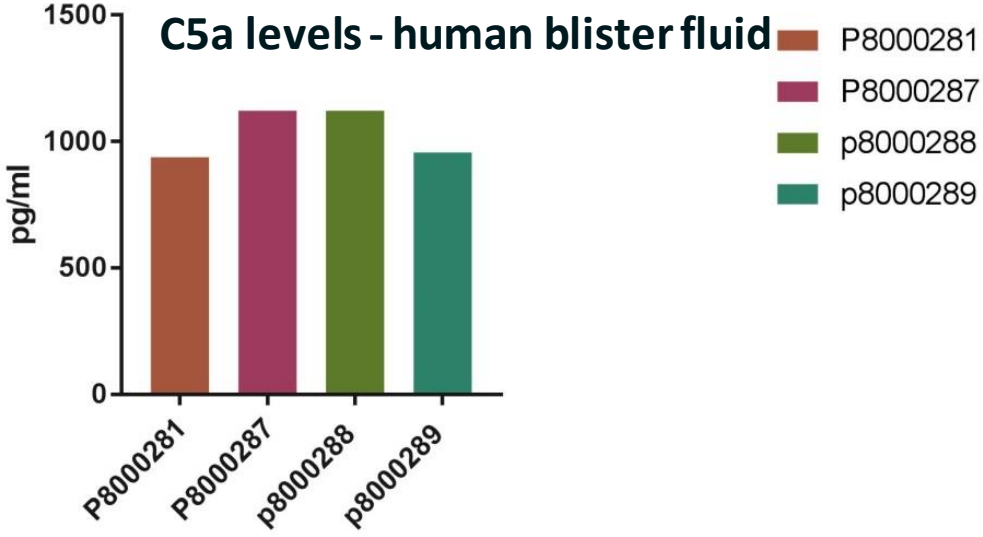
Preclinical passive mouse model of epidermolysis bullosa acquisita (EBA) from Dr. Sadik in Lubeck, Germany a leading Bullous Pemphigoid center



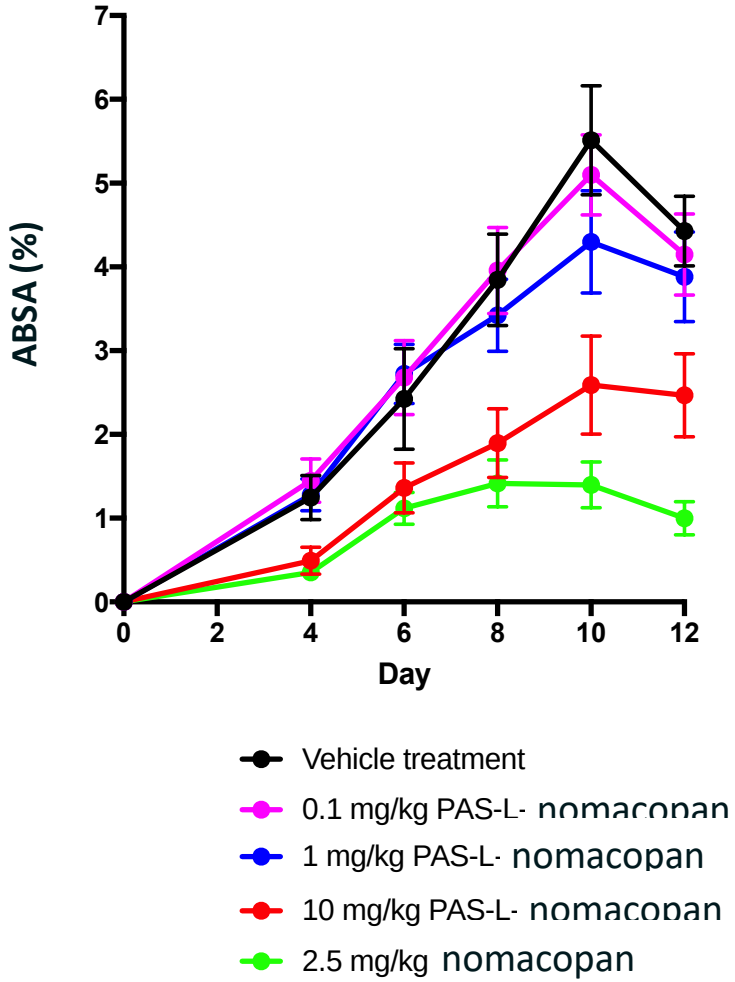
- Clear dose response
- ~60% reduction in affected area on nomacopan (SC) compared to vehicle or steroid
- P=0.0023 between vehicle and 250 µg/kg



Elevated levels of LTB4 and C5a indicate activation of complement and synthesis of LTB4 in blisters



Dual Acting Nomacopan Significantly More Effective Than Long-Acting Nomacopan Only Inhibiting LTB4



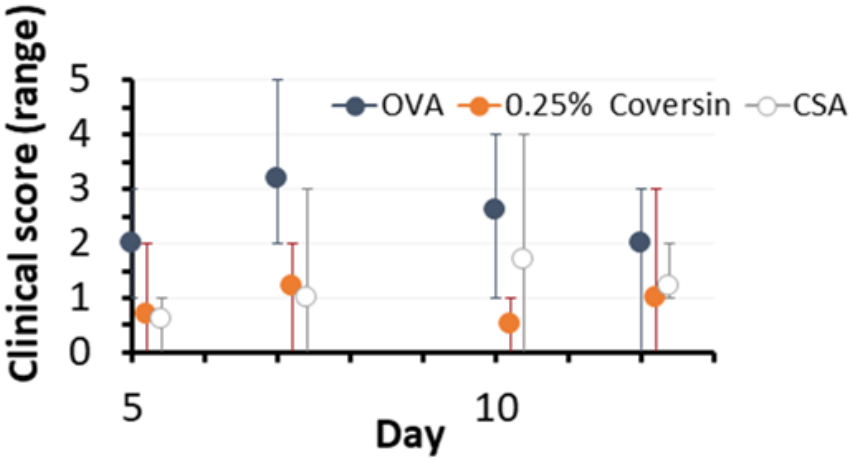
- Long acting LTB4 only nomacopan (10mg/kg) ameliorates blister formation but is less effective than the molar equivalent dose of nomacopan (2.5mg/kg)
- 1mg/kg long acting LTB4 only nomacopan ineffective whereas molar equivalent dose of 0.25mg nomacopan effective (see previous slide)

CONCLUSION:
Both C5 and LTB4 inhibition needed for full efficacy of nomacopan in EBA model

Significant Benefit Demonstrated in Late-Phase Eye Surface Inflammation

Collaboration with Institute of Ophthalmology, London, UK

EIC pre-clinical model of severe eye surface inflammation (Dr Virginia Calder)



- Treatment model of established severe eye surface inflammation
- Significant improvement in clinical scores
- Histology and cytology data support beneficial role of nomacopan
- Reduction in Th9 cells
- Reduction in IL-9

- *C57/Bl6 mice sensitized to OVA for 14 days*
- *Eye surface challenged with OVA for 12 days post sensitization (days 0 – 12)*
- *Nomacopan and cyclosporin A (CSA) applied once daily on days 4 – 12 after inflammatory response well established*
- *Maximum effect seen after 10 days of nomacopan treatment (late phase of inflammation)*

Bullous Pemphigoid Ongoing Phase II Study Patients with Mild-to-Moderate Disease

- **Trial approved Netherlands (2 sites) and Germany (6 sites)**
- **Amendment planned to treat ≤9 additional moderate-severe patients for ≤ 90 days**

Current Study design

- Phase II Single arm (n = 9); 42 days treatment
- Test role of C5 & LTB4 dual inhibition in improving BP outcomes
- Active bullous pemphigoid; newly diagnosed or recurrent

Treatment

- Nomacopan
- Day 1: 60 mg and 30 mg 12 hours later, Day 2-42: 30 mg od

Primary endpoint	Secondary endpoints
Safety	Efficacy evaluated by BPDAI (BP disease activity index) and QoL at day 42

AKC Phase I/II Proof-of-Principle Trial Design

Study design

- Phase I/II randomized, double masked, placebo-controlled, safety and efficacy study
- Part A : 3 patients open label - safety
- Part B : 16 randomized patients
- All patients on maximum cyclosporine

Treatment

- Nomacopan topical eye drops twice daily for 2 months
- Placebo eye drops

Primary endpoint	Secondary endpoints
• Safety, comfort	• Composite score of clinical signs (6) and symptoms (5)

Investor Presentation

September 2019