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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

**C5**
- MAC Activation
- C5a Activation
- Direct damage - MAC
- Cell lysis / damage
- Granulocyte population at inflammatory site

**LTB4**
- Synthesis
- Activation
- Recruitment to local site & amplification
- Granulocytes at distal sites
- Direct damage

C5 & LTB4 granulocyte activation

PNH

TMA  AKC  BP
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 ≤ 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

<table>
<thead>
<tr>
<th></th>
<th>Phase I</th>
<th>Phase II</th>
<th>Phase III</th>
</tr>
</thead>
<tbody>
<tr>
<td>PNH&lt;sup&gt;1&lt;/sup&gt;</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>TMA-HSCT&lt;sup&gt;2&lt;/sup&gt;</td>
<td></td>
<td><strong>Named Patients</strong></td>
<td></td>
</tr>
<tr>
<td>TMA-aHUS&lt;sup&gt;3&lt;/sup&gt;</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>BP&lt;sup&gt;4*&lt;/sup&gt;</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>AKC&lt;sup&gt;5*&lt;/sup&gt;</td>
<td></td>
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</tbody>
</table>

- 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

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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

<table>
<thead>
<tr>
<th>Trial Enrollment Completed</th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>PNH Resistant (chronic) Phase II</td>
<td></td>
</tr>
<tr>
<td>4 and 32 Months</td>
<td>2 patients</td>
</tr>
<tr>
<td>PNH naïve (chronic) Phase II and long-term safety</td>
<td></td>
</tr>
<tr>
<td>11-21 Months</td>
<td>6 patients</td>
</tr>
<tr>
<td>18 Months</td>
<td>1 patient</td>
</tr>
<tr>
<td>TMA (HSCT) Named Patients</td>
<td></td>
</tr>
<tr>
<td>2 Months</td>
<td>2 patients</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Enrollment Ongoing (target patient numbers and trial duration)</th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>Bullous Pemphigoid Phase II</td>
<td></td>
</tr>
<tr>
<td>3 Months</td>
<td>9+9 patients</td>
</tr>
<tr>
<td>AKC Phase II</td>
<td></td>
</tr>
<tr>
<td>2 Months</td>
<td>11 patients</td>
</tr>
<tr>
<td>TMA-aHUS Phase II</td>
<td></td>
</tr>
<tr>
<td>6 Months</td>
<td>5 patients</td>
</tr>
<tr>
<td>PNH Phase III (Naive only)</td>
<td></td>
</tr>
<tr>
<td>9 Months</td>
<td>30 patients</td>
</tr>
</tbody>
</table>
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

- 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)

<table>
<thead>
<tr>
<th>TMA Marker</th>
<th>Patient</th>
<th>Baseline</th>
<th>Day 7</th>
<th>Day 14</th>
<th>Day 28</th>
<th>Day 60</th>
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<tbody>
<tr>
<td>Hemolytic anaemia</td>
<td>1</td>
<td>Yes</td>
<td></td>
<td></td>
<td></td>
<td>Resolved</td>
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<tr>
<td></td>
<td>2</td>
<td>No</td>
<td></td>
<td></td>
<td></td>
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<tr>
<td>Red blood cell fragments</td>
<td>1</td>
<td>Yes</td>
<td></td>
<td></td>
<td></td>
<td>Resolved</td>
</tr>
<tr>
<td></td>
<td>2</td>
<td>Yes</td>
<td></td>
<td></td>
<td></td>
<td>Resolved</td>
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<tr>
<td>Thrombocytopenia</td>
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<td>Yes</td>
<td></td>
<td></td>
<td></td>
<td>Resolved</td>
</tr>
<tr>
<td></td>
<td>2</td>
<td>Yes</td>
<td></td>
<td></td>
<td></td>
<td>Resolved</td>
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<tr>
<td>Increased LDH</td>
<td>1</td>
<td>Yes</td>
<td></td>
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<td></td>
<td>Resolved</td>
</tr>
<tr>
<td></td>
<td>2</td>
<td>Yes</td>
<td></td>
<td></td>
<td></td>
<td>Resolved</td>
</tr>
<tr>
<td>Proteinuria and/or increased creatinine</td>
<td>1</td>
<td>Yes</td>
<td></td>
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<td></td>
<td>Resolved</td>
</tr>
<tr>
<td></td>
<td>2</td>
<td>Yes</td>
<td></td>
<td></td>
<td></td>
<td>N/A</td>
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<tr>
<td>Hypertension</td>
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<td>Yes</td>
<td></td>
<td></td>
<td></td>
<td>Resolved</td>
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<td></td>
<td>2</td>
<td>Yes</td>
<td></td>
<td></td>
<td></td>
<td>Resolved</td>
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<tr>
<td>Neurology</td>
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<td>Yes</td>
<td></td>
<td></td>
<td></td>
<td>Resolved</td>
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<tr>
<td></td>
<td>2</td>
<td>No</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>GI bleed</td>
<td>1</td>
<td>No</td>
<td></td>
<td></td>
<td></td>
<td>Resolved</td>
</tr>
<tr>
<td></td>
<td>2</td>
<td>Yes</td>
<td></td>
<td></td>
<td></td>
<td>Resolved</td>
</tr>
</tbody>
</table>

**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

**Antiphospholipid syndrome model - area of induced thrombi**

- tgG-APS/PBS vs IgG-NHS/PBS: p < 0.001
- IgG-APS/PBS vs IgG-APS/Coversin: p < 0.001

**Effect of C5 inhibitors on aHUS serum-activated C5b-9 deposition on ADP-activated HMEC-1**
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

HSCT-TMA → aHUS → TMA+SLE → Anti-phospholipid syndrome → Vasculitis

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 Bertillumab® (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

<table>
<thead>
<tr>
<th>Primary endpoint</th>
<th>Secondary endpoints</th>
</tr>
</thead>
<tbody>
<tr>
<td>Safety</td>
<td>Efficacy evaluated by BPDAI (BP disease activity index) and QoL at day 42</td>
</tr>
</tbody>
</table>
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

AKC → VKC, PKC, SAC, SJS, MMP
### 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

<table>
<thead>
<tr>
<th>Primary endpoint</th>
<th>Secondary endpoints</th>
</tr>
</thead>
<tbody>
<tr>
<td>Safety, comfort</td>
<td>Clinical signs and symptoms</td>
</tr>
<tr>
<td></td>
<td>Reduction in inflammatory markers (MMP-9)</td>
</tr>
<tr>
<td></td>
<td>Cytology</td>
</tr>
</tbody>
</table>
PNH Clinical Programs: Staged Phase III Commitment
Focus on PNH Patient Convenience

**Resistant Program**
- Recruiting
- 2 patients currently on treatment in Holland & USA

**Phase II COBALT**
- FDA post-Phase II review meeting completed

**Phase III CAPSTONE**
- Naïve patients, recruiting
- 6 month treatment
- Up to 30 patients
- End points: transfusion independence & hemoglobin

**Phase III ASSET**
- Treated patients, planned
- Preceded by an initial proof of principle study
- End point: transfusion & hemoglobin

**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

Cell recruitment to Lung Induced by LPS

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

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
Investor Presentation

October 2018