Scientific meeting of the

# International Pemphigus & Pemphigoid Foundation (IPPF)
# &
# the PEGASUS Research Group

**-Online-**

## September 19-21, 2021

***Program***

**Program Committee:**
Barbara Horvath | Luca Borradori | Rüdiger Eming | Jens Waschke | Michael Hertl

**Abstract Committee:**
Roberto Maglie | Christian Möbs | Farzan Solimani | Ritva Tikkanen
Franziska Vielmuth | Jens Waschke | Holger Garn | Michael Hertl

**Meetings starts SUN-TUES at CEST 14.00/2 pm**

**i.e. WST -9 hours (San Francisco) 5.00/5 am, EST -6 hours (New York) 8.00/8 am, CST +6 hours (Beijing) 20.00/8 pm, JST +7 hours (Tokyo) 21.00/9 pm**

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DFG Deutsche Forschungsgemeinschaft
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## Sunday 19th September 2021:

**14:00** **Welcome notes**
**Kevin Mead** (Executive Director IPPF)
**Michael Hertl** (Speaker PEGASUS)

**14:15** **Introductory Talk**
**Marc Yale** (Advocacy & Research Coordinator IPPF)
"Patients' need and expectations, current challenges and initiatives"

**14:30 – 16:00** **Session I** **Skin homeostasis and loss of adhesion**
Chairs: John Stanley (Philadelphia), Luis Diaz (Chapel Hill)
Moderators: Zhi Liu (Chapel Hill), Donna Culton (Chapel Hill)

14:30 **Masayuki Amagai** (Tokyo): Immune pathogenesis of pemphigus

15:00 **Detlef Zillikens** (Lübeck): Immune pathogenesis of the pemphigoids

15:20 **Rüdiger Eming** (Marburg/Koblenz): Pemphigus - preclinical models

15:40 **Hideyuki Ujiie** (Sapporo): The active mouse model for bullous pemphigoid

**16:00 – 16:20** **Oral abstract session I**

16:00 **Shirin Emtenani** (Lübeck): Cutaneous kinase activity correlates with treatment outcomes following PI3K delta inhibition in mice with experimental pemphigoid diseases

16:10 **Eliane Müller** (Berne): Pemphigus vulgaris antibodies activate stem cells (CS): a new role of Dsg3 transadhesion in the regulation of the bulge SC compartment

**16:20 – 16:30** **Coffee break**

**16:30 – 17:30** **Session II** **Diagnostic criteria of pemphigus and pemphigoid: practice gaps**
Chairs: Barbara Horvath (Groningen), Victoria Werth (Philadelphia)
Moderators: Branka Marinovich (Zagreb), Valeria Aoki (Sao Paulo)
**This session is dedicated to Marcel Jonkman, Groningen.**

16:30 **Luca Borradori** (Berne): Practical gaps in bullous pemphigoid

16:50 **Jane Setterfield** (London): Mucous membrane pemphigoid

17:10 **Enno Schmidt** (Lübeck): European Guidelines - status quo

**17:30 – 18:00** **Oral abstract session II**

17:30 **Hendri Pas** (Groningen): The autoimmune IgG subclass response defines the IgG deposition pattern in pemphigus patient skin

17:40 **Grant Sprow** (Philadelphia): Mucous membrane pemphigoid flares and disease conversions in patients following Covid-19 vaccination: a case series

17:50 **Avani Kolla** (New York): A retrospective cohort study using Methotrexate as a steroid-sparing or single agent in the treatment of pemphigus vulgaris and foliaceus

**18:00** **Barbara Horvath** (Groningen)**:** Closing remarks

## Monday 20th September 2021:

**14:00 – 15:00** **Session III** **Epidemiology and Genetics**
Chairs: Russel Hall (Chapel Hill), Saleh Ibrahim (Lübeck)
Moderators: Johannes Schumacher (Marburg), Daniel Mimouni (Petach Tikva)

14:00 **Eli Sprecher** (Tel Aviv): The genetic basis of pemphigus vulgaris

14:20 **Animesh Sinha** (Buffalo): Predisposing factors in pemphigus and regional differences: an update

14:40 **Kasia Tasanen** (Oulu): Comorbidities and trigger factors in bullous pemphigoid: recent advances

**15:00 – 15:30** **Oral abstract session III**

15:00 **David Chang** (Philadelphia): A phase 1 trial of desmoglein 3 chimeric autoantibody receptor T cells (DSG3-CAART) for targeted B cell depletion in patients with mucosal-dominant pemphigus vulgaris

15:10 **Kristina Seiffert-Sinha** (Bufallo): Anti-thyroid peroxidase antibodies target a cytoplasmic protein in keratinocytes and may contribute to blister formation in pemphigus vulgaris

15:20 **Enno Schmidt** (Lübeck): Efficacy and Safety of Efgartigimod PH20 subcutaneous in adult patients with pemphigus vulgaris and pemphigus foliaceus: ADDRESS, a global phase 3 clinical trial

**15:30 – 15:40** **Coffee break**

**15:40 – 17:10** **Session IV** **Impaired adhesion: mechanisms**
Chairs: Takashi Hashimoto (Osaka/Kurume), Yasuo Kitajima (Gifu)
Moderators: Rizaak Ahmed (Boston), Eliane Müller (Berne)

15:40 **Kathleen Green** (Chicago): Desmosomes in cell adhesion and beyond

16:05 **Jens Waschke** (Munich): Desmosomal signaling in pemphigus

16:30 **Andrew Kowalczyk** (Hershey): Desmosomal homeostasis

16:50 **Sergei Grando** (Irvine): Non-desmosomal autoantigens in pemphigus

**17:10 – 17:50** **Oral abstract session IV**

17:10 **Franziska Vielmuth** (Munich): Apremilast is protective against PV-IgG-induced loss of intercellular adhesion and keratin retraction in human epidermis

17:20 **Thomas Schmitt** (Munich): $Ca^{2+}$ signalling is critical for antibody-induced blistering of human epidermis in pemphigus

17:30 **Anna Zakrzewicz** (Gießen): Humanized anti-Desmoglein-3 antibodies as tools for research on the role of the neonatal Fc receptor in pemhigus vulgaris

17:40 **Roberta Lotti** (Modena/Milan): PC111, a monoclonal anti-Fas Ligand antibody, blocks blister formation in human pemphigus

**18:00** **Jens Waschke** (Munich)**:** Closing remarks

**18:10 – 20:00** **Poster session (live event with breakout rooms for each poster)**

## Tuesday 21st September 2021:

**14:00 – 14:40** **Session V** **Adaptive and innate immune responses in autoimmune bullous diseases**
Chairs: Philippe Musette (Paris), Dagmar Simon (Berne)
Moderators: Jun Yamagami (Tokyo), Claudio Feliciani (Parma)

14:00 **Ralph Ludwig** (Lübeck): Adaptive and innate immune responses in pemphigoid

14:20 **Meng Pan** (Shanghai): T/B cell interaction in pemphigus

**14:40 – 15:00** **Oral abstract session V**

14:40 **Jussi Tuusa** (Oulu): Cytokine spectra of DDP4 inhibitor-induced and regular bullous pemphigoid skin are different

14:50 **Ayeong Lee** (Seoul): Activated cytotoxic $CD4^+$ T cells produce CXCL13 in a TCR-independent manner in tertiary lymphoid structures of chronic skin lesions in pemphigus

**15:00 – 16:25** **Session VI** **Treatment: from current evidence to future perspectives**
Chairs: Janet Fairley (Iowa), C. Chams Davatchi (Teheran), Miklos Sardy (Budapest)
Moderators: Emiliano Antiga (Florence), Katerina Patsatsi (Thessaloniki)

15:00 **Pascal Joly** (Rouen): Rituximab in pemphigus and the pemphigoids

15:25 **Amy Payne** (Philadelphia): CAART cells in pemphigus

15:45 **Michael Hertl** (Marburg): Therapeutic targeting of T cells in pemphigus

16:05 **Deedee Murrell** (Sydney): BTK inhibitors in pemphigus

**16:25 – 16:45** **Oral abstract session VI**

16:25 **Karolin Wieber** (Marburg): Characterization of Desmoglein 3 reactive $CD4^+$ T cells in pemphigus vulgaris

16:35 **Morna Schmidt** (Aachen): Impact of innate immunity on pemphigus vulgaris disease manifestation

**16:45 – 17:00** **Coffee Break**

**17:00 – 17:10** **Best poster and best oral abstract awards**

**17:10 – 18:00** **Session VII** **What is new in autoimmune bullous diseases? Young investigator forum**
Chairs: Giovanni Di Zenzo (Rome), Frederic Caux (Paris)
Moderator: Karen Harman (Leicester)

17:10 **Joost Meijer** (Groningen): Clinical heterogeneity of bullous pemphigoid

17:30 **Kyle Amber** (Chicago): The role of eosinophils in bullous pemphigoid

17:50 **Khalaf Kridin** (Haifa): Dipeptidyl-peptidase IV inhibitor-associated bullous pemphigoid

**18:10** **Michael Hertl** (Marburg)**:** Closing remarks

# Posters:

**(1) Alexandra Polakova** (Marburg): Rapid isolation of lymphocytes from human skin biopsies

**(2) Anna Zakrzewicz** (Gießen): Humanized anti-Desmoglein-3 antibodies as tools for research on the role of the neonatal Fc receptor in pemphigus vulgaris

**(3) Antti Nätynki** (Oulu): Use of dipeptidyl-peptidase IV inhibitors is associated with decreased SDF-1 levels in type 2 diabetes mellitus and bullous pemphigoid

**(4) Avani Kolla** (New York): A Retrospective Cohort Study Using Methotrexate as a Steroid-Sparing or Single Agent in the Treatment of Pemphigus Vulgaris and Foliaceus

**(5) Ayeong Lee** (Seoul): Activated cytotoxic CD4+ T cells produce CXCL13 in a TCR-independent manner in tertiary lymphoid structures of chronic skin lesions in pemphigus

**(6) Dae San Yoo** (Seoul): Long-term survival of patients with bullous pemphigoid treated with rituximab

**(7) Daniel Seiler** (Lübeck): C5aR2 deficiency ameliorates inflammation in antibody transferexperimental epidermolysis bullosa acquisita and reveals promoting of C5aR1 signaling outcomes

**(8) Dario Didona** (Marburg): A particular case of linear IgA bullous dermatosis: when the clinic plays a pivotal role

**(9) Dario Didona** (Marburg): Dyshidrosiform pemphigoid induced by ustekinumab

**(10) David Chang** (Philadelphia): A Phase 1 trial of desmoglein 3 chimeric autoantibody receptor T cells (DSG3-CAART) for targeted B cell depletion in patients with mucosal-dominant pemphigus vulgaris

**(11) Desalegn Egu** (Munich): Lessons from ultrastructural studies on pemphigus pathomechanisms

**(12) Dipankar De** (Chandigarh): Psychological morbidity in pemphigus patients in clinical remission and its clinico-demographic determinants

**(13) Eliane Mueller** (Berne): Pemphigus vulgaris antibodies activate quiescent stem cells (SC): a new role of Dsg3 transadhesion in the regulation of the bulge SC compartment

**(14) Emtemani Shirin** (Lübeck): Detailed evaluation of complement activation in the skin and blood of patients with pemphigoid diseases

**(15) Franziska Vielmuth** (Munich): Apremilast is protective against PV-IgG-induced loss of intercellular adhesion and keratin retraction in human epidermis

**(16) Grant Sprow** (Philadelphia): Mucous Membrane Pemphigoid Flares And Disease Conversions In Patients Following COVID-19 Vaccination: A Case Series

**(17) Hanan Rashid** (Groningen): Insights in clinical and diagnostic findings and treatment responses in patients with mucous membrane pemphigoid, a retrospective cohort study.

**(18) Hendri Pas** (Groningen): The autoimmune IgG subclass response defines the IgG deposition pattern in pemphigus patient skin

**(19) Ivo Stoykov and Peter Verheesen** (Ghent): Efficacy and Safety of Efgartigimod PH20 Subcutaneous in Adult Patients with Pemphigus Vulgaris (PV) and Pemphigus Foliaceus (PF): ADDRESS, a Global Phase 3 Clinical Trial in Progress

**(20) Ji Yeon Hong** (Seoul): DWP212525, a novel JAK3 and Tec family kinase inhibitor, has the ability to reduce the severity of PV disease through autoantibody inhibition

**(21) John David Baker** (Buffalo): Three North American cases of cutaneous Pemphigus vulgaris with no history of mucosal disease.

**(22) Jong Hoon Kim** (Seoul): Pathogenicity and Antigen Specificity of $ICOS^{+}CXCR5^{+}PD\text{-}1^{+}$ T helper cells in Pemphigus Vulgaris

**(23) Julia Hinterseher** (Marburg): Refractory pemphigus vulgaris of the face – think of apples and oranges!

**(24) Jussi Tuusa** (Oulu): Cytokine spectra of DDP4 inhibitor-induced and regular bullous pemphigoid skin are different

**(25) Karolin Wieber** (Marburg): Characterization of Desmoglein 3 reactive CD4+ T cells in pemphigus vulgaris

**(26) Kristina Seiffert-Sinha** (Buffalo): Anti-thyroid peroxidase antibodies target a cytoplasmic protein in keratinocytes and may contribute to blister formation in Pemphigus vulgaris

**(27) Manuela Pigors** (Lübeck): Increased fibrosis in a mouse model of anti-laminin 332 mucous membrane pemphigoid remains unaltered by inhibition of aldehyde dehydrogenase

**(28) Marian Dmochowski** (Poznan): Serum IgG antibodies to desmoglein 1 are detected more often than those to desmoglein 3 in pemphigus patients having IgG deposits: A laboratory experience of a single tertiary referral center

**(29) Merced Leiker** (Buffalo): A novel Pemphigus vulgaris patient-derived antibody with sequence homology to antibodies directed against desmosomal and non-desmosomal targets induces keratinocyte dissociation

**(30) Michael Fuchs** (Munich): Dsg2 is upregulated in pemphigus and undergoes functionally different interactions with desmosomal and classical cadherins

**(31) Morna Schmidt** (Aachen): Impact of innate immunity on pemphigus vulgaris disease manifestation

**(32) Nina van Beek** (Lübeck): Anti-BP230 IgE-autoantibodies in bullous pemphigoid correlate with disease activity

**(33) Nina van Beek** (Lübeck): Site- and autoantigen- specific associations in mucous membrane pemphigoid- lessons learned from a retrospective bicentric observational study

**(34) Peter Sliwiak** (Marburg): Redox enzymes of the thioredoxin family as potential and novel markers in pemphigus

**(35) Pia Stüssel** (Lübeck): Drug repurposing Propranolol as an effective topical and systemic treatment option for experimental epidermolysis bullosa acquisita

**(36) Ramie Fathy** (New Haven): Bullous pemphigoid-like disease arising after COVID-19 vaccination

**(37) Roberta Lotti** (Modena/Milan): PC111, a monoclonal anti-Fas Ligand antibody, blocks blister formation in human pemphigus

**(38) Sabrina Patzelt** (Lübeck): RECAPITULATION OF CLINICAL AND IMMUNOPATHOLOGICAL FINDINGS BY IMMUNIZATION-INDUCED EXPERIMENTAL ANTI-LAMININ 332 MUCOUS MEMBRANE PEMPHIGOID

**(39) Sanjeev Khindri** (London): ARREST-BP: A Randomised Placebo Controlled Phase 3 Study to Evaluate the Efficacy and Safety of Nomacopan Therapy in Bullous Pemphigoid Patients Receiving Adjunct Oral Corticosteroid

**(40) Seedeh Ghorbanalipoor** (Lübeck): Cutaneous kinase activity correlates with treatment outcomes following PI3K delta inhibition in mice with experimental pemphigoid diseases

**(41) Sonal Shah** (New York): Diagnostic Dilemma: Rare Oral Autoimmune Diseases Presenting in Hispanic Males

**(42) Stephanie Goletz** (Lübeck): Sera of patients with bullous pemphigoid and mucous membrane pemphigoid react with C-terminal epitopes on BP180 by immunoblotting and ELISA

**(43) Stephanie Goletz** (Lübeck): Comparison of two diagnostic assays for the detection of anti-laminin 332 autoantibodies in patients with mucous membrane pemphigoid

**(44) Tomas Cunha** (Marburg): The history is now - Walter Lever's classification of blistering diseases

**(45) Veleria Bumiller Bini** (Lübeck): INVESTIGATION OF C5AR1 AND C5/5A IN PEMPHIGUS AND IN HUMAN SKIN ORGAN CULTURE MIMICKING PEMPHIGUS

**(46) Vitus Brix and Lovely Pierre** (Buffalo): Smoking cessation is significantly correlated with disease