Summary of Topline Phase II Results
Nomacopan for Treatment of Bullous Pemphigoid

April 2020

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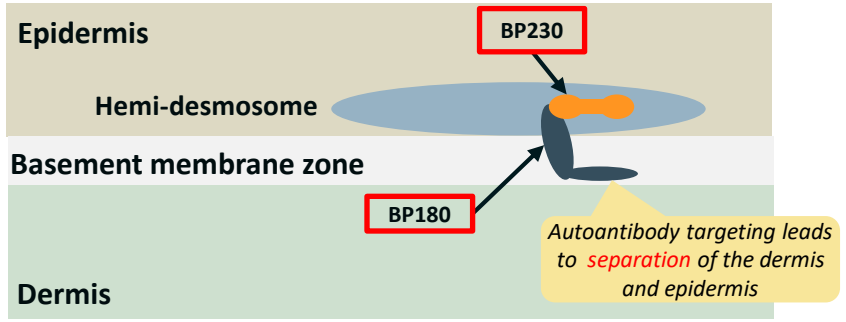
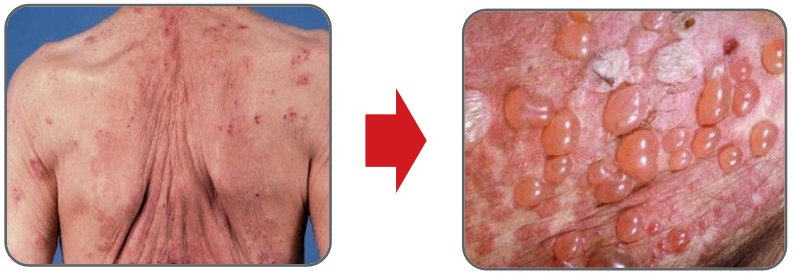
Bullous Pemphigoid
Significant Unmet Need

Schematic of dermal-epidermal junction



Key molecular targets that lead to BP pathophysiology

Presentation of bullous pemphigoid



Urticarial-like skin rash

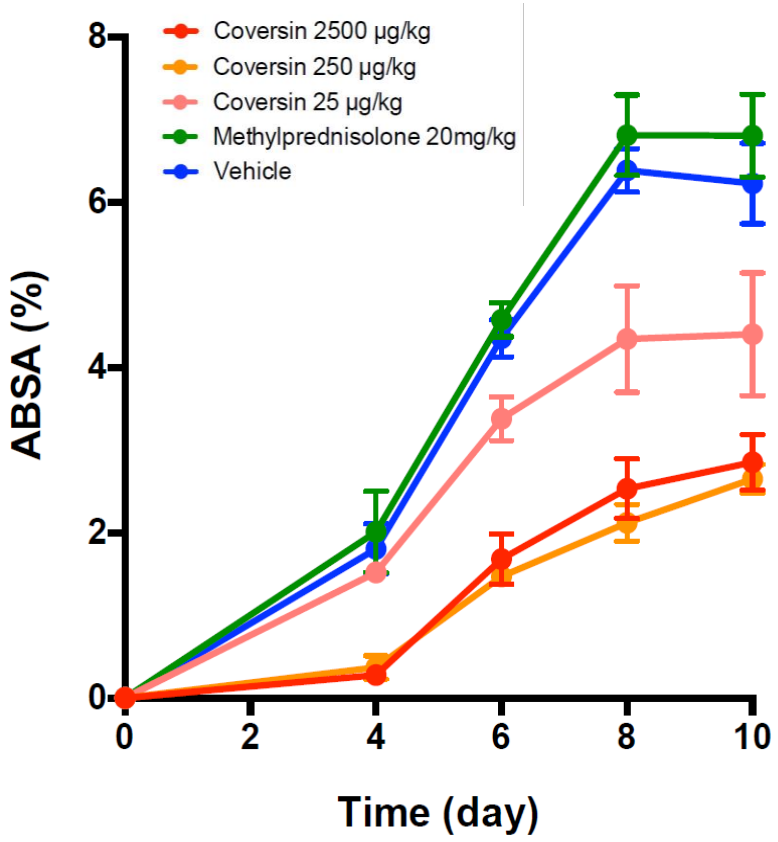
Fluid-filled blisters

- Severe autoimmune blistering skin disorder primarily of the elderly, caused by immune complex mediated inflammation
- **No approved therapy. Current treatment with chronic corticosteroids poses substantial health concerns and 3 – 4-fold increase in mortality**
- **KOLs: essentially no change in standard of care in decades**
- Understanding of BP etiology has grown, but specific targeted therapies are lacking
- Evidence (pre-clinical and human tissue) that C5 and LTB4 have a beneficial and additive role in disease control*
- Orphan condition, with a prevalence of circa 120K patients in US and EU

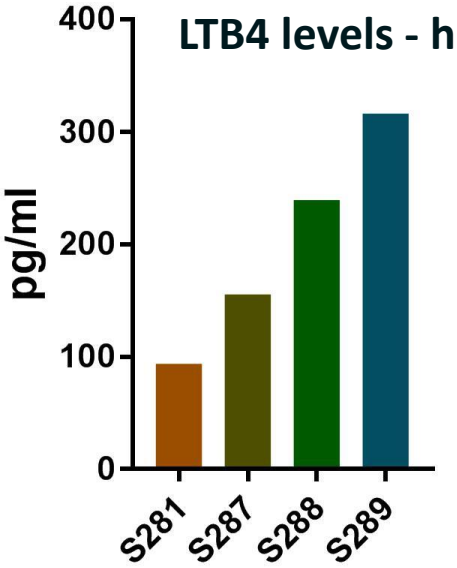
* Preclinical studies with Nomacopan showed a dose response and combined C5 & LTB4 more effective than LTB4 alone: Sezin (2019) *JCI insight 4: e*128239. Akari also found elevated C5/LTB4 in an *ex-vivo* study with Lubeck University of BP patients

Rational for nomacopan in BP
Preclinical Efficacy and Elevated C5a/LTB4 (in Human)

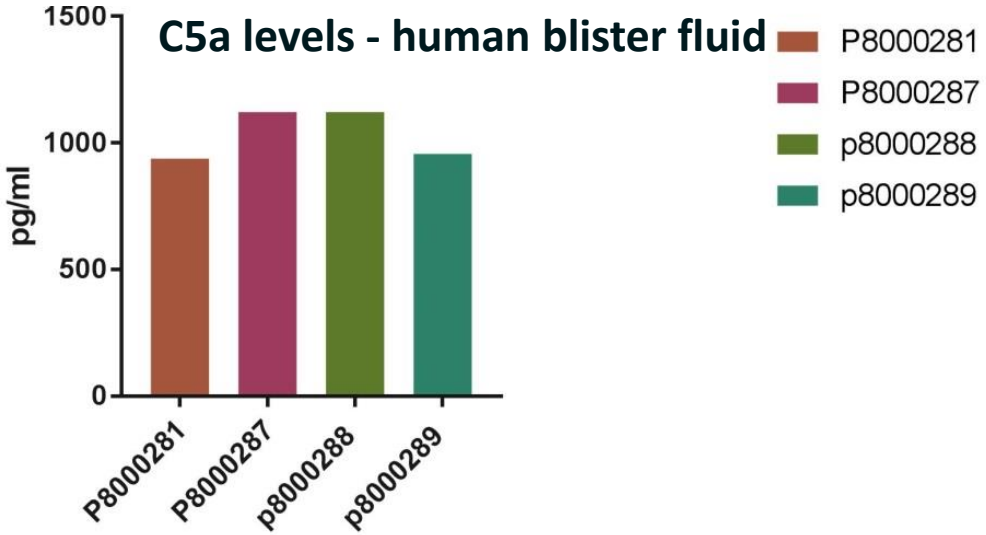
Preclinical passive mouse model of epidermolysis bullosa acquisita (EBA) from Dr. Sadik in Lubeck, Germany; leading Bullous Pemphigoid center



- Clear dose response
- ~60% reduction in affected area on nomacopan (SC) compared to vehicle or steroid
- P=0.002 between vehicle and 250 µg/kg



Elevated levels of LTB4 and C5a indicate activation of complement and synthesis of LTB4 at sites of blister formation



Phase II Study
nomacopan in Bullous Pemphigoid

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

Trial approved in Netherlands (2 sites) and Germany (6 sites)

Study design
• Phase II Single arm (n = 9); 42 days treatment • Active bullous pemphigoid; newly diagnosed or recurrent
Treatment
• Nomacopan SQ dosing • Day 1: 60 mg and 30 mg 12 hours later, Day 2-42: 30 mg once daily

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

- Day 1 to 21: nomacopan (30 mg once daily) + lesional mometasone only
- Day 21 to 42: nomacopan only
- Any use of mometasone or any other steroid after Day 21 considered rescue therapy

Summary of Fully Recruited Phase II BP Data

Patient Demographics

- 7 moderate and 2 mild BP patients; 5 female, 4 male; median age 75; median weight 89.4kg; median BMI 30.2
- Median Karnofsky score 80 (range 60 – 90)
- 5 patients newly presenting BP and 4 relapsing disease
- Typical comorbidities and concomitant medications of an elderly patient population

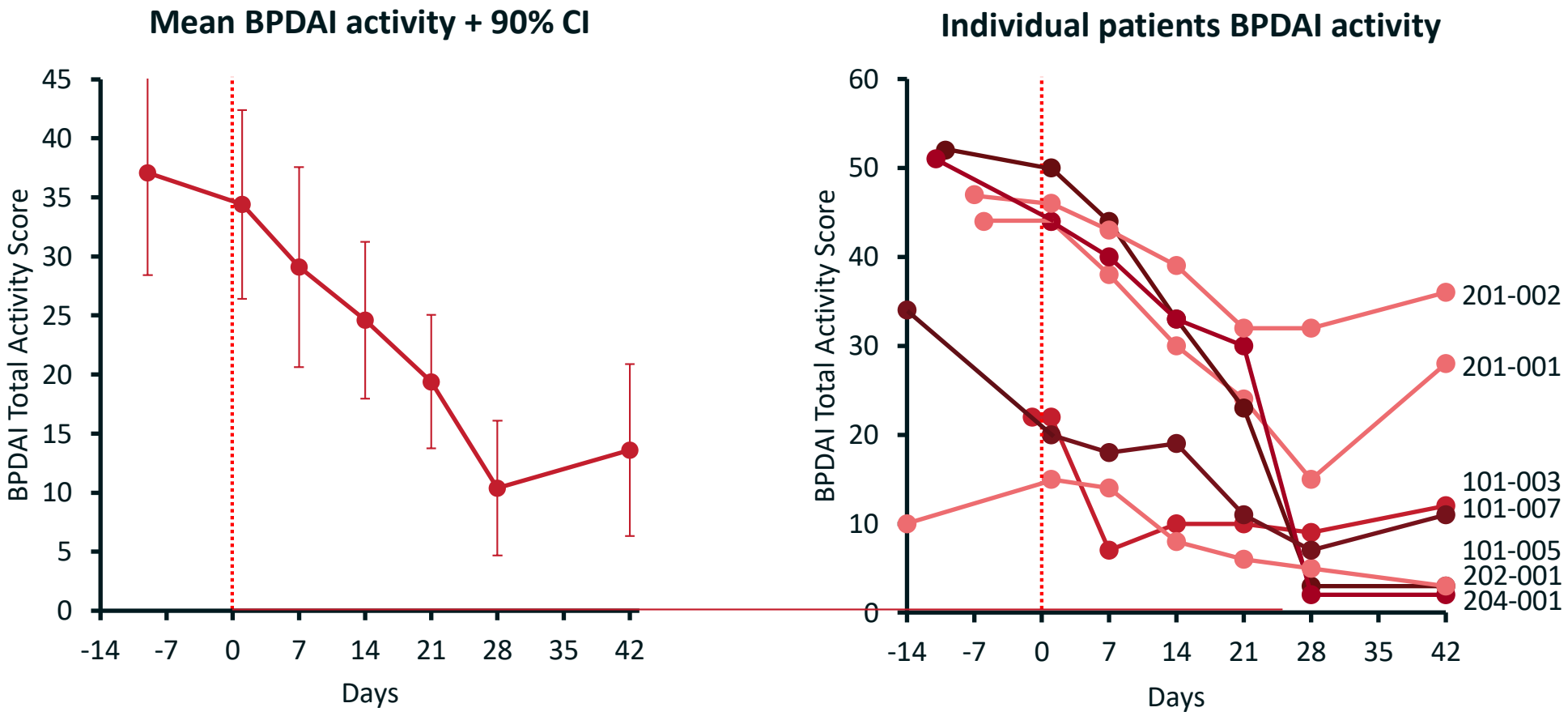
Patient Safety

- No deaths
- None of 9 patients reported grade 3, 4 or 5 treatment-related adverse events

Efficacy

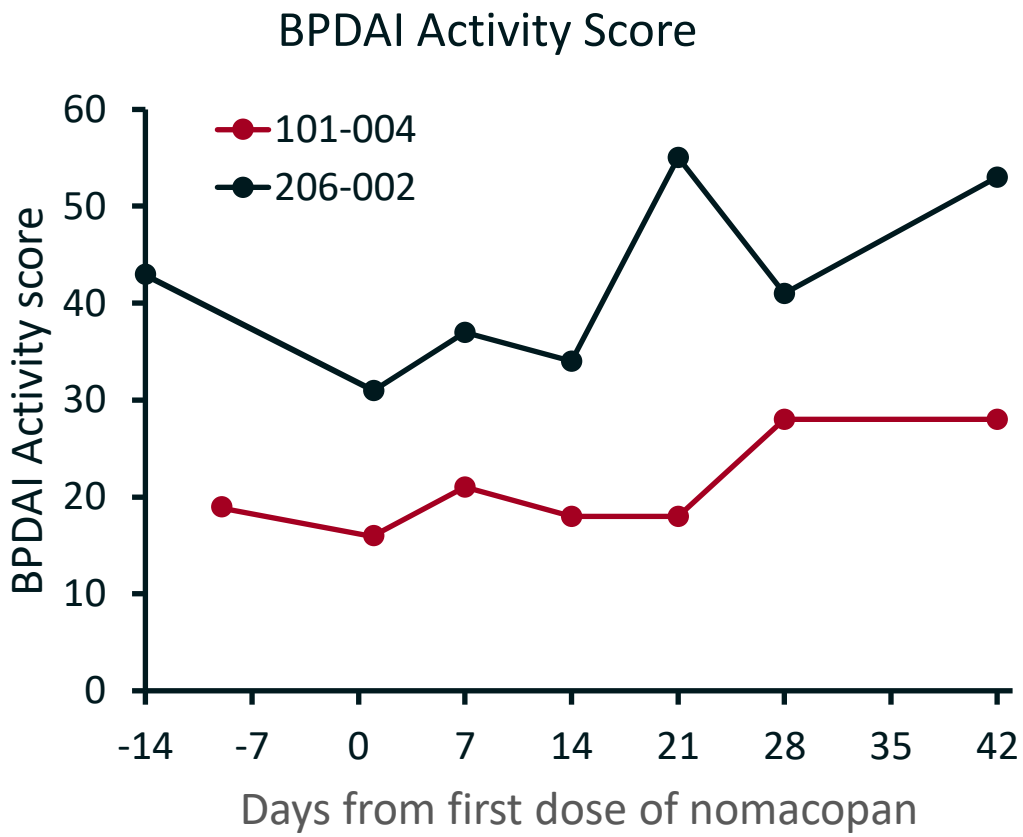
- 7 of 9 patients responded to nomacopan - with 3 showing > 80% reduction in BPDAI activity & 4 showing > 70% reduction in pruritis by Day 42
- 2 of 9 patients were non-responders

Patient BPDAI Activity Scores (n=7) Excluding Two Non-Responders



- Prior treatment withdrawn approximately one week prior to treatment with nomacopan
- Lesional mometasone permitted until Day 21. Mometasone is a moderate topical steroid that was given at a low dose which can help to stabilise BP but is not used to control BP

Patient BPDAI Activity Scores Two Non-Responders



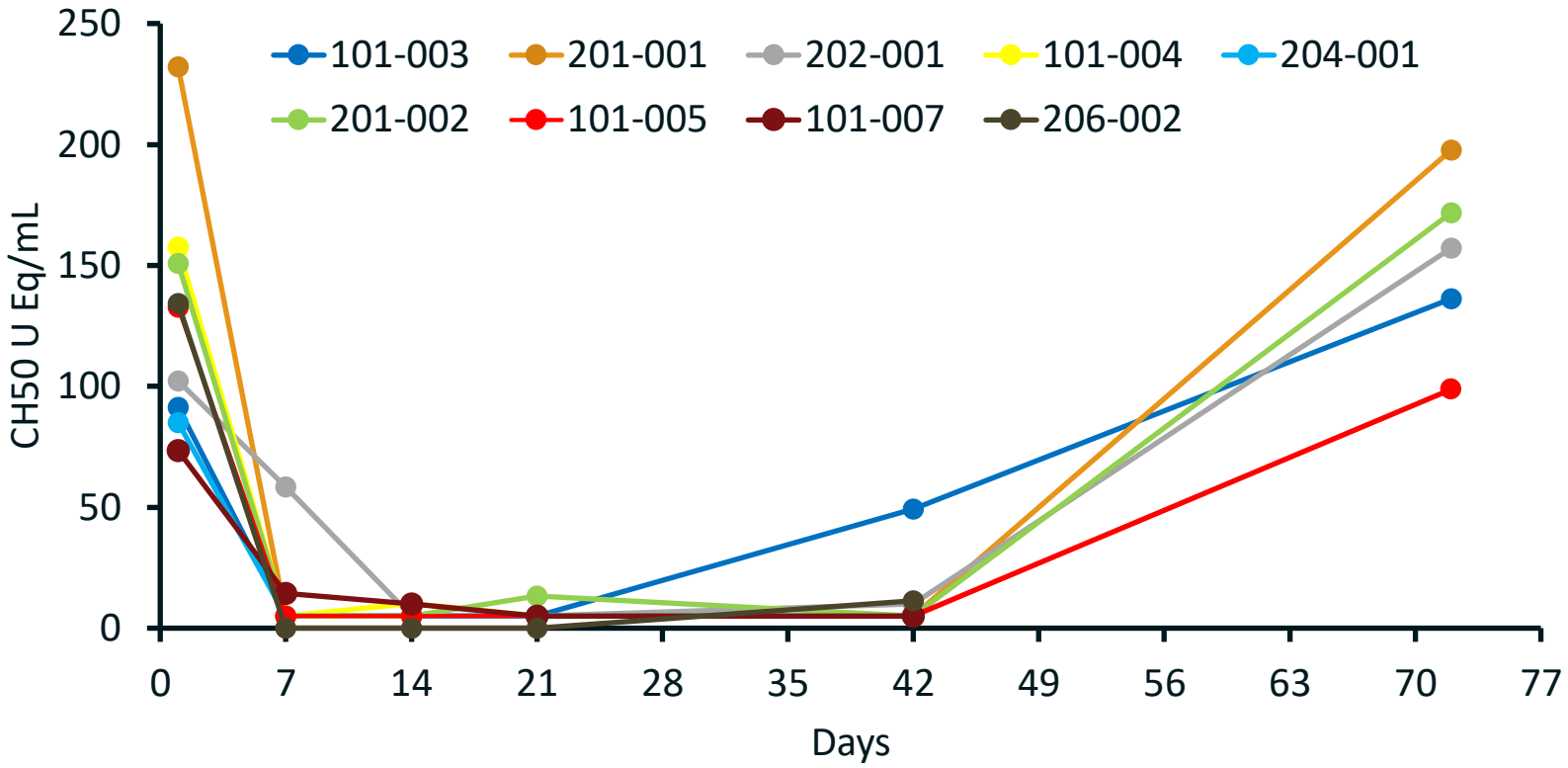
- Both non-responders had long term relapsing disease on admittance
- Combined decline in mean BPDAI activity score at day 42 for all nine patients including non responders was circa 40% at Day 42
- Patient 101-004 received rescue therapy with betamethasone at Day 28 of nomacopan dosing which was ineffective so initiated oral prednisone on Day 39 and topical clobetasol on Day 43. Both super potent steroids were still being used at follow up (Day 72)
- Patient 206-002 received prednisone at Day 62

Concomitant Treatments

- Patients on BP typically are referred to a hospital and, as such, may be on treatment prior to dosing. Oral steroid treatments were an exclusion from the Phase II study
  - 5 out of 9 patients on treatment prior to nomacopan dosing
  - 3 were on topical clobetasol (a relatively strong tropical steroid) which was stopped 2 to 7 days prior to treatment with nomacopan to avoid residual effects
  - None on oral steroids

- Mometasone provided topically to lesions for up to day 21 as this was the first study of nomacopan in BP patient
  - Experts consider that mometasone may stabilize but not improve BP symptoms
  - Mometasone is a short-acting, mid-strength topical steroid not typically given as a treatment for BP

Terminal Complement Activity - BP Patients



- Ablating dose Day 1: 60mg and 30 mg 12 hours later
- Maintenance dose: 30mg once daily until Day 42

Notes:

a) Data to Day 42 for all 9 subjects and follow up data at Day 72 for 5 patients

b) 202-001 did not receive a full ablating dose (60mg total on Day 1 rather than 90mg)

c) 101-003 had low (near normal) total C5 on Day 42 suggesting that the patient may recently have missed one or more doses of nomacopan

Key Phase II Conclusions

Primary Observations

1) Rapid clinical response on nomacopan
2) Quantum of response similar to potent oral steroids
3) Indications that remission may be achievable with 6 weeks of treatment
4) Supports a Phase III design of rapid steroid tapering
5) Good safety profile and well tolerated

Unknowns

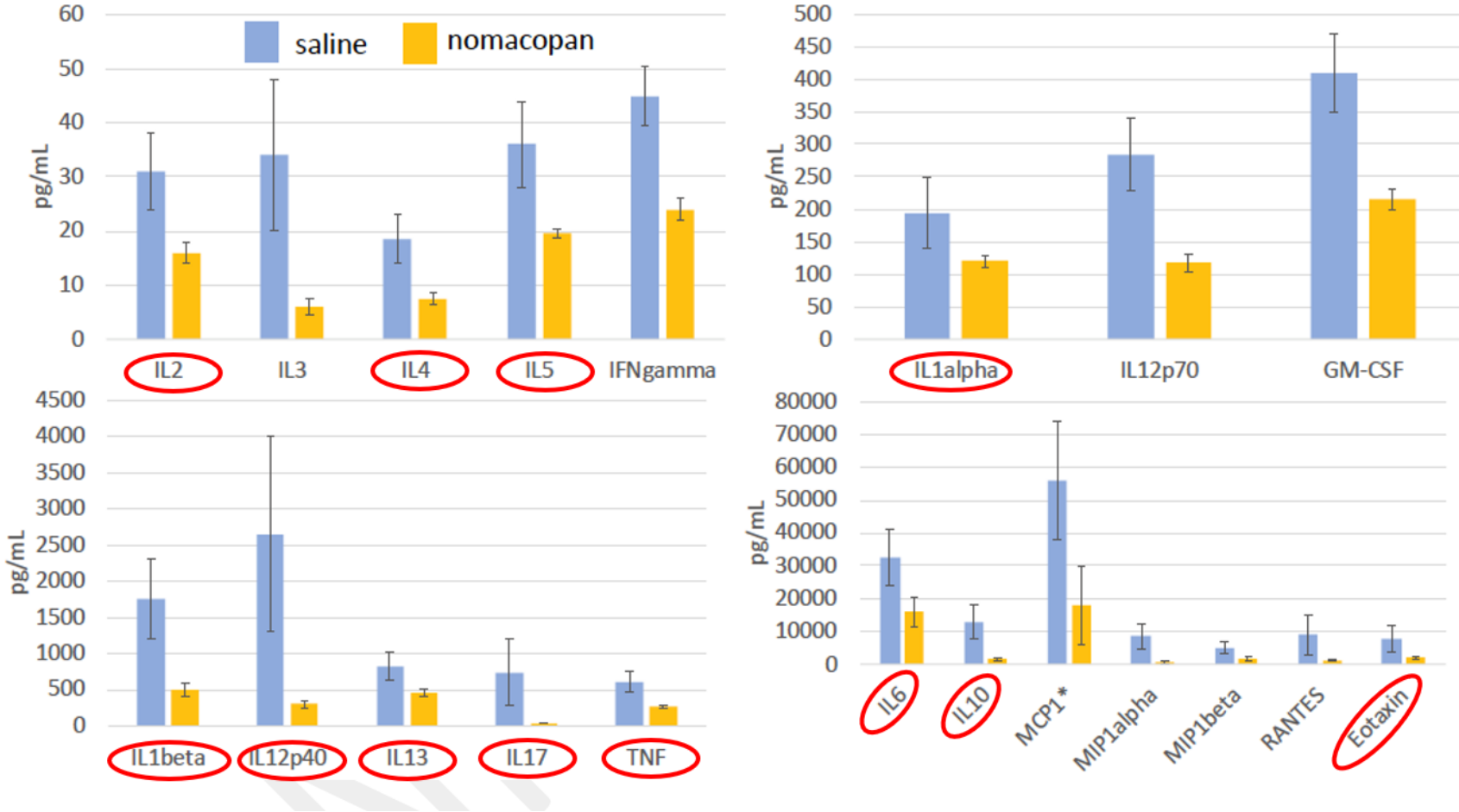
1) Longer term remission rates
2) Additive effect of nomacopan and oral steroids

Therapeutic Approaches to BP

- Major unmet medical need of the elderly in aging patient population
- No approved therapy for BP and no new treatment in last 40 years
- BP treated with steroids increases risk of mortality compared to age-matched controls by ~3-4X; causes hospitalization/clinical and home nurse home care
- Several drugs have been tested in trials for BP, and are believed to not have subsequently progressed into further development:
  - Novartis – ligelizumab - anti Ig E
  - GSK - mepolizumab - IL5 inhibition
  - Bertilimumab - Eotaxin-1 antibody
  - Eli Lilly - Taltz - Anti-IL17a mAb
  - Rituximab : targets CD20
- Few other known biological drugs currently in development
  - Dupilumab – IL4 and IL13 inhibition

Nomacopan Inhibits Downstream Cytokine & Chemokine Signalling Cascades Implicated in BP Pathology

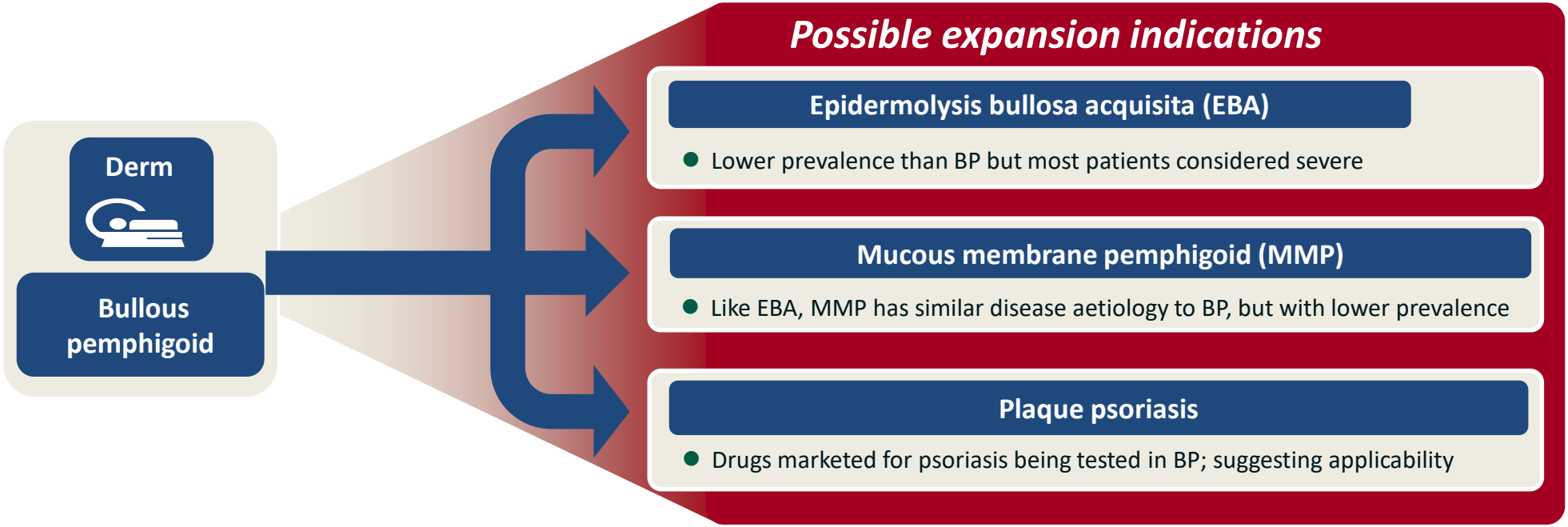
Inflammatory markers significantly inhibited by nomacopan in mouse model of polymicrobial sepsis. Cytokines/chemokines circled in red have been associated with BP



Data adapted from: Figures 1, 2 and 3 of Huber-Lang et al, *J Immunol.* 2014; 192: 5324 – 5331

Notes: *value shown for MCP1 is scaled to10-fold less than measured value to allow use of same y-axis scale; chemokines are MCP1 (CCL2); MIP1alpha (CCL3); MCP1beta (CCL4); RANTES (CCL5) and eotaxin-1 (CCL11)

BP: Gateway to Other Indications



- Other autoimmune blistering skin diseases are categorized by auto-antibodies to either structural epidermal proteins or dermal-epidermal junction proteins
- Symptoms follow that of BP and are generally moderate to severe
- Generally, physicians treating BP will also treat other AIBDs such that approval in BP may drive physician interest in nomacopan for other diseases

Source: LEK 2018 Report

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