

Association Pemphigus Pemphigoïde France

PATIENT JOURNEY – Pemphigus Vulgaris (auto-immune blistering disease - AIBD), a rare skin and or mucous membrane autoimmune disease

Stage of Journey	Timeline	Clinical presentation / symptoms	Identify patient needs	Ideal outcome/ support
First symptoms	A few months before diagnosis – people of all ages but primarily in patients over 50 yrs old; extremely rare in children	<ol style="list-style-type: none"> 1. Warning signs that cannot yet be attributed to the disease - these appear, disappear, reappear and then it " flares up "; intermittent flare-ups 2. Painful white blisters in the mouth, under the tongue, on the gums, in the cheeks; these blisters burst and leak out 3. Shedding of skin from tongue, gums and cheeks 4. Progressive difficulty in eating, swallowing and breathing, in sleeping flat to avoid choking 5. Scabs in the nose and bleeding 6. Cough 7. Anal bleeding 8. Spitting up a lot 	<ol style="list-style-type: none"> 1. Need to be quickly diagnosed 2. Need to find the right expert: going to several appointments with general practitioners, stomatologists, ENT (both local and in hospital), occupational physician; none know what we have and usually say so, and, take photos; these visits are often useless - we prefer that a doctor tell us that he doesn't know what we have and direct us to someone who may know 3. Ineffectiveness of the treatments given because of lack of diagnosis 	<ol style="list-style-type: none"> 1. Avoiding diagnostic delay (6 to 9 months) 2. Being able to get support, kindness and attention from the doctors we meet; this is not always the case because many imply that what is happening to us is "in our head" 3. Taking into account the pain of the disease and its secondary symptoms, the difficulties we have in eating and the anxiety linked to multiple painful examinations (biopsies, samples, blood exams) 4. Taking into account fatigue, sleeping difficulties, anxiety related to diagnostic delays and misdiagnosis. These uncertainties are difficult to deal with, as well as the problems due to treatment errors by doctors who do not know how to treat this rare disease 5. Taking into account the impact of the disease on patients' sexual life 6. Dealing with precautionary measures taken to protect one's family in the event of death due to these symptoms
Diagnosis	<ol style="list-style-type: none"> 1. Pemphigus vulgaris 2. Usually established between a 3 to 12 month delay. In patients with exclusive 	<ol style="list-style-type: none"> 1. White blisters in the mouth and on the skin that burst, leak and leave painful wounds; scabs on the scalp; pieces of the inside 	<ol style="list-style-type: none"> 1. Preparing for your first appointment with the AIBD dermatologist: if possible, go with someone and with a list of questions prepared in advance 	<ol style="list-style-type: none"> 1. Feeling more confident: knowing that the doctor knows the pathology is crucial

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	<p>oral lesions, the diagnosis of pemphigus is frequently evoked after other more common diseases of the oral cavity have been first suspected, (oral ulcers, in particular) by GP, dentists or stomatologists.</p> <p>3. First appointment with the AIBD dermatologist</p>	<p>cheek, gums and tongue that shed off</p> <p>2. Oral, nasal, vaginal and anal mucous membranes may be affected. Eyes are rarely involved in pemphigus</p>	<p>2. Being informed of the next appointments - their time and places, in order to be able to organize family and professional life</p> <p>3. Needing to know what you have, to be able to give a name to the disease and to know that you are going to be treated</p> <p>4. Needing to start learning a whole new vocabulary to be able to understand doctors: B cells, lymphocytes, immune system, auto antibodies, biotherapies, and so on</p> <p>5. Seeing an AIBD trained specialist for each affected part of the body – dermatologist, stomatologist, proctologist, ophthalmologist, rheumatologist, gynecologist, ENT</p> <p>6. Feeling relief from pain, itching and burning sensation on the skin</p>	<p>2. Understanding medical examinations in order to “own” one’s own health</p> <p>3. Being able to consult AIBD dermatologists and specialists (ENT, rheumatologist, gynecologist, ophthalmologist, have a bone density test done), if possible, all at once (in a day care hospital, for example, or, through coordinated consultations) and hope that these different doctors communicate their results among themselves</p> <p>4. Being able to benefit from psychological support</p> <p>5. Begin understanding that life and daily life will change and start getting informed about what needs to change in daily activities - especially at work</p> <p>6. Having a disrupted sex life when you have painful blisters and lesions in the vagina or on the penis; and, finding it difficult to talk to doctors about this</p> <p>7. Being able to share with others who have gone through the same experiences – for support and understanding from peers and patient organisations</p>
<p>Treatments</p>	<p>Before diagnosis is established (waiting for the results of the biopsy, for histological examination and direct immunofluorescence</p>	<p>1. Weakening and thinning skin but pain relief</p>	<p>1. Understanding the disease</p> <p>2. Being able to have access to a patients' association that explains how the disease works, that refers to the right doctors and that allows</p>	<p>1. Being informed, better understanding our illness in order to become an active participant in our treatment</p>

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	<p>which usually needs a 7-15 days delay):</p> <ol style="list-style-type: none"> 1. antiseptic mouthwashes 2. topical steroids (skin, mouthwashes, nose...) 3. small amounts of topical lidocaine erase the symptoms and allow you to eat <p>AFTER:</p> <ol style="list-style-type: none"> 1. oral corticosteroids for a duration of between 6months (when corticosteroids are combined with rituximab) to several years (in patients treated with corticosteroids alone or combined with conventional immunosuppressants), usually associated with potassium , proton pump inhibitors , calcium and vitamin D3, or biphosphonate (in patients with a high risk of osteopenia) 2. Rituximab: initial treatment based on 2 infusions 2 weeks apart followed by maintenance infusions 	<ol style="list-style-type: none"> 1. Corticosteroids quickly relieve flare-ups, but trigger a state of excitability and of great fatigue; feeling of having fewer blisters in the mouth; easier breathing 2. Immuno-suppressive drugs and biologics allow rapid reduction of corticosteroid therapy; lesions usually heal within 2 to 3 months after rituximab infusions 	<p>patients to ask the questions they don't dare ask the doctors</p> <ol style="list-style-type: none"> 1. Understanding treatments and their side effects to know if these are normal or if they should alert us 2. Learning how to manage pain, itching and lack of sleep 3. Getting help from weekly physiotherapy or balneo-therapy sessions to help with the difficulties to move around 4. Checking osteoporosis 5. Getting psychological follow-up sessions 6. Re-learning to eat - following a new salt-free and low-sugar diet and not eating anything that is likely to irritate the mouth 7. Understanding blood sampling related to the use of certain drugs 8. Careful monitoring of blood test to track and diagnose early the potential side effects of treatments 9. Applying non-adhesive, tubular dressings 	<ol style="list-style-type: none"> 1. Understanding side effects because they are upsetting; being able to recognize their importance so that you know when to inform your doctors 2. Being able to contact one's AIBD specialist and family doctor, and/or a referring nurse in case of need 3. Treating pain, excitability, lack of sleep other than by additional allopathic treatments - favoring hypnosis, relaxation, and so on; and, getting financial coverage for these sessions 4. Learning how to manage your diet with the help of dieticians so as to avoid excessive weight gain or loss 5. Not minimizing the effects of steroid withdrawal 6. Getting to know the limits between what is normal and what is essential 7. Being cared for by professionals who deal with osteoporosis
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	<p>at months 6, 12 & 18 depending on the evolution of clinical lesions and the change in serum anti-desmoglein antibodies</p> <p>3. Conventional immunosuppressants agents (most often mycophenolate mofetil or azathioprine) if rituximab is contraindicated, unavailable or not reimbursed</p> <p>4. IVIGs: In Europe, and according to the European guidelines, this treatment is given in the case of a refractory pemphigus</p>			<p>8. Being cared for by professionals for local care</p>
<p>Relapses</p>	<p>1. New itching or pain which are early signs of new blisters (on skin and mucous membranes)</p> <p>2. Relapses may occur within a few months after stopping corticosteroids (in particular in patients with persistent high titers of anti-desmoglein antibodies) or during</p>	<p>1. New blisters or scabs on the skin, scalp and mucous membranes</p> <p>2. These relapses do not always affect the same part of the body as the first outbreaks: they can attack the mucous membranes instead of the skin and vice versa</p>	<p>1. Figuring out how many new blisters does it take, in a given time, to consider it a relapse and notify doctors</p> <p>2. Learning to know how to recognize a relapse if it is located in a place that has not been affected previously</p>	<p>1. Having the possibility to contact the specialist doctor or a referring person in order to know if the relapse is minor and only requires monitoring, or, if a quick consultation with an AIBD trained specialist (dermatologist, rheumatologist, gynecologist, dentist, ENT etc...) is needed</p> <p>2. For patients who have received rituximab as a first-line treatment, understanding why the dermatologist suggests a preventive infusion if necessary (if</p>

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	<p>corticosteroid dose tapering. Most relapses occur after tapering corticosteroid lower than 15mg/day; or after reaching a lower level</p> <p>3. Maintenance rituximab infusions aim at reducing and avoiding relapses</p>			<p>the antibody level changes) or a regular monitoring (an antibody check every three months and a check-up every 6 months)</p>
<p>Follow-up and burden of the disease</p>	<p>1. ELISA blood test performed regularly (antibody check)</p> <p>2. Consultation with AIBD dermatologist (every 3 to 6 months in patients with active lesions and thereafter every 6-12 months in patients in remission)</p> <p>3. Throughout the course of the disease</p>	<p>1. Increase, decrease or absence of blisters and sores</p> <p>2. Side effects of treatment</p> <p>3. Occurrence of bacterial or viral super-infections (streptococci, staphylococci, herpes, fungi)</p> <p>4. Frequent feeling of sensitivity, fragility and physical discomfort in the mucous membranes and skin – even after the disappearance of blisters and erosions</p>	<p>1. Preparing a list of questions before each appointment with your AIBD doctor</p> <p>1. Getting regular blood tests to check for anti-desmoglein antibodies (disease activity), to check for kidney, and liver functions; to check for blood cell count, potassium, glycemia and so on, depending on the treatment received (tolerance of treatment)</p> <p>2. Need for a comprehensive management of disease symptoms and side effects - need for physiotherapy; in case of super-infections, treat them promptly</p>	<p>1. Understanding your blood test results so that you can notify your doctor if necessary</p> <p>2. Understanding your antibody levels and the following stages of treatments and the evolution of the disease</p> <p>3. Being able to stay in contact with the health care professionals that follow you in order to handle the side effects resulting from the treatments and those that are ongoing</p> <p>4. Establishing a communication link between your general practitioner and your specialist doctor</p> <p>5. Providing a quick relief from super-infections</p> <p>6. Improving nurse care: dressings take time to do - and nurses don't have enough time to take on patients with PV</p>

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		<p><i>Everyday problems, issues and concerns:</i></p> <ol style="list-style-type: none"> 1. Problems relating to the way other people look at you – burden of others’ gaze: issue of rejection, exclusion from one’s social and/or family environment 2. Intimate life related issues: all dimensions of sexuality are affected - because of a degraded self-image, intense fatigue, erection problems, lubrication problems and so on... 3. Considering pregnancy? Young women have to think about planning one because rituximab 	<ol style="list-style-type: none"> 3. Being informed about vaccines: knowing which ones are necessary: for instance, the flu and the pneumococcal vaccines must be administered before rituximab and immunosuppressant agents; knowing which ones are not recommended and which ones are contra-indicated – like living vaccines: for instance, the one against yellow fever cannot be administered to patients taking immunosuppressants agents or rituximab <p><i>Everyday problems, issues and concerns:</i></p> <ol style="list-style-type: none"> 1. Being able to get psychological counseling 2. Difficulty in sharing sexuality issues with our doctors 3. Being able to manage a pregnancy with pemphigus 	<p><i>Everyday problems, issues and concerns:</i></p> <ol style="list-style-type: none"> 1. Being able to have access to psychological and group support - being able to contact people who are going through the same situation in order to ask practical questions and exchange tips and tricks that help in everyday life – getting support from official patient organisations (peer support) 2. Same as 1 3. To be able to carry out the pregnancy without difficulty for the baby and the mother: need for
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		<p>and some immunosuppressant agents are contraindicated during pregnancy (steroids and azathioprine are allowed)</p> <p>4. Administrative problems related to the illness and their consequences on daily life (difficulty in obtaining loans – for instance, a loan for home ownership; additional costs from private insurance companies ...)</p> <p>5. Difficulty in combining work, family life and personal and social life with the disease - at work people think we are pretending to be tired and we are seen as lazy</p> <p>6. Financial problems, outstanding balances due to out-of-cost care</p> <p>7. Foreign travel/travel arrangements (see above for vaccines)</p>	<p>4. Getting help with administrative tasks and checking the availability of social coverage</p> <p>5. Adapting working hours to the intensity and complications of the illness</p> <p>6. Finding a solution to the very high cost of care: the remaining expenses are significant and have an impact on the family's financial health</p> <p>7. Being able to set up these trips requires careful organization</p>	<p>ongoing communication between the gynecologist, the dermatologist and the general physician.</p> <p>4. Being able to get help from administrative medical staff</p> <p>5. Getting support to manage challenges at work - accommodating the workstation; understanding frequent absences, exhaustion; getting support from occupational health physicians; getting help with household chores</p> <p>6. Finding the necessary contacts in the necessary institutions to push forward the reimbursement policy for our care, which is very costly</p> <p>7. Making sure you have your medical records with you so that you can deal with any emergency in the event of a relapse Traveling with sufficient medication for the duration of the trip</p>
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