

**PEMPHIGUS: A DISEASE STAMPED IN THE SKIN**

Aline Bicalho Matias, Ana Maria Ferreira Roselino

*School of Medicine of Ribeirão Preto, University of São Paulo, São Paulo, Brazil***Source of Support:**

FAPESP (Fundação de Apoio à  
Pesquisa do Estado de São Paulo),  
Process number: 2011/03230-1.

**Competing Interests:**

None

**Corresponding author:** M.Sc. Aline Bicalho Matias[aline\\_bmatias@yahoo.com.br](mailto:aline_bmatias@yahoo.com.br)

Our Dermatol Online. 2013; 4(Suppl. 3): 601-605

Date of submission: 08.09.2013 / acceptance: 13.10.2013

**Abstract**

**Introduction:** Pemphigus are autoimmune blistering diseases that affect the skin and mucous membranes. The blisters characteristics of pemphigus tend to break, causing painful erosions that easily bleed. This study aimed to identify the experiences involved in the triggering of pemphigus and how patients face the illness and treatment.

**Material and Methods:** The study included 31 patients diagnosed with pemphigus foliaceus and vulgaris, under a standardized pulsetherapy treatment. Data collection was through semi-structured interviews, which were audio-recorded, transcribed and analyzed using a qualitative approach.

**Results:** During the journey in search of the phenomenon, were defined two thematic categories, subdivided into eleven subcategories. Months before the triggering of the illness, patients experienced feelings of losses, familiar conflicts and concerns; also showed an experience permeated by heartache and disappointments. After the first signs of pemphigus, patients experienced a long journey until the correct diagnose; worsening of the lesions after the communication of the diagnosis; feelings of isolation, shame and prejudice; interruptions of the future plans; lack of information about the disease and treatment; difficult adherence to the pulsetherapy and the appearance of new lesions or worse by stressful events.

**Discussion:** It might be observed that the disease is not just a biological deviation, but also a social deviance, which explicit the need to adapt to the new reality of the disease and face the isolation, prejudice and shame of living with a stigmatizing disease.

**Key words:** pemphigus foliaceus; pemphigus vulgaris; psychological factors; psychological interview

**Cite this article:**

Aline Bicalho Matias, Ana Maria Ferreira Roselino: Pemphigus: a disease stamped in the skin. *Our Dermatol Online*. 2013; 4(Suppl.3): 601-605.

**Introduction**

The term pemphigus, from the greek pemphix (blister), describes a group of autoimmune blistering diseases involving the skin and mucous membranes, the histologically characteristic is the formation of acantholytic intraepidermal blister and the deposition of IgG autoantibodies on the surface of epidermal keratinocytes. Two main types of pemphigus are stands out: pemphigus vulgaris (PV), in which acantholysis at the basal layer; and pemphigus foliaceus (PF) with subcorneal acantholysis of the granular layer [1].

Pemphigus foliaceus (PF) affects only the skin, and the autoantibodies recognize desmoglein 1, causing surface blisters [2]. It can be sporadic, known as Cazenave Pemphigus, with worldwide distribution, or endemic, known as “Fogo Selvagem”. Pemphigus vulgaris (PV) affects the skin and mucous membranes, the autoantibodies are directed against desmoglein 1 and 3, respectively. The desmoglein 3 is more expressive in the lower layers of the epidermis and in the mucous membranes, the blisters formed are strained and more severe [3]. Its distribution is worldwide, occurs similarly in both sexes, has a peak incidence in the fourth to sixth decade of life and can affect

any age group. In the northeast region of the state of São Paulo (Brazil), the PV has become more frequent, compared to PF, affecting younger patients [4].

Although the morphology of pemphigus is well established, the etiology for the loss of immune tolerance is still under investigation [5].

The blisters and vesicles characteristics of pemphigus tend to break, causing painful erosions or ulcers that easily bleed. In the treatment is common to use glucocorticoids, due to its anti-inflammatory and immunosuppressive proprieties. Although effective in controlling the disease, glucocorticoids can lead to side effects, changing the organic metabolism, which, in turn, can cause accumulation of centripetal fat, reduce the tolerance to carbohydrates, vascular and skin fragility, muscle weakness, hypertension, osteoporosis and increased susceptibility to infections [6].

Thus, there are significant changes in the quality of life of patient, including the absence from work and social life, by the side effects of medications such as the characteristics of the disease [7].

In addition, the skin is considered the largest organ of the human body, sensorial receptors base, responsible for capturing stimuli of cold, heat, touch, pressure and pain and it is responsible for the physiological functions of organic defense, thermal regulation, control of blood flow and gas exchange [8]. As a sensorial organ, it is fundamental in the socialization processes throughout life and it is an important organ of communication, responsive to a variety of emotional stimuli and can affect body image and self-esteem [9].

Through the skin, affects, feelings and conflicts are expressed. It also demonstrates the internal and external organs and plays an important symbolic role of protection, delimiting the self and not self, between the inner world and the outer [10,11].

Such notes have raised questions about how patients experience their illness, in which moment of life noted the first symptoms and the consequences of illness and treatment in your life. This study aimed to identify the experiences involved in the triggering of pemphigus and how patients face the illness and treatment.

### Materials and Methods

In compliance with Resolution No. 196/96, of the Brazilian National Board of Health, regarding research involving human subjects, this study was approved by the Ethics Committee in Research of the Hospital of the Faculty of Medicine of Ribeirão Preto, University of São Paulo (HC-FMRP-USP, process number 503/2011).

The objectives of the study and the conditions of professional secrecy were informed in advance, and the individuals who freely agreed with the work signed the consent form.

### Participants

The study included 31 patients, 20 women and 11 men, diagnosed with pemphigus foliaceus (17 participants) and pemphigus vulgaris (14 participants), with a mean of 4.5

years of symptoms, under a standardized pulsetherapy with dexamethasone and/or cyclophosphamide in the Dermatology division of HC-FMRP-USP.

### Instruments

Data collection was through semi-structured interviews, conducted individually, in face to face situation, following two guiding questions previously defined (How did you noticed something different in your body? How is having a disease such as pemphigus?), allowing openness to new questions that arose during the interview.

The interviews were audio-recorded, with the authorization of the participant. Subsequently, the interviews were transcribed, the material was read and analyzed using a qualitative approach to identifying the concepts, beliefs, values, motivations and attitudes of participants. A thematic content analysis was used as a method [12,13].

To capture the meanings of the experiences, of the perceptions that humans have of their own experiences, attributing meanings followed by feelings [14], the steps proposed by Valle [15] were followed: Search information provided by the subject; Comprehensive analysis of the descriptions, by reading the material, seeking global understanding; Careful reading of the material in order to define the units of meaning; The agreements and disagreements among the units of meaning gave rise to the themes; Description of the structure of the studied phenomenon. During the journey in search of the phenomenon, were defined, initially, two categories of analysis, subdivided into twenty sub-categories, which were presented to three judges with college degrees, two psychologists and a medical doctor, which compared each category with the other, confirming or regrouping them according to their contextual equivalence. Finally, the final cast was composed of two thematic categories, Events Prior to Illness and Existing-in-world with Pemphigus, subdivided into eleven subcategories (Tabl. I).

Categories	Subcategories
Events Prior to Illness	Losses
	Familiar Conflicts
	Concerns
	Heartache/ Disappointments
Existing-in-world with Pemphigus	The journey to diagnose
	Worsening of the lesions after the communication of the diagnosis
	Isolation / Shame / Prejudgment
	Interruption of plans
	A lack of information about the disease and treatment
	The difficult adherence to the pulsetherapy

Table I. Categories and Subcategories.

## Methods

### 1. Events Prior to Illness

The perception of the first lesions was associated with marked and significant events in the lives of patients, mainly related to loss events, family conflict, concerns and heartache/disappointments. The impact and intensity that these events represent in their lives suggest how the events were challenging of their psychological resources to deal with them [16].

Losses: Beyond the the concrete death, the symbolic death

*I guess I never had losses, had always won. Left one, then the other and then the other [children went to college] and I have been getting ... alone and I don't think I knew how to manage these things. I can say it was a drop of water. (Female, 50 years, PF).*

Family conflicts

Family conflicts, between husband and wife and between parents and children/grandchildren were also a recurring theme among the patients interviewed.

*My husband cheated on me, but I suffered silently, not vented, just cried, spoke to no one (...). Soon it [pemphigus] has begun, a month as well. (Female, 33 years, PF).*

Concerns

*I discovered I was pregnant, it was intended (...). I got my baby, soon got cold and he got sick (...). I was afraid to let him in hospital, he had only 21 days. After 15 days my bronchitis attacked and gave bronchus pneumonia. I thought I was going to die, I was bad. I improved. After two months gave me this [pemphigus]. (Female, 32 years, PF).*

Heartache/ Disappointments

Patients also showed an experience permeated by heartache and disappointments, mainly related to the relationship with a aggressive father.

*When I was 2 months my mother dropped my dad and I do not know her. My brother stayed with her, I also do not know him. Then I lived with my father and came a time he took advantage of me, I had no one for me. (...) I feel like a rejected person. (Female, 43 years, PF).*

### 2. Existing-in-world with Pemphigus

The illness causes an accidental crisis in the existence and constitutes a subjective phenomenon, complex, multidetermined, rarely anticipated and experienced in different ways, suffering cultural and environmental influence. The culture in which the being is inserted influences the perception, reaction and communication of the disease [17].

Perceptions and interpretations of the world and the existence occur exclusively in terms of an experiential understanding of being-in-the-world, in which the body has a central role, since it is through this body that the being-in-the-world is revealed. It is the bodily experience that conceptions of health and mental illness are perceived and created.

The journey to diagnose

Patients underwent several medical professionals until receive the diagnostic of pemphigus and be referred for appropriate treatment.

*Seemed thrush, tonsillitis, doctors spoke it was some resistance problem. (...) A doctor began to suspect of lupus, leukemia and*

*started making more complex tests. (...) The doctor said it was SIDA (...) They started to treat gastritis. (Female, 21years, PV).*

The worsening of the lesions after the communication of the diagnose

Patients realized a quantitative and qualitative worsening of lesions after confirmation and communication of the diagnostic of pemphigus.

*They confirmed that it was the Pemphigus that it took the entire body. (Female, 63 years, PV).*

Isolation/ Shame/ Prejudgment

Experience of isolation feelings, shame and prejudice were observed. A skin disease, severe and chronic, carries symbolic associations that can influence how a person identifies their illness and the behavior of others. Thus, it is observed that pemphigus is not just a biological deviation, but also a social deviance.

*It was that thing, small town, people scared. Departed. I suffered a lot of prejudice. (Female, 54 years, PV).*

Interruption of plans

*It seems that our lives finish. It closes in that little world and just (...) I already said that I do not make plans as I used to do. I think my life stopped, it is standing there. (Female, 46 years, PF).*

Appearance of new lesions or worse by stressful events

The reporting of appearance of new lesions by the experience of stressful events was frequently observed.

*I had the graduation from my older son and when it was closer I got worse. I talked to him I was going to ruin his album and he said 'Mom I want you.' (Female, 50 years, PF).*

*Be very happy it [pemphigus] attacks, be very sad it attack, all too much. (Female, 32 years, PV).*

A lack of information about the disease and treatment

A significant gap in the knowledge that patients have about pemphigus and treatment they are subjected to, as well as a difficulty in understanding the information given by medical professionals were observed.

*So, that's what I was told, that it is a chemotherapy [the pulse], but I do not have cancer. (Female, 33 years, PF).*

*This pemphigus is a fungal infection that we handle. (...) it had to treat on the remedy base to cure. (Male, 49 years, PF).*

The difficult adherence to pulsetherapy

We observed a close relationship between adherence to treatment and effective communication between doctor and patient.

*The pulsetherapy is the worst, as I was reluctant to do it! I was not improving with medication and doctors asked to start the pulse. (...) In the first consultation I sat beside a lady and a girl and I heard the girl mention that I also had pemphigus.*

*The lady started crying and told my mom 'Do not let her do the pulse, my daughter died doing it' (...) I understood that the pulse have killed her and it created a huge resistance. (...) The first impression was that the pulse would kill me. I tried a long time with home remedies and there was no improvement and I did not accept the pulse, but there came a time that I could not resist anymore. (Female, 21 years, PV).*

## Discussion

All patients showed stressful life events, striking and significant preceding the triggering of pemphigus, events related to losses, family conflicts and concerns. It was also identified the presence of former stressful life events that marked the history of life of these patients, mainly related to hurts and disappointments.

With the clinical scene installed, patients experienced a real journey in search of answers and appropriate diagnostic Pemphigus, especially the PV to affect the mucous membranes, is considered a serious autoimmune disease due to following a clinical concern when not diagnosed and treated in its early stage [18]. Nevertheless, due to the low incidence, the time between the first symptoms and the diagnosis can be long.

Our way of being, in general, does not provide for the sick. And a skin disease, chronic, stigmatized and of an aggressive treatment represents a disruption in the order to exist. There have been many reports of the interruption of future plans, changes in body image, in the social role and in lifestyle. Abrupt onset and surrounded by striking and significant stressful life events, pemphigus had its evolution permeated by feelings of shame, isolation and prejudice, making the arduous path of adaptation to the reality of illness and treatment.

The analysis of the material obtained in interviews allowed to unveil people of various ages caught by the involvement of a stigmatizing disease, having as one of its consequences the absence of socializing (isolation), revealing the need to spare the enjoyment of others, the social gaze, aesthetic of failure, of the suffering duplicate that they believe they have both in the disease and in the shame of the other's gaze full of strangeness, curiosity and compassion.

Being-in-world with other beings is an ontological fundamental constitution of the human being and the involvement by a chronic illness carries symbolic associations and can influence on how the person identifies their illness and the behavior of others and the easy visibility of a disease skin, such as pemphigus, increases the probability of stigmatization [19].

Thus, the disease is not just a biological deviation, but also a social deviance, which explicit the need to adapt to the new reality of the disease and face the isolation, prejudice and the shame of living with a stigmatizing disease.

Planning the life, the human being, in general, sees a promising future. However, inserted into a different reality, permeated by the destruction of his vanity, autonomy, confidence and significant changes in your body image, might feel defeated before the world [20].

Dominated by pemphigus patients expressed their anguish against their existential conditions, impeding them from being authors of their own history, given to this feeling of imminent possibility of being in shock. It is like if the patient with pemphigus decreed his „death prior” a form of „social death”, as if seeking a hiding to protect him from an attack on his estimates, and thereby surrenders to a process of withdrawal of life, that might mean, the approach of death. The absence of future perspective and the inability to project themselves in this future, to anticipate your image at a time to come, make the present moment empty [21].

The news of a chronic disease such as pemphigus, mobilizes the need to adapt to the new reality, to the social stigma and the implications of the clinical chronicity, factors that can produce overload, conflicts, feelings of disbelief, loss of control and fear

[22,23].

Silva, Castoldi and Kijner [10] suggest that the impact of the diagnostic, the disfiguring appearance and the chronicity of some skin disease might be stressful events and bring serious damages to the quality of life of the individual. Patients suffering from skin diseases experience feelings of inadequacy and stigma forward the current demands of aesthetics. And this feeling of inadequacy and discrimination raises dissatisfaction with himself, and might be focus of stress [24].

Associated to the impact of diagnostic was observed a gap in the patients' knowledge about their disease and treatment, contributing to difficult adherence to the treatment for some patients. According to some reports they postpone the agreement to start pulsetherapy because of the lack of information or of misunderstanding of the information received.

According to Kübler-Ross [25], there are five stages involved in receive and elaboration of bad news. At first, patients experience the initial shock and denial, goes through moments of anger, bargaining, depression, when finally can come to accept the new condition.

Upon to the initial shock, at the diagnostic moment, the way, the language of communication and not open to the clarification of doubts might lead to misinterpretation of diagnosis, prolonging the stages involved in the elaboration and acceptance of the new reality.

The standard pulsetherapy with dexamethasone and/or cyclophosphamide consist in the administration of high doses of drugs during short periods of time in monthly hospitalizations. It is chosen as treatment when the exclusive use of daily doses of corticosteroids is not effective in controlling the disease or carries severe side effects, such as Metabolic Syndrome.

Although the treatment has its efficacy increased, the pulse, in long term, might lead side effects such as sterility by cyclophosphamide, increased blood pressure and weight gain by dexamethasone. Impacted by news of the necessity of this new treatment modality it was observed difficulty in understanding the benefits of therapy, complicating treatment adherence. Adherence that can be defined as the active collaboration between patient and medical team, in cooperative work, with the objective of achieving therapeutic success and can be expressed as the patient's behavior corresponds to the opinion, information or carefully, following instructions for proper treatment [26]. Thus, it can be noted an intimate relationship between adherence and empathetic relationship and effective communication between medical team and patients.

Afraid to constantly question your doctor about diagnosis and treatment the patient keep in silence [27]. Evidencing, therefore, the urgent need to develop more open communication between doctors and patients, enabling higher quality in the relationship. In this direction, in the context of public health, highlights the crucial importance to realize the integrality of the individual in yours bio psychosocial and spiritual context, with the central focus in the improvement of their quality of life. Aiming the multidisciplinary care and taking into account the inter-relationship between mind and body, Rolland [28] points out that the treatment of a chronic illness must go beyond strategies related to the biological understanding of the disease, psychosocial issues should also be considered, as well as family involvement in illness. In the treatment of chronic diseases, the isolated understanding of the case is inefficient, making it necessary the interface of the knowledge.

According to Souza et al. [29], psychotherapy, combined with medical care, might help patients with chronic skin diseases changing your posture and attitudes of in stressful situations, reflecting positively on their quality of life, and consequently of their skin. According to the authors, psychological assistance focused on the identification and management of stressful life events, especially those of intern origin, might enable a new way of symbolizing in which the skin is no longer the vehicle of expression of suffering.

## Conclusions

Just like the skin, one of the major function of the immune system is to distinguish self from the non-self [10,11,30,31]. The epidermal intercellular adhesion plays a vital role. When the mechanism of self-tolerance is broken, breaking occurs due to the binding of autoantibodies to the epidermal self-antigens, resulting in the formation of blisters [32].

Thus, pemphigus, an autoimmune disease with antibodies directed against specific proteins, specifically affecting cells of the epidermis, resulting in a disease that is expressed in the skin leads us to reflect on the existential meaning of this disease for the patient affected by it: a disease in which the body's own components become to identified as aggressors to the immune system. Moreover, it has broken the limitation between the inner world and the outer, between self and non-self. The organism does not recognize and destroy the body's own structures and the individual loses some of its identification through the disfigurement resulting from chronic disease.

Pemphigus mobilizes the need to adapt to the new reality, to the social stigma and the implications of the clinical chronicity. The time after diagnosis and the journey of living with the disease involve a relearning of the meaning of life and an appropriation of consciousness to accept the possibility and the need to maintain hope and the desire to strengthen the development of its existence, creating favorable conditions for treatment.

## REFERENCES

1. Grando SA: Pemphigus autoimmunity: Hypotheses and realities. *Autoimmunity*. 2012; 45:7-35.
2. Cunha PR, Barraviera SRCS: Dermatoses bolhosas auto-imunes. *An Bras Dermatol*. 2009; 84:111-24.
3. Rocha-Alvarez R, Ortega-Loayza AG, Friedman H, Campbell I, Aoki V, Rivitti EA, et al: Endemic Pemphigus Vulgaris. *Arch Dermatol*. 2007;143:895-9.
4. Gonçalves GAP, Cumming Brito MM, Salathiel AM, Ferraz TS, Alves D, Ferreira Roselino AM: Incidência do pêfígo vulgar ultrapassa a do pêfígo foliáceo em região endêmica para pêfígo foliáceo: análise de série histórica de 21 anos. *An Bras Dermatol*. 2011;86:1109-12.
5. Kalantari-Dehaghi M, Molina DM, Farhadieh M, Morrow WJ, Liang X, Felgner PL, et al: New targets of pemphigus vulgaris antibodies identified by protein array technology. *Exp Dermatol*. 2011;20:154-6.
6. da Cunha DF, da Cunha SF, Monteiro JP, Ferreira TP, dos Santos JA, Furtado RA, et al: Nutritional evaluation of pemphigus foliaceus patients on long term glucocorticoid therapy. *Rev Inst Med Trop*. 2000;42:23-6.
7. Timóteo RP, Marques LS, Bertinello D: Intervenção fisioterapêutica promove melhora na qualidade de vida de indivíduos com pêfígo. *Revista da Sociedade Brasileira de Medicina Tropical*. 2010;43:580-3.
8. Anzieu D: *O eu-pele*. São Paulo(SP): Casa do Psicólogo, 1989.
9. Picardi A, Abeni D: Stressful Life Events and Skin Diseases: Disentangling Evidence from Myth. *Psychother Psychosom*. 2001;70:118-36.
10. Silva AK, Castoldi L, Kijner LG: A pele expressando o afeto: uma intervenção grupal com pacientes portadores de psicodermatoses. *Contextos Clínicos*. 2011;4:53-63.
11. Muller MC: *Psicossomática: uma visão simbólica do vitiligo*. São Paulo: Editora Vetor, 2005.
12. Minayo MCS: *O desafio do conhecimento: Pesquisa qualitativa em saúde*. São Paulo(SP): Hucitec; 1994.
13. Triviños ANS: *A pesquisa qualitativa em educação*. São Paulo(SP): Atlas; 1992.
14. Forghieri Y: *Fenomenologia do existir de uma professora universitária (Tese - Livre-Docência)*. São Paulo: Instituto de Psicologia - Universidade de São Paulo, 1991.
15. Valle E: *Câncer infantil: compreender e agir*. Campinas(SP): Psy; 1997.
16. Lazarus RS, Folkman S: *Stress, appraisal, and coping*. New York(NY): Springer, 1984.
17. Lustosa MA: *A família do paciente internado*. *Rev SBPH*. 2007;10:3-8.
18. Kavusi S, Daneshpazhooh M, Farahani F, Abedini R, Lajevardi V, Chams-Davatchi C: Outcome of pemphigus vulgaris. *J Eur Acad Dermatol Venereol*. 2008;22:5804.
19. Barankin B, Dekoven J: Psychosocial effect of common skin diseases. *Canadian Family Physician*. 2002;48:712-6.
20. Leite Almeida CS, Donini Silva JD, Silva VA, Pagliarini Waidman MA: Qualidade de vida sob a ótica de pessoas em tratamento antineoplásico: uma análise fenomenológica. *Rev Eletr Enf*. [Online]. 2011;13:250-8.
21. Birman J: *Futuro de todos nós: temporalidade, memória e terceira idade em psicanálise [w:] Terceira idade: um envelhecimento digno para o cidadão do futuro*. Veras R [res]. Rio de Janeiro(RJ): Relume-Dumará; 1995.
22. Pereira MAO, Pereira Júnior A: Transtorno mental: dificuldades enfrentadas pela família. *Rev Esc Enferm USP*. 2003;37:92-100.
23. Lipp MEN: *Mecanismos neuropsicofisiológicos do stress: teorias e aplicações clínicas*. São Paulo(SP): Casa do Psicólogo, 2010
24. Silva JDT, Muller MC: Uma integração teórica entre psicossomática, stress e doenças crônicas de pele. *Estud. Psicol. (Campinas)*. 2007;24:247-56.
25. Kubler-Ross E: *Sobre a Morte e o Morrer*. São Paulo(SP): Martins Fontes, 1992.
26. Drotar D: Promoting adherence to medical treatment in chronic childhood illness: concepts, methods, and interventions. Nova York(NY): Lawrence Erlbaum, 2000.
27. Petean EBL, Pina Neto JM: Investigações em aconselhamento genético: impacto da primeira notícia: a reação dos pais à deficiência. *Medicina*. 1998;31:288-95.
28. Rolland JS: *Doença crônica e o ciclo de vida familiar. [w:] As mudanças no ciclo de vida familiar: uma estrutura para terapia familiar*. Carter B, Mcgoldrick M (red). Porto Alegre: Artes Médicas, 1995.
29. Souza APFS, Carvalho FT, Rocha KB, Lages, MN, Calvetti PU, Castoldi L: Associação de eventos estressores ao surgimento ou agravamento de vitiligo e psoríase. *PSICO*. 2005;36:167-74.
30. Abbas AK, Lichtman AH, Pillai S: *Cellular and Molecular Immunology*. Philadelphia: Elsevier/Saunders, 2012.
31. Volich RM: *Psicossomática: de Hipócrates à Psicanálise*. São Paulo: Casa do Psicólogo, 2000.
32. Aoki V, Huang MH, Périgo AM, Fukumori LM, Maruta CW, Santi CG, et al: Endemic pemphigus foliaceus (fogo selvagem) and pemphigus vulgaris: immunoglobulin G heterogeneity detected by indirect immunofluorescence. *Rev. Hosp. Clin*. 2004;59:251-6.