



Pemphigus and Pemphigoid Priority Setting Partnership Survey

Welcome

Thank you for your interest in the Pemphigus & Pemphigoid Priority Setting Partnership. We want to find out what people think are the most important areas for future research on the treatment of:

- Bullous Pemphigoid (BP)
- Pemphigus Vulgaris (PV)
- Mucous Membrane Pemphigoid (MMP)

Research studies are often designed and implemented by medical academics and researchers, but we wanted ask the pemphigus and pemphigoid community (patients, their families/friends and health professionals) what should be researched to improve treatments for these conditions.

Previously, we collected questions about the treatment and management of blistering skin diseases from people with bullous pemphigoid, pemphigus vulgaris and mucous membrane pemphigoid, their families and the people who care for them and treat them.

We are now asking you to help prioritise the questions by voting for the questions that are most important to you.

Please take part if you are:

- A person who has pemphigoid or pemphigus
- A carer/relative of someone who has pemphigoid and pemphigus
- A healthcare professional working with people with pemphigoid and pemphigus conditions
- From an organisation representing the interests of people with pemphigoid and pemphigus.

Further information about this project can be found [on the website](#)

This survey should only take a few minutes to complete.



Questions for research

Please tick those questions where you would like to see more research. The list is fairly long so it is important that you read through to the end. You can select as many as you like but try not to select them all, and you do not need to select one in each section. If there are some words you do not understand please refer to the [glossary of terms here](#).

Topical /Local treatments

- How effective and safe are topical treatments for different parts of the body in BP/MMP/PV, and which are the most effective for each part?
- What are the best personal care products to use in BP/MMP/PV including toothpaste, mouthwash, moisturiser and wash products?
- What is the best way to treat skin wounds in BP/MMP/PV including how should blisters/ erosions be best washed and managed and does treatment vary according to body site?
- Are steroid injections into mouth lesions more effective than steroid mouthwashes in pemphigus and pemphigoid?
- How should persistent mouth lesions be best treated in pemphigus and pemphigoid?
- What are the benefits of good oral hygiene and frequent dental hygienist treatments for the oral manifestations of BP/MMP/PV?

Predicting treatment response, treatment duration, remissions and relapses

- Can we predict the response to treatment in BP/MMP/PV and what factors affect this?
- In patients with BP/MMP/PV, how long does it take to achieve remission, how is it best maintained, how long does it last and what is the likelihood of relapse?
- What factors predict relapses in BP/MMP/PV, how can the risk of relapse be reduced and how are relapses best treated?
- How long should tablet treatments for BP/MMP/PV be continued before starting to reduce the dose?

Drug treatments and monitoring

- What are the risks and benefits of the different tablet and injection treatments used to treat BP/MMP/PV? (such as azathioprine, mycophenolate mofetil, methotrexate, cyclophosphamide, chlorambucil, nicotinamide, dapsone, intravenous immunoglobulin, plasmapheresis)
- Can the duration of steroid therapy be reduced by the early introduction of additional oral or injection treatments in BP/MMP/PV?
- Are outcomes for patients with BP/MMP/PV better if treatment is started earlier and with 'stronger' treatments, such as an immunosuppressant or biologic, rather than escalating from 'milder' treatments if they do not work?
- What tests and checks are required and how often should these be done to safely use the drugs required to treat BP/MMP/PV?
- What are the best ways to monitor the response to treatment and progression of disease in BP/MMP/PV?
- Is it possible to identify drugs that block the specific immune pathways for BP/MMP/PV rather than treat them with broad immunosuppressive drugs?

Steroid tablets and injections

- What is the most effective and rapid treatment strategy using steroids (oral or injection) to induce disease control in BP/MMP/PV?
- What are the risks/side effects and benefits of the different forms of steroid tablets and injections and how can the side-effects be best monitored and managed?
- What is the best/most effective dose to prescribe for steroid tablets in BP/MMP/PV including the starting dose, when and how quickly to reduce the dose, and when to stop?
- How effective are steroid tablets compared with topical steroids (creams) in BP/MMP/PV?
- Which patients benefit most from steroids and which patients should avoid them and what other treatments can be used instead of steroids?

Rituximab

- How effective, safe and cost-efficient is rituximab (or similar biologics) in BP/PV/MMP compared to standard steroid/immunosuppressant use, when should it be started and should it be a 1st line treatment?
- Is rituximab (or similar biologic) effective when used as a single therapy in BP/MMP/PV?
- What is the best dose for rituximab (or similar biologics) in BP/MMP/PV including what criteria should determine if and when infusions are repeated?
- Is it safe to combine conventional immunosuppressants with rituximab (or similar biologics)?

Antibiotics including tetracyclines

- Which is the best tetracycline antibiotic to use for the treatment of BP/MM/PV?
- How effective is doxycycline used as a single therapy compared with standard treatments in each of BP/MMP/PV ?
- Is there benefit in the use of doxycycline, or other tetracyclines, as an additional therapy in combination with immunosuppressants for the management of BP/MMP/PV?
- What is the best dose and duration of tetracycline antibiotics in BP/MMP/PV both as single therapy and combined therapy?
- Is Doxycycline effective in preventing the development or reducing the severity of disease in patients with pre-bullous pemphigoid?

Pain/Itch/patient circumstances

- What are the best treatments for BP/MMP/PV in patients with other medical conditions, in the frail elderly, when planning a pregnancy or when pregnant?
- What is the best management of pain at all stages of disease in BP/MMP/PV, whilst minimising the side effects of treatments for pain?
- How can itching be best relieved in BP/MMP/PV?

Mucous Membrane Pemphigoid

- What is the best treatment for preventing and repairing scarring in MMP (medical and surgical)?
- What are the risks and benefits of repeated procedures to reverse the effects of scarring in the upper airway and oesophagus in MMP?
- Can conjunctival scarring be prevented or reversed in ocular MMP?
- Are vision restoring techniques (e.g stem cell therapy, corneal transplant, artificial cornea, gene therapy) effective in the management of ocular MMP?
- What are the most effective procedures for treating ingrowing eyelashes in ocular MMP? Which ones give the best cosmetic results?
- Can dry eye disease in MMP be reversed?
- What are the best topical treatments for dry or swollen/sore eyes in ocular MMP?
- Can topical treatments maintain remission in ocular MMP?

Complementary therapies/ lifestyle/education

- What is the best diet to support patients with BP/MMP/PV? Are there any foods to avoid or take? Do dietary supplements and/or probiotics improve or trigger BP/MMP/PV?
- What lifestyle and environment changes, including exposure to sunlight/ultraviolet light, help in the management of BP/MMP/PV?
- What are the risks and benefits of complementary therapies for BP/MMP/PV?
- What mental health and wellbeing techniques help to manage BP/MMP/PV and the impact of these diseases on mental health?
- What is the quality and availability of patient information on BP/MMP/PV and their treatments and what are the benefits providing this information?



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Of these questions you have selected as important, please choose up to a maximum of ten that are the MOST important to you.

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About you

It would be helpful for us to know a little more about you. Please answer the following questions:

What is your age?

- 16 and under
- 17 - 24 years
- 25 - 44 years
- 45 - 59 years
- 60+
- prefer not to say

What is your gender?

- Male
- Female
- prefer not to say

Which of these best describes you?

- Person who has pemphigus or pemphigoid
- Family/friend/carer of person who has pemphigus or pemphigoid
- Dermatologist
- Oral medicine doctor
- Ophthalmologist
- Geriatrician
- GP
- Nurse in primary/ community care
- Nurse in secondary care
- Other (please specify)



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Please tell us which blistering skin disease you have:

- Bullous Pemphigoid
- Pemphigus Vulgaris
- Mucous Membrane Pemphigoid
- Not sure
- Other (please specify)



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Priority Setting Workshop

Would you be interested in attending our workshop?

We need a few volunteers to help with the final priority setting process to produce a jointly agreed Top Ten research questions. This will involve joining two half day workshops on 21 and 22 September 2022 (online via Zoom). Please provide your contact details below, if you are interested in hearing more about this.

Name

Email Address

Phone Number

Results of this prioritisation exercise will be made available on our website. If you would like to receive the results directly then please provide your email address here

Data Protection:

Your contact details will be held solely for the purpose of this research project and accessed only by the research team at University of Nottingham. Your details will not be shared with any other companies or institutions. You can contact us at any time to remove your information and we will delete your details.



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Thank you

Thank you for completing our survey. You can contact the research team at the University of Nottingham for more information about this exercise - email: ukdctn@nottingham.ac.uk

More information about the PEM Priority Setting Partnership can also be found on [the University of Nottingham website](#)

The Pemphigus & Pemphigoid Priority Setting Partnership is an independent group of patients and health professionals funded by the Nottingham Hospitals Charity. It is overseen and monitored by the James Lind Alliance (www.jla.nihr.ac.uk), a not-for-profit organisation managed by the National Institute for Health Research Evaluation, Trials and Studies Co-ordinating Centre, and co-ordinated by the UK Dermatology Clinical Trials Network, University of Nottingham.