Pemphigus Vulgaris (auto-immune blistering disease - AIBD) PATIENT JOURNEY

European Reference Networks

A rare skin and or mucous membrane autoimmune disease that affects people of all ages but primarily those over 50



1. First symptoms

Blisters and erosions on the skin that burst and leave the skin raw

Blisters and erosions on the mucous membranes (mouth, esophagus, nose, eyes, genitals...)

2. Diagnosis

Blisters in the mouth burst and leak Pieces of cheeks and gums come off Painful wounds Scalp scabs Anal and vaginal ulcers

3. Treatment

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 (rituximab) induce long-term remission IVIG, immunoadsorption in a relapsing/refractory setting

Emerging therapies: clinical trials are available in specialized centers

Consequences:

- Weakened, painful skin that tears easily
- Salt free and low sugar diet to avoid diabetes and associated
- comorbidities e.g. glaucoma
- Sleep disturbance
- Osteoporosis; difficulties to move around
- Careful monitoring of blood tests to track and diagnose early the potential side effects of treatments

Needs:

- Being able to contact one's AIBD specialist & family doctor and/or
- a referring nurse in case of need
- Getting dietary advice
- Financial coverage for relaxation, physiotherapy, balneotherapy sessions, etc.
- Understanding therapy for better observance

Consequences:

Pain; difficulty eating, breathing, sleeping Ideally:

- 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 :
- .. in the pain of the disease & its secondary symptoms
- .. in the impact of medical examinations (skin biopsies, blood exams)
- .. in the intense fatigue
- .. in the anxiety

Multiple additional medical exams
Many appointments with different AIBD trained

Consequences:

- specialists (dermatologist, stomatologist, proctologist, ophthalmologist, rheumatologist, gynecologist) Needs:
- 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)

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monitoring, or, if a quick consultation with an AIBD trained specialist

(dermatologist, ENT specialist, gynecologist, dentist, etc.) is needed

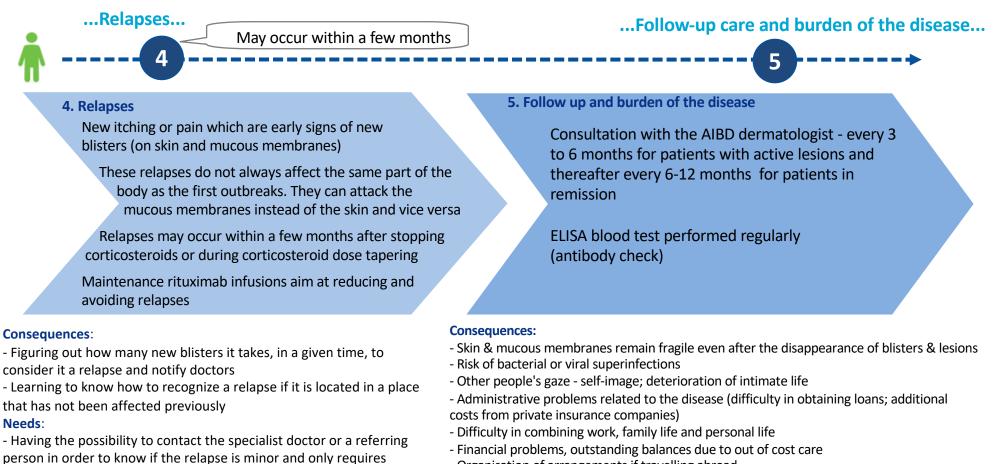
- For patients who have received rituximab as a first-line treatment,

necessary or just regular monitoring

understanding why the dermatologist suggests a preventive infusion if



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- Organisation of arrangements if travelling abroad

Needs:

- 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 patients 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, please contact us: coordination@ern-skin.eu