

NATIONAL INSTITUTE FOR HEALTH AND CARE EXCELLENCE

Health Technology Evaluation

Spesolimab for treating generalised pustular psoriasis flares

Final scope

Final remit/evaluation objective

To appraise the clinical and cost effectiveness of spesolimab within its marketing authorisation for treating generalised pustular psoriasis flares in adults.

Background

Psoriasis is an inflammatory skin disease that typically follows a relapsing and remitting course. The most common form of psoriasis is plaque psoriasis which is characterised by raised plaques on the skin. Generalised pustular psoriasis (GPP), also known as von Zumbusch psoriasis, is a rare form of psoriasis characterised by recurrent episodes of widespread eruption of pustules. This leads to large surface areas of the skin becoming inflamed, red, and developing pustules over the body, specifically in the bends or folds of the skin, genital regions, and fingertips. GPP can be life threatening if left untreated as it can lead to organ failure. Although it is a chronic and persistent condition, its course may be unpredictable, with flare-ups and remissions.^{1,2,3}

Historically, GPP has been classified as a variant of psoriasis consequently assessment and treatment have commonly involved elements from guidance developed for plaque psoriasis. Evidence from genetic, histological, immunological and clinical studies suggest that GPP is a distinct disease as it has unique skin and systemic symptoms.³ The genetic mutations commonly associated with the development of this condition are IL36RN and CARD14. Alteration in these genes can trigger overly active signalling pathways that promote skin inflammation.⁴

The prevalence of GPP in England is unclear. A French study estimated a prevalence of 1.76 patients per million, while a study from Japan estimated a prevalence of 7.46 patients per million. Estimating the number of people affected by GPP is difficult because rare diseases are often misdiagnosed, and patient numbers are small.^{5,6}

There are no specific NICE guidelines for generalised pustular psoriasis. Suspected generalised pustular psoriasis flares should be managed as a medical emergency and immediate same-day specialist dermatology assessment must be arranged. NICE clinical guideline 153 on psoriasis in general recommends that people with psoriasis should be offered topical therapies such as corticosteroids, systemic non-biological therapies, and systemic biological therapies. Some of the treatments recommended for other types of psoriasis might be used for generalised pustular psoriasis flares. However, there are currently no licensed treatments specifically for this condition. In clinical practice, this condition is normally managed in hospital care where topical therapy is provided as well as systemic non biological therapy such as ciclosporin and biological therapies from the IL-17 and 23 family inhibitors.

The technology

Spesolimab (Spevigo, Boehringer Ingelheim Ltd) is a humanised monoclonal antibody that inhibits the action of the interleukin-36 receptor (IL-36R). It is administered intravenously.

Spesolimab currently has a conditional marketing authorisation in the UK for the treatment of flares in adult patients with generalised pustular psoriasis (GPP) as monotherapy.

Intervention(s)	Spesolimab
Population(s)	Adult patients with generalised pustular psoriasis presenting with flares
Comparators	Established clinical management without spesolimab which may include: <ul style="list-style-type: none">• Systemic non biological therapies such as ciclosporin• Biological therapies (such as TNF-alpha inhibitors, IL-17 and IL-23 family inhibitors)
Outcomes	The outcome measures to be considered include: <ul style="list-style-type: none">• symptoms specific to GPP including pain• severity of flares• mortality• response rate• duration of response• relapse rate• adverse effects of treatment• health-related quality of life.
Economic analysis	<p>The reference case stipulates that the cost effectiveness of treatments should be expressed in terms of incremental cost per quality-adjusted life year.</p> <p>The reference case stipulates that the time horizon for estimating clinical and cost effectiveness should be sufficiently long to reflect any differences in costs or outcomes between the technologies being compared.</p> <p>Costs will be considered from an NHS and Personal Social Services perspective.</p> <p>The availability of any commercial arrangements for the intervention, comparator and subsequent treatment technologies will be taken into account.</p>

<p>Other considerations</p>	<p>Guidance will only be issued in accordance with the marketing authorisation. Where the wording of the therapeutic indication does not include specific treatment combinations, guidance will be issued only in the context of the evidence that has underpinned the marketing authorisation granted by the regulator.</p>
<p>Related NICE recommendations</p>	<p>Related Technology Appraisals</p> <p>‘Deucravacitinib for treating moderate to severe plaque psoriasis’ NICE technology appraisals guidance TA907.</p> <p>‘Bimekizumab for treating moderate to severe plaque psoriasis’ (2021) NICE technology appraisals guidance 723. Review date: September 2024.</p> <p>‘Risankizumab for treating moderate to severe plaque psoriasis’ NICE technology appraisals guidance 596. Review date: 2022.</p> <p>‘Tildrakizumab for treating moderate to severe plaque psoriasis’ NICE technology appraisals guidance 575. Review date: 2022.</p> <p>‘Certolizumab pegol for treating moderate to severe plaque psoriasis’ NICE technology appraisals guidance 574. Review date: 2022.</p> <p>‘Guselkumab for treating moderate to severe plaque psoriasis’ (2018) NICE Technology Appraisal 521. Review date: TBC.</p> <p>‘Brodalumab for treating moderate to severe plaque psoriasis’ (2018) NICE Technology Appraisal 511. Review date: TBC.</p> <p>‘Dimethyl fumarate for treating moderate to severe plaque psoriasis’ (2017) NICE Technology Appraisal 475. Review date: TBC.</p> <p>‘Ixekizumab for treating moderate to severe plaque psoriasis’ (2017) NICE Technology Appraisal 442. Review date: TBC.</p> <p>‘Apremilast for treating moderate to severe psoriasis [rapid review of technology appraisal guidance 368]’ (2016) NICE Technology Appraisal 419. Review date: TBC.</p> <p>‘Secukinumab for treating moderate to severe plaque psoriasis’ (2015) NICE Technology Appraisal 350. Static list.</p> <p>‘Ustekinumab for the treatment of adults with moderate to severe psoriasis’ (2009) NICE Technology Appraisal 180. Static list.</p> <p>‘Adalimumab for the treatment of adults with psoriasis’ (2008) NICE Technology Appraisal 146. Static list.</p> <p>‘Infliximab for the treatment of adults with psoriasis’ (2008) NICE Technology Appraisal 134. Static list.</p> <p>‘Etanercept and efalizumab for the treatment of adults with psoriasis’ (2006) NICE Technology Appraisal 103. Static list.</p>

	<p>Note: guidance for efalizumab has now been withdrawn.</p> <p>Appraisals in development</p> <p>‘Spesolimab for preventing generalised pustular psoriasis flares’ NICE Technology Appraisal Guidance [ID6216]. Expected publication date: TBC</p> <p>Related Guidelines</p> <p>‘Psoriasis: assessment and management’ (2012) NICE guideline 153. No new evidence identified in June 2017. Review date to be confirmed.</p> <p>Related Interventional Procedures</p> <p>‘Grenz rays therapy for inflammatory skin conditions’ (2007) NICE interventional procedures guidance 236.</p> <p>Related Quality Standards</p> <p>‘Psoriasis’ (2013) NICE quality standard 40.</p>
<p>Related National Policy</p>	<p>The NHS Long Term Plan, 2019. NHS Long Term Plan</p> <p>NHS England (2023) Manual for prescribed specialist services (2023/2024) Chapter 61: Specialist dermatology services</p>

References

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2. Generalised Pustular Psoriasis. Psoriasis treatment (2015) available from: <https://patient.info/doctor/generalised-pustular-psoriasis> [Accessed June 2024]
3. Burden D, Eng Choon S, Gottlieb A, Navarini A and Warren R (2022) Clinical Disease Measures in Generalized Pustular Psoriasis. American Journal of Dermatology pp.39-50.
4. Generalized pustular psoriasis, NIH Medline plus (2017) available from: <https://medlineplus.gov/genetics/condition/generalized-pustular-psoriasis/#causes> [Accessed June 2024]
5. Augey F, Renaudier P, Nicolas J (2006) Generalized pustular psoriasis (Zumbusch): a French epidemiological survey. European Journal of Dermatology 16(6):669-73.
6. Zheng M, Jullien D and Eyerich K (2022) The prevalence and disease characteristics of generalized pustular psoriasis. American Journal of Dermatology pp.5-12.