

Summary of Burden and Needs: Pemphigus Vulgaris Patient Journey

Auto-immune blistering disease - AIBD

Pemphigus Vulgaris – a rare skin and/or mucous membrane autoimmune disease that affects people of all ages but primarily those over 50

Symptoms	Consequences	Patients and families' needs
<i>First symptoms – Period of diagnostic delay</i>		
<ul style="list-style-type: none"> • Blisters and erosions on the skin that burst and leave the skin raw • Blisters and erosions on the mucous membranes (mouth, oesophagus, nose, eyes, genitals...) 	<ul style="list-style-type: none"> • Pain • Difficulty eating • Difficulty breathing • Difficulty sleeping... 	<ul style="list-style-type: none"> • Obtaining a quick and accurate diagnosis (average diagnostic delay ranges from 6 to 9 months) • Receiving support and care from health care professionals • Getting doctors well informed and trained <ul style="list-style-type: none"> ○ in the pain of the disease & its secondary symptoms AND in the impact of medical examinations (skin biopsies, blood exams) ○ in the intense fatigue ○ in the anxiety
<i>Diagnosis finally established after multiple diagnostic errors</i>		
<ul style="list-style-type: none"> • Blisters in the mouth that burst and leak • Pieces of cheeks, gums come off • Painful wounds • Scalp scabs • Anal and vaginal ulcers 	<ul style="list-style-type: none"> • Multiple additional medical exams • Many appointments with different AIBD trained specialists (dermatologist, stomatologist, proctologist, ophthalmologist, rheumatologist, gynecologist) 	<ul style="list-style-type: none"> • Understanding medical examinations in order to “own” one's own health • Feeling confident with all the different doctors and if possible being able to consult them all the same day • Being able to benefit from psychological support • Adapting one's daily life • Being able to share with other patients & getting support from a patient organization (peer support)
<i>Treatments</i>		
<ul style="list-style-type: none"> • First symptoms & flares relieved by corticosteroids, but long-term use, in turn, may cause excitability, great fatigue, muscular pains... • Immunosuppressants decrease the need of corticosteroids but have side effects (mainly risks of infections) • Biologics, eg. Rituximab, induce long-term remission • IVIG, immunoadsorption in a relapsing/refractory setting • Emerging therapies: clinical trials are available in specialized centers 	<ul style="list-style-type: none"> • Weakened, painful skin that tears easily • Salt free and low sugar diet to avoid diabetes and associated comorbidities, eg. glaucoma • Sleep disturbance • Difficulties to move around • Osteoporosis • Careful monitoring of blood tests to track and diagnose early the potential side effects of treatments 	<ul style="list-style-type: none"> • Being able to contact one's AIBD specialist and family doctor and/or a referring nurse in case of need • Getting dietary advice • Financial coverage for relaxation sessions • Financial coverage for physiotherapy, balneotherapy sessions etc • Being able to understand therapy for better observance
<i>Follow-up care and burden of illness</i>		
	<ul style="list-style-type: none"> • Skin and mucous membranes remain fragile even after the disappearance of blisters and lesions • Risk of bacterial or viral superinfections • Other people's gaze - self-image • Deterioration of intimate life • Administrative problems related to the disease (difficulties in obtaining loans; additional costs from private insurance companies) • Difficulty in combining work, family life and personal life • Financial problems, outstanding balances due to out of cost care • Organization of arrangements if travelling abroad 	<ul style="list-style-type: none"> • Getting support from administrative medical staff • Getting support from occupational physicians • Keeping medical records in case of emergencies or relapses while away from home • Getting help with household chores • Getting support from official patient organizations (peer support)

This patient journey represents the collective patient perspective and experience, and, was prepared by the ERN SKIN AIBD ePAG representative from France with the help of the Italian, UK and German patient groups and in cooperation with the ERN SKIN AIBD doctors.

For more information and a detailed version of this journey, please contact us at: <https://ern-skin.eu/>