



Investor Presentation

October 2018

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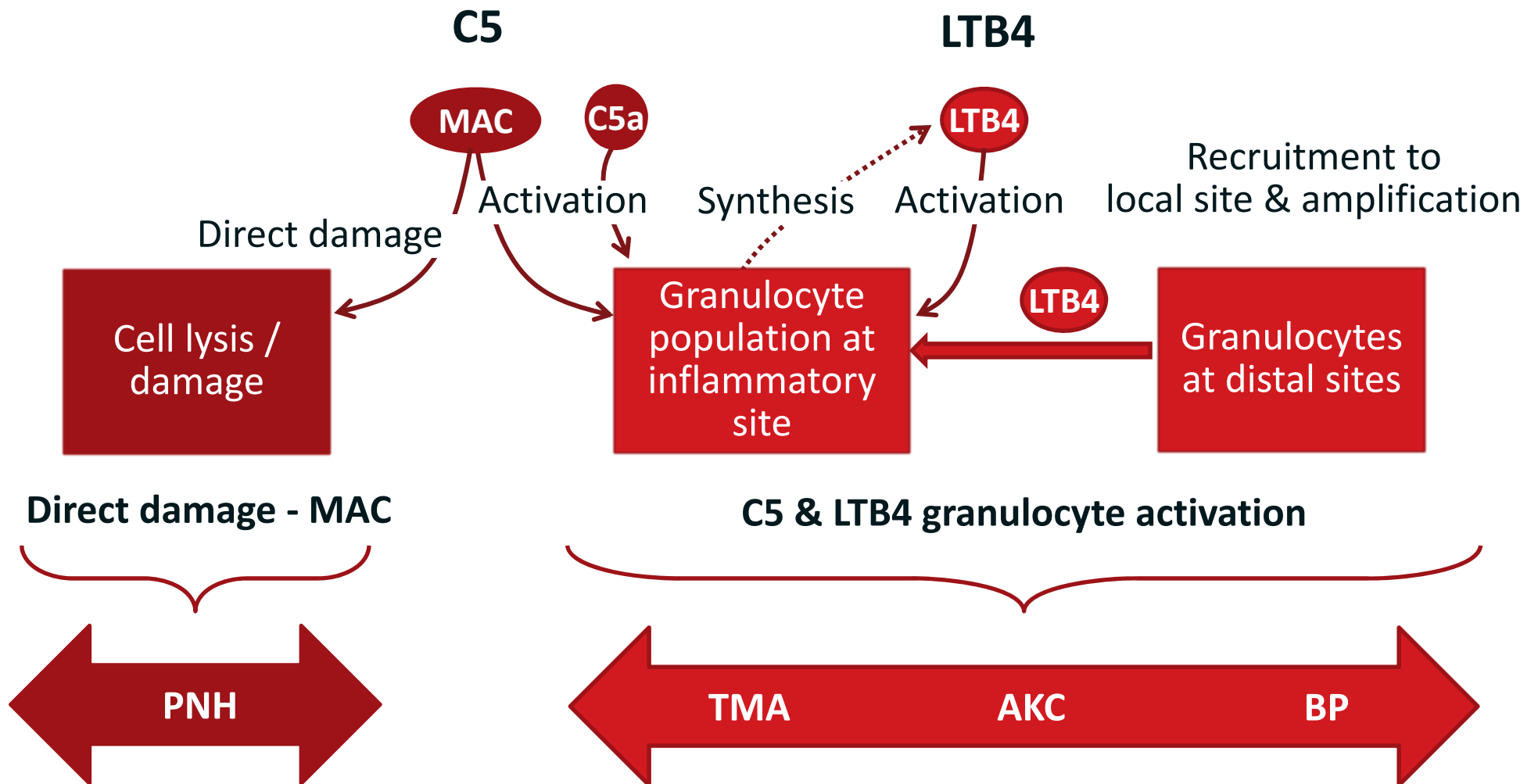
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Akari Overview

- Creating drugs for acute and chronic orphan inflammatory diseases by modulating complement/C5 and leukotriene/LTB4 pathways
- Lead drug – Coversin has unique dual MOA in both C5 & LTB4
- Diversified portfolio : four target orphan inflammatory diseases
 - Bullous pemphigoid (BP)
 - Atopic keratoconjunctivitis (AKC)
 - Severe thrombotic microangiopathy (TMA)
 - Paroxysmal nocturnal haemoglobinuria (PNH)
- Advancing clinical development
 - One Phase I/II, two phase II, and one Phase III clinical trial ongoing
 - AKC and BP initial readout in Q1 2019
- Ongoing development program prioritization
 - Potential to partner one or more active clinical programs
 - Active C5/LTB4 pipeline development for additional indications

Coversin: Synchronous C5 and LTB4 Inhibition

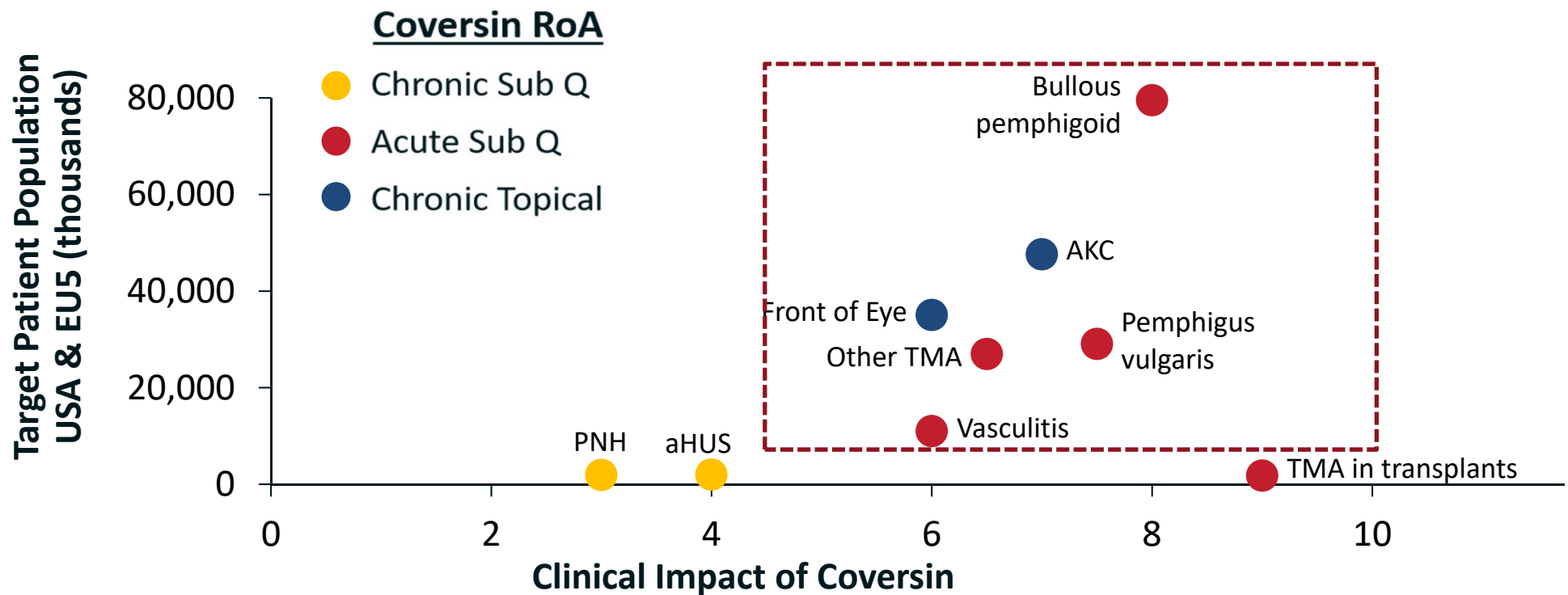
Coversin Independently Binds Both C5 & LTB4



Growing Clinical Data Set Supports Coversin Development for Range of Inflammatory Conditions

- ✓ **Positive safety profile**
 - Eleven patients on ongoing or completed treatment for between 2 and 32 months; cumulative total of over 13 patient years of treatment
 - No SAEs related to Coversin and no neutralizing antibodies
- ✓ **Proven C5 binding across disease categories**
 - Met PNH Phase II primary efficacy – reduction in LDH $\leq 1.8X$ ULN at day 28
 - CH50 below limit of quantification (from day 1) in PNH and TMA patients
 - Resolution of a wide range of clinical markers in TMA patients
- ✓ **Proven LTB4 binding in wide range of models**
 - Inhibits LTB4 induced human neutrophil migration
- ✓ **Patients successfully self administering by daily subcutaneous injection**

Focused on Orphan Inflammatory Indications with High Unmet Need



- Four lead programs : Pemphigoids (incl BP), Surface of the eye (incl AKC), TMAs, and PNH all with peak sale potentials of \$500m+
- Most disease target conditions have no approved treatment
- Smaller diseases such as HSCT – TMA provide potential gateway into other TMAs

Source: CSM Consulting, LEK

Portfolio Builds on Coversin PNH Clinical Results

Focus on Maximizing Return on Capital

	Phase I	Phase II	Phase III
PNH ¹	▶		
TMA-HSCT ²	▶ Named Patients		
TMA-aHUS ³	▶		
BP ^{4*}	▶		
AKC ^{5*}	▶		

- Initial PNH Phase II and recent TMA patient data provided clinical, safety and dosing validation for Coversin
- \$20m equity facility from Aspire focused on extending Akari's financial runway through completion of ongoing Phase II trials in TMA, BP, and AKC
- Exploring external partnering program in parallel to internal funding

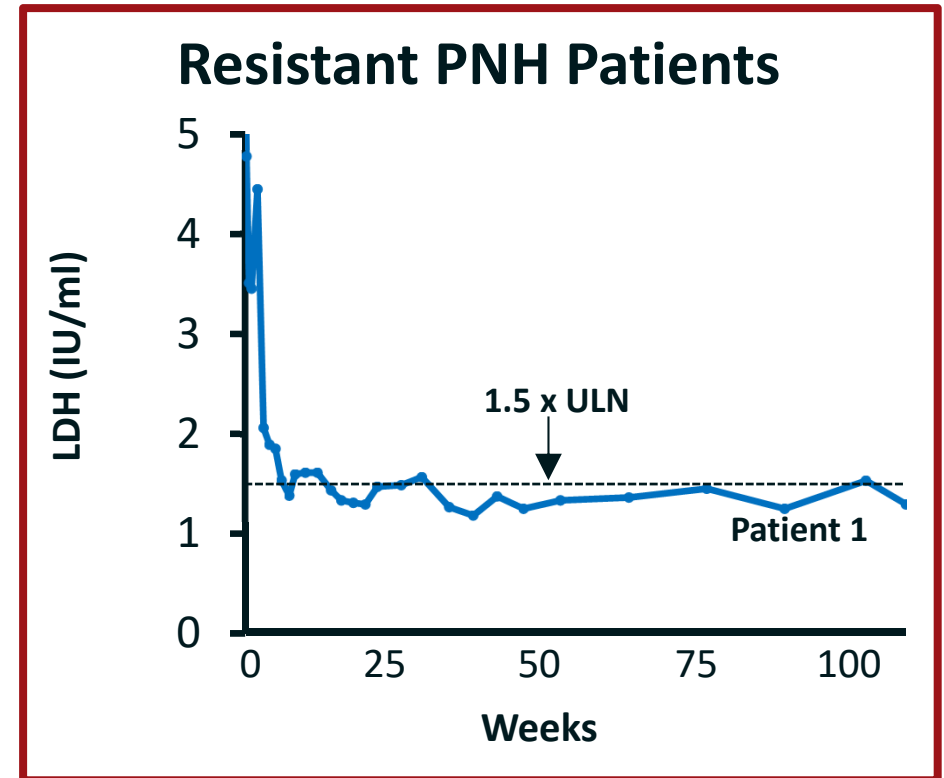
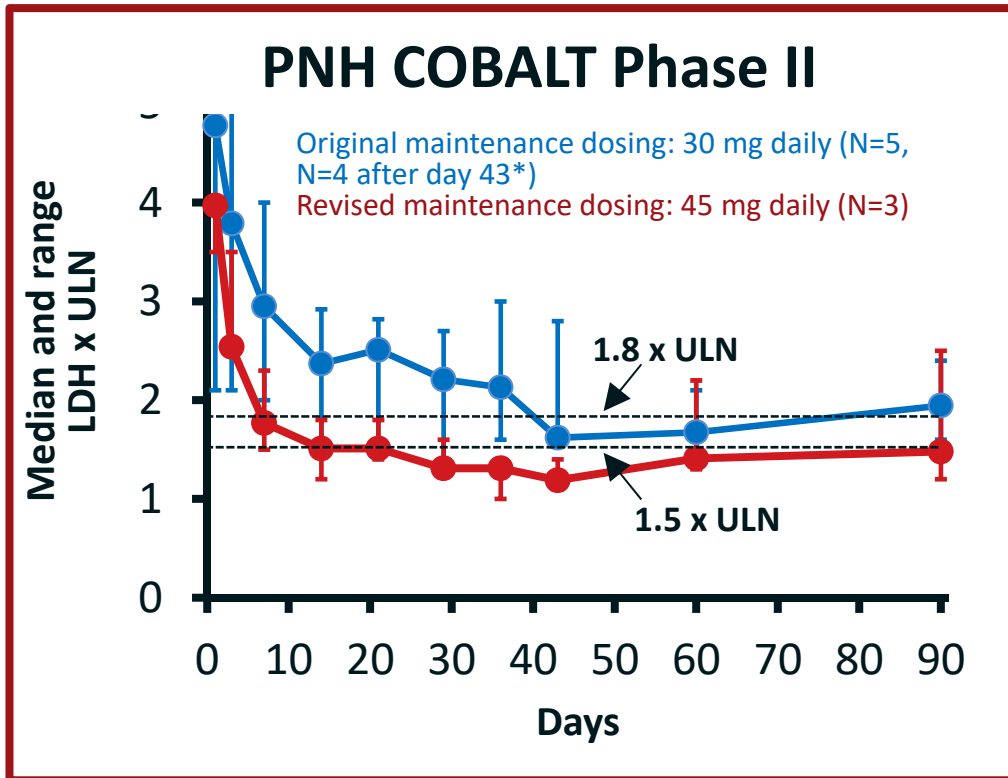
1. PNH: Paroxysmal nocturnal hemoglobinuria; 2. TMA: Thrombotic Microangiopathy; HSCT Hematopoietic stem cell transplant
 3 aHUS: Atypical hemolytic-uremic syndrome; 4. BP: Bullous Pemphigoid; 5. AKC: Atopic keratoconjunctivitis; * Dual action MOA: C5 & LTB4

Clinical Data

Patients Being Treated With Coversin Across Four Primary/Lead Indications

Trial Enrollment Completed		
PNH Resistant (chronic) Phase II 4 and 32 Months	2 patients	Ongoing
PNH naïve (chronic) Phase II and long-term safety 11-21 Months	6 patients	Ongoing
18 Months	1 patient	Complete
TMA (HSCT) Named Patients 2 Months	2 patients	Complete
Enrollment Ongoing (target patient numbers and trial duration)		
Bullous Pemphigoid Phase II 3 Months	9+9 patients	Recruiting
AKC Phase II 2 Months	11 patients	Recruiting
TMA-aHUS Phase II 6 Months	5 patients	Recruiting
PNH Phase III (Naive only) 9 Months	30 patients	Recruiting

C5 Inhibition Demonstrated in PNH patients treated with Coversin



- Naïve patients treated for 90 days
- All completing patients entered long term safety study
- No drug related SAE

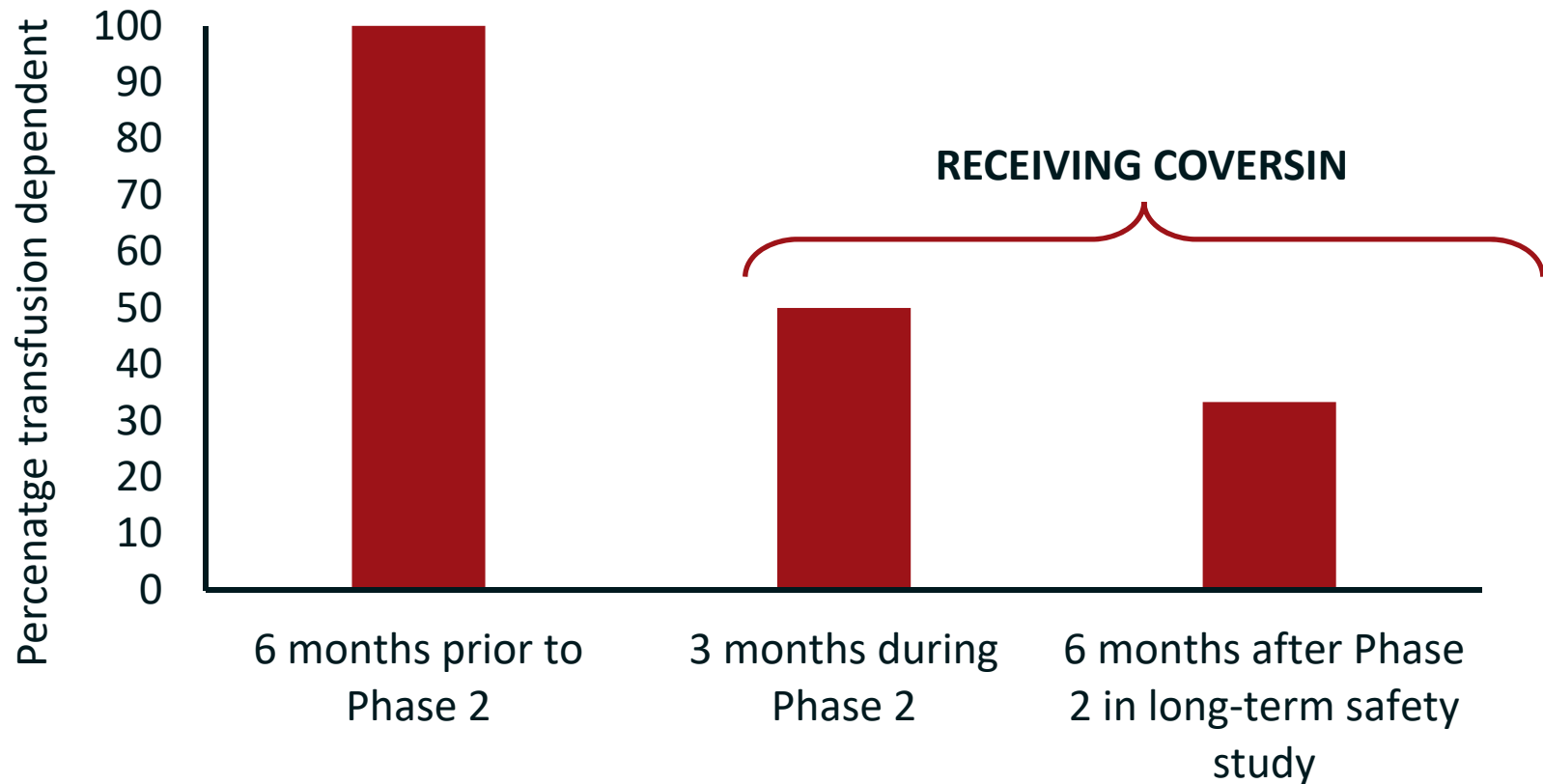
- Two patients with C5 polymorphisms resulting in treatment resistance to Eculizumab
- First patient treated for 32 months (graph above)
- Second patient treated for 4 months – LDH 1.4X ULN at day 90

Note : LDH x ULN Day 28 : 1.4, 2.2, 2.3, 1.3, 1.4, 2.7, 1.6, 1.3 :Day 60 : 1.5, 2.1, 1.8, 2.2, 1.5, 1.4, 1.3
Day 90 : 1.6, 2.4, 2.0, 2.5, 1.9, 1.5, 1.2 - *Patient withdrawn with suspected co-morbidity unrelated to treatment

Phase II PNH Clinical Endpoint

67% Decline in Transfusion Dependence

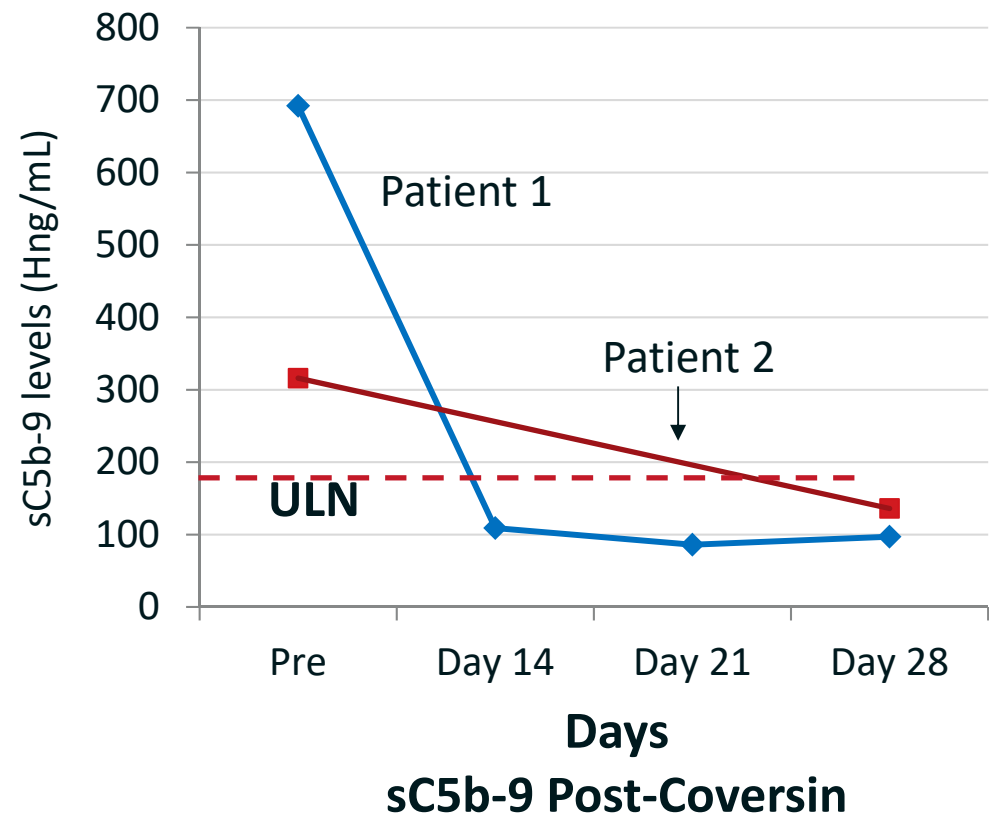
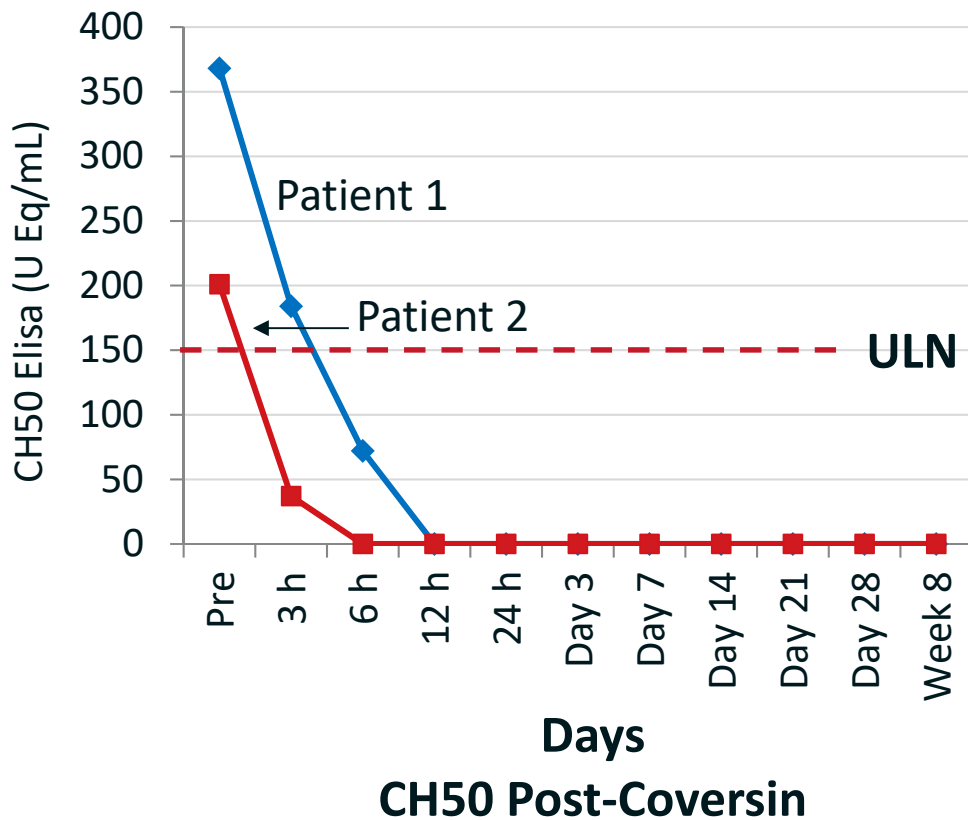
Percentage Phase II patients transfusion dependent (n = 6)
at entry who remain transfusion dependent



- 6 patients transfused in 6 months prior to entering Phase II
- 3 became transfusion independent during Phase II
- 4 transfusion independent during long-term safety study

Pediatric TMA: Post-Bone Marrow Transplant Complement Inhibition demonstrated with Coversin

Complete and rapid C5 inhibition as seen in PNH patients
Pediatric dosing regime effective



Note : data up to day 28 for Patient 2

Coversin Activity Demonstrated in HSCT–TMA Across a Range of Outcome Measures (2 Patients)

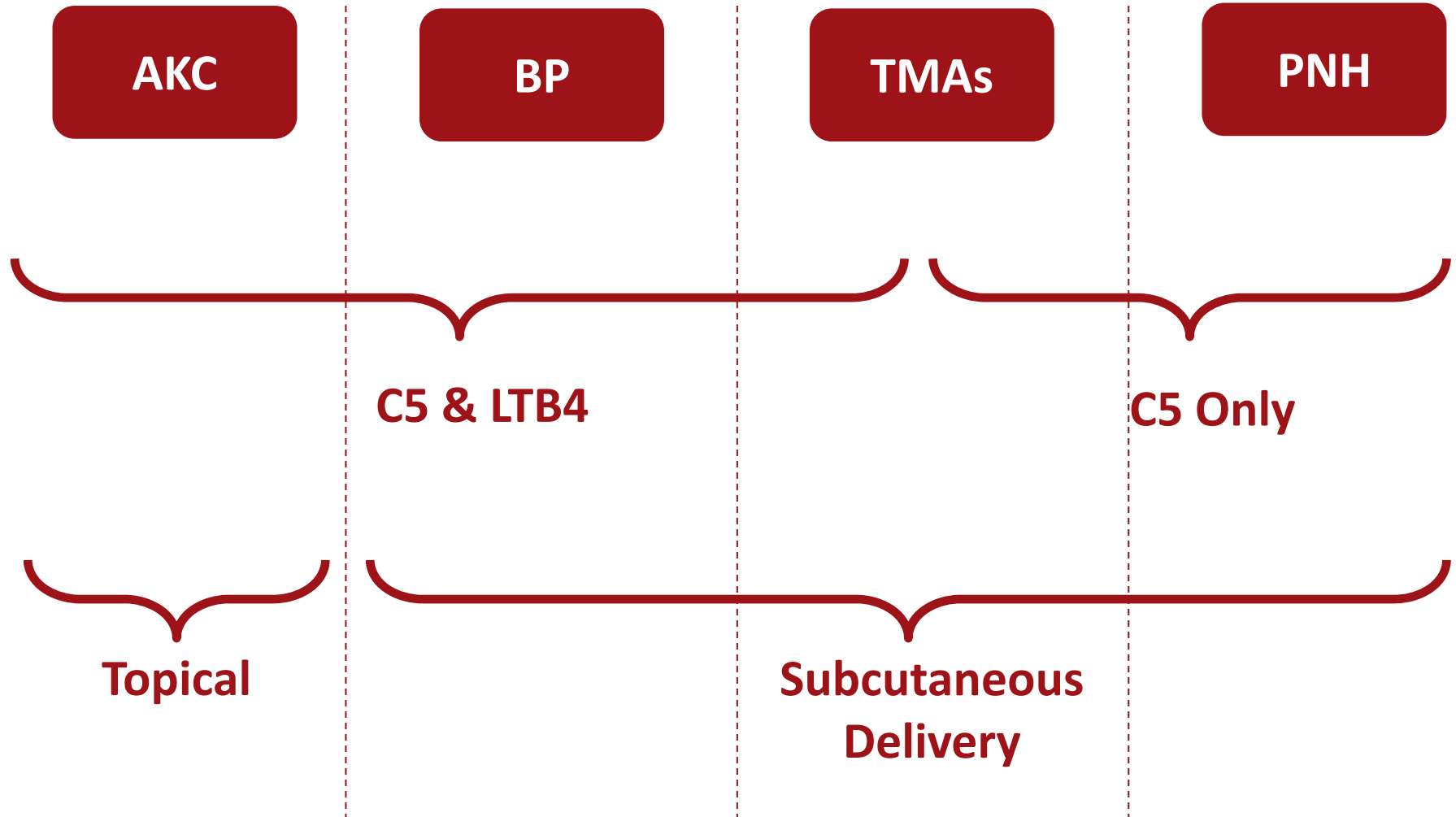
TMA Marker	Patient	Baseline	Day 7	Day 14	Day 28	Day 60
Hemolytic anaemia	1	Yes	→			Resolved
	2	No				
Red blood cell fragments	1	Yes	→			Resolved
	2	Yes	→		Resolved	
Thrombocytopenia	1	Yes	→			Resolved
	2	Yes	→	Resolved		
Increased LDH	1	Yes	→		Resolved	
	2	Yes	→	Resolved		
Proteinuria and/or increased 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 Coversin 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 Coversin. Note - data for patient 2 is up to day 28

Clinical Programs

Clinical Programs focused on C5 & C5/LTB4 Targets And Both SQ and Topical Delivery



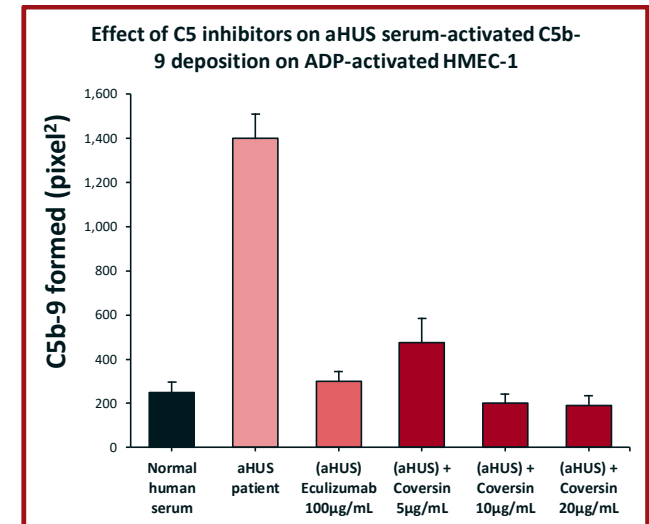
AKC: Atopic keratoconjunctivitis; BP: Bullous Pemphigoid; TMA: Thrombotic Microangiopathy;
PNH: Paroxysmal nocturnal hemoglobinuria

Thrombotic Microangiopathies (TMA)

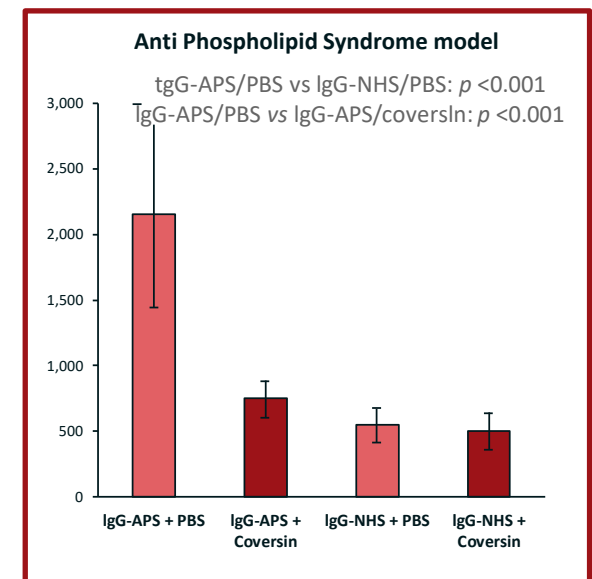
Several Diseases With Unmet Need

- Thrombotic Microangiopathies
 - Range of orphan disease, in most cases no approved therapy
 - Numerous conditions considered complement driven with LTB4 implicated in several
- Atypical hemolytic-uremic syndrome (aHUS)
 - Ongoing Phase II program in naïve patients
- Hematopoietic stem cell transplant (HSCT TMA)
 - Named patient program; 2 patients to date
 - Gateway condition
- Other TMAs
 - Preclinical data in anti-phospholipid syndrome
 - Potential for treatment with Coversin

aHUS – *ex vivo* endothelial cell surface model



Antiphospholipid model - area of induced thrombi



Thrombotic Microangiopathy (TMA) After Transplant: Significant Unmet Need

- Orphan condition in which there is evidence that both terminal complement activation and possibly LTB4 have a role in driving disease
- Complications following bone marrow transplantation (BMT) in up to 30% of patients
- In severe cases, mortality in excess of 80%
- Use of eculizumab in TMA, post bone marrow transplant appears to offer better outcomes compared to SoC
- Coversin used on a named patient basis for Pediatric HSCT-TMA

TMA offers opportunity to expand into related conditions



Bullous Pemphigoid (BP)

- Orphan condition in which there is evidence that both terminal complement activation (C5) and LTB4 have a role in driving disease
- TPP – moderate to severe patients, newly presenting or relapsing
- Other biologicals with different MOA are also in Phase II development for BP (e.g. – Taltz[®] (anti-IL17a) and Bertilimumab[®] (anti-eotaxin))
- Focus of biologicals is steroid sparing and rapid reduction symptoms
- Elevated C5/LTB4 in ex-vivo study of BP patients

BP offers opportunity to expand into related conditions



Bullous Pemphigoid Phase II Trial Design in Patients with Mild-to-Moderate Disease

- Trial approved Netherlands (2 sites) and expected soon in Germany (6 sites)
- Amendment planned to increase trial size with 9 additional moderate-severe patients

Study design

- Phase II Open label single arm (n = 9); 42 days treatment
- Test role of C5 & LTB4 dual inhibition in improving BP outcomes
- Active; newly diagnosed or recurrent, mild to moderate treated with topical mometasone

Treatment

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

Primary endpoint

Safety

Secondary endpoints

Efficacy evaluated by BPDAI (BP disease activity index) and QoL at day 42

Atopic Keratoconjunctivitis (AKC)

- Severe eye surface inflammation causing infiltration of immune cells (neutrophils & T cells) – a major cause of blindness
- Topical drugs, such as steroids or cyclosporin, often not effective or cannot be given chronically
- Progresses to affect cornea; may lead to vision loss
- Both complement and LTB4 known to be involved
- Preclinical model demonstrated greater inflammatory reduction than typically achieved by cyclosporin

AKC offers opportunity to expand into related conditions



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

Study design

- Phase I/II randomized, double blind, placebo-controlled, safety and dose finding study – 11 active 8 placebo

Treatment

- Coversin topical eye drops three times daily for 2 months
 - High (0.25%), medium (0.125%) and low (0.063%) doses
- Placebo eye drops

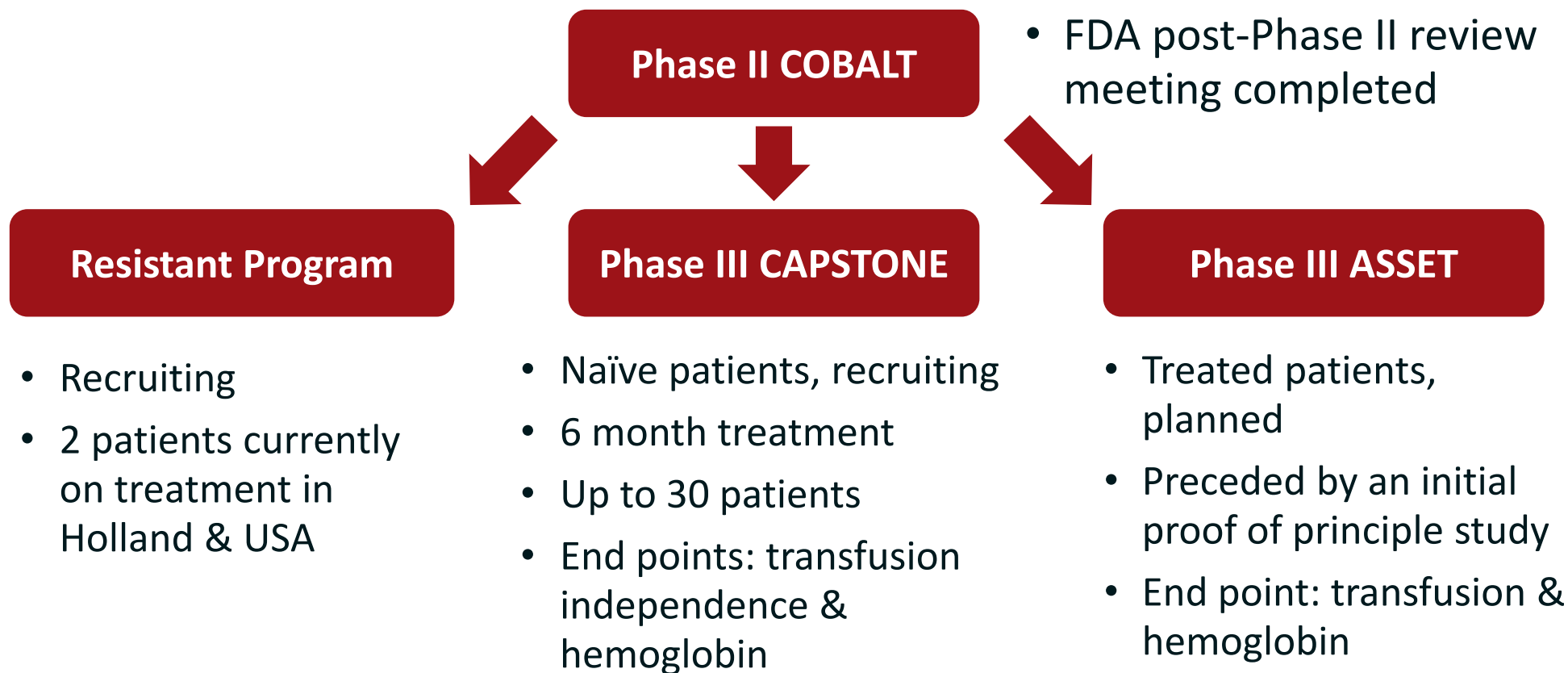
Primary endpoint

- Safety, comfort

Secondary endpoints

- Clinical signs and symptoms
- Reduction in inflammatory markers (MMP-9)
- Cytology

PNH Clinical Programs: Staged Phase III Commitment Focus on PNH Patient Convenience



Development program allows ongoing data readouts and staged allocation of resources

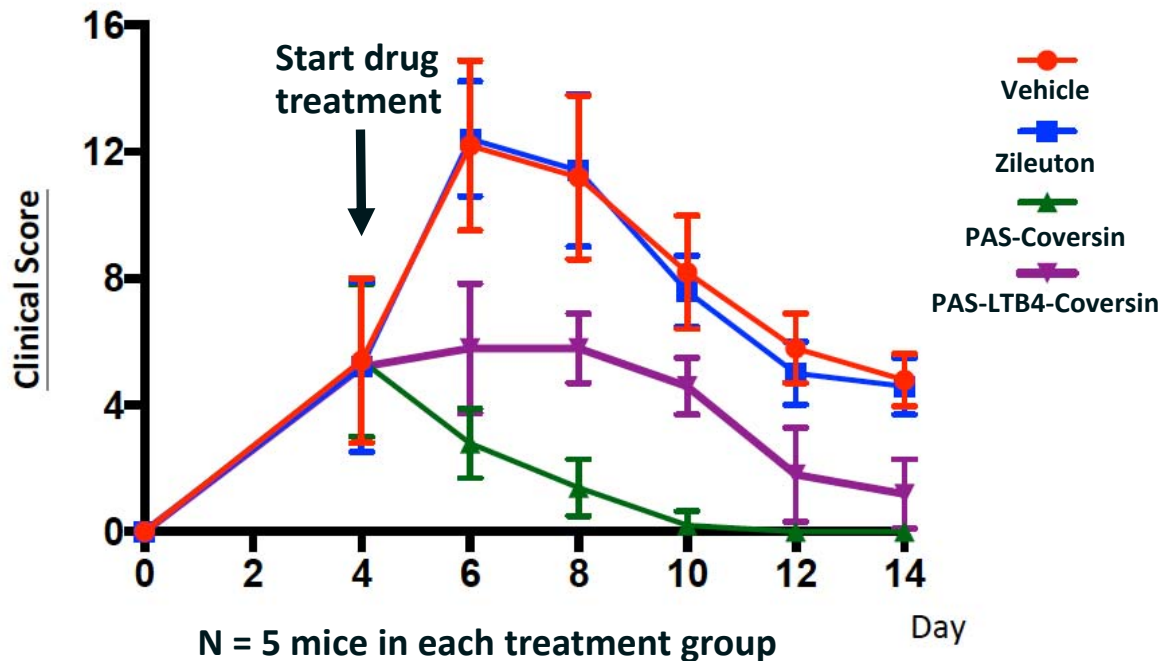
**Focus on patient convenience : SQ, highly soluble, stable at room temp,
New formulation in development for Auto injector pen with 1 week dosing**

Expanding Pipeline Focused on Indications Where C5 and LTB4 Both Involved

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

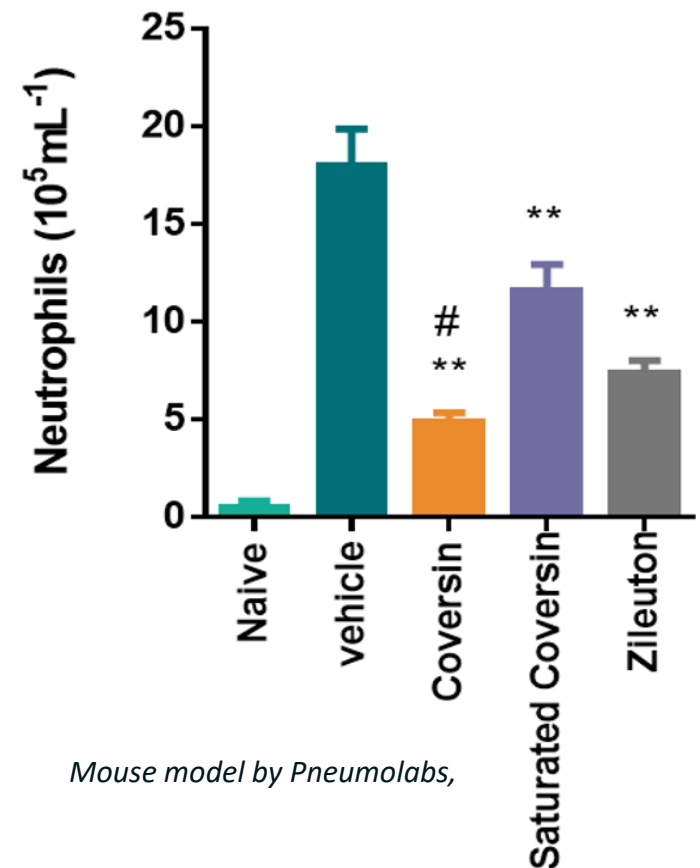
Dual action Coversin more effective than C5-only Coversin (or Zileuton®) alone

Rheumatoid Arthritis Therapeutic model



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

Cell recruitment to Lung Induced by LPS



Mouse model by Pneumolabs,

Akari Summary

- Unique mode of action - inhibiting both C5 and LTB4
- Diversified pipeline :
 - AKC and BP - initial readouts Q1 2019
 - TMAs - ongoing readouts in aHUS and TMA post HSCT – H1 2019
 - PNH - staged readouts in 2019
 - Preclinical - ongoing programs in lung, RA, trauma
- \$20 million financing facility with Aspire Capital
- \$15 million in cash at June 30, 2018



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