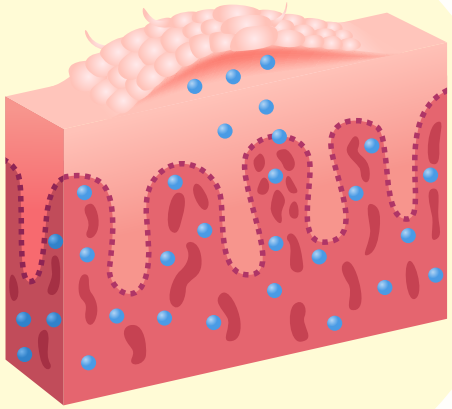
Generalised pustular psoriasis (GPP) is a rare, heterogenous (of different origin) and potentially life-threatening neutrophilic (high white blood cell count) skin disease, which is clinically distinct from plaque psoriasis. GPP is caused by neutrophils (a type of white blood cell) accumulating in the skin, resulting in painful, sterile pustules all over the body.

The clinical course varies, with some people having a relapsing disease with recurrent flares, and others having a persistent disease with intermittent flares. While the severity of GPP flares can vary, if left untreated they can be life-threatening due to complications such as sepsis and multisystem organ failure. This chronic, systemic disease has a substantial quality of life impact for patients and increased healthcare burden. GPP has a varied prevalence across different geographical regions and more women are affected than men.



A new therapy has recently been approved for GPP by the U.S. Food and Drug Administration (FDA), called spesolimab and manufactured by Boehringer Ingelheim under the brand name Spevigo. The treatment is also subject to a UK licence application with the Medicines and Healthcare products Regulatory Agency (MHRA) and, at some stage, will be assessed by both the National Institute for Health and Care Excellence (NICE) and the Scottish Medicines Consortium (SMC).

As the first approved treatment option for generalised pustular psoriasis (GPP) flares in adults, spesolimab is a novel, selective antibody that blocks the activation of the interleukin-36 receptor (IL-36R), a key part of a signalling pathway within the immune system shown to be involved in the cause of GPP.

"GPP flares can greatly impact a patient's life and lead to serious, life-threatening complications," said Mark Lebwohl, M.D., lead investigator and publication author, and dean for clinical therapeutics at Icahn School of Medicine at Mount Sinai, Kimberly and Eric J. Waldman Department of Dermatology, New York. "The approval of Spevigo is a turning point for dermatologists and clinicians. We now have an FDA-approved treatment that may help make a difference for our patients who, until now, have not had any approved options to help manage GPP flares."

It is estimated that one out of every 10,000 people has GPP. Given that it is so rare, recognising the signs and symptoms can be challenging and consequently lead to delays in diagnosis.

In the 12-week pivotal Effisayil 1 clinical trial, people experiencing a GPP flare (N=53) were treated with spesolimab or placebo. After one week, 54% of those treated with spesolimab showed no visible pustules compared to 6% taking the placebo.

With all new treatments, safety is a key part of the research process. Spesolimab is no exception. In Effisayil 1, the most common adverse reactions (around 5%) in people that received spesolimab were asthenia (weakness; lack of energy and strength), fatigue, nausea and vomiting, headache, pruritus and prurigo, infusion site bruising, and urinary tract infection.

"GPP can have an enormous impact on patients' physical and emotional wellbeing. With the FDA approval of this new treatment, people living with GPP now have hope in knowing that there is an option to help treat their flares," said Thomas Seck, M.D., senior vice president, medicine and regulatory affairs, Boehringer Ingelheim. "Spesolimab represents Boehringer Ingelheim's commitment to delivering meaningful change for patients living with serious diseases with limited treatment options."

PAPAA will be submitting evidence to appraisals for this treatment, so if you have any experience of living with GPP, please send us your personal perspective, which we will add to our lived-experience evidence submission. See page 2 for how to contact us.

Source:
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