

# Chapter 5

## Analysis of Pemphigus Vulgaris

### Clinical Variants of Pemphigus Vulgaris

There are two main different variants of pemphigus vulgaris: one is the mucocutaneous variant and the other is the mucosal dominant variant [1]. The mucosal dominant variant of pemphigus vulgaris involves autoantibodies against only desmoglein 3, whereas the mucocutaneous variant involves autoantibodies against both desmoglein 1 and 3. Some patients, however, report never having mucosal lesions and are classified as having cutaneous-only disease.

Pemphigus vegetans is also a clinical variant of pemphigus vulgaris that involves erosions that evolve into fungoid or papillomatous growths [1]. These fungoid or vegetative growths are mainly seen on the scalp or face. This clinical variant is fairly rare and involves two subtypes: mild Hallopeau and severe Neumann types.

### Classification of Pemphigus Disease Activity

According to the “Consensus Statement on Definitions of Disease Endpoints and Therapeutic Response for Pemphigus,” all pemphigus diseases can be classified according to the level of disease activity [2]. Knowing the disease activity level of pemphigus vulgaris can aid physicians in their decisions about which treatment to start and when in the course of the disease.

## ***Early Endpoints***

The classification of disease activity is divided into four big categories: (1) early endpoints, (2) late endpoints, (3) relapse or flare, and (4) treatment failure [2]. Early endpoints include three stages of disease activity: baseline, control of disease activity, and end of consolidation phase. The baseline is referred to as the level of disease activity on day 1 of when the doctor begins the treatment on the patient with pemphigus vulgaris. Control of disease activity is when the current lesions that the patient has are starting to heal and there are no new lesions cropping up. The end of the consolidation phase of disease activity is when the patient has had no new lesions for at least 2 weeks, and the already existing lesions have healed over. This stage is clinically important because the physician may be able to start weaning the patient off of the immunosuppression therapies at this point.

## ***Late Endpoints***

The second part of this classification includes the late endpoints [2]. Late endpoints of pemphigus diseases include complete remission off therapy, complete remission on therapy, partial remission off therapy, and partial remission on minimal therapy. Complete remission off therapy is the classification that we give the disease when there have been no new lesions and/or established lesions for at least a period of 2 months without any treatment. The “complete remission on therapy” classification is a bit more complicated however. It simply means that there are no new lesions cropping up and no previously established lesions present while the patient is on treatment.

Complete remission on therapy can be further divided into the following categories: minimal therapy and minimal adjuvant therapy [2]. When the patient has complete remission on minimal therapy, the guidelines are referring to the “minimal therapy” as up to a maximum of 10 mg/day of prednisone, an equivalent treatment, or the use of “minimal adjuvant therapy” for at least 2 months. Minimal adjuvant therapy is when half the full therapeutic adjuvant dose is given to the patient.

Partial remission off therapy is when a patient, who has received no forms of treatment for a period of at least 2 months, gets new lesions that then heal within a period of a week without restarting any forms of treatment. Partial remission on minimal therapy is when a patient gets new lesions that heal within 1 week of erupting [2].

Relapse/flare is the third type of disease activity. It is when a patient gets three new lesions within the time frame of a month and they do not heal within a week without any treatment. It also can be defined as a patient who has been in the “control of disease activity” early endpoint, whose established lesions start getting worse instead of healing [2].

Treatment failure is the last type of disease activity discussed in the guidelines. It is basically defined as the failure of bringing the patient to the “control of disease activity” early endpoint despite using the maximum therapeutic doses accepted by the guidelines [2].

## References

1. Bologna JL, Jorizzo JL, Rapini RP, editors. *Dermatology*. 2nd ed. Philadelphia: Mosby; 2007.
2. Murrell DF, Dick S, Ahmed AR, Amagai M, Barnadas MA, Borradori L, et al. Consensus statement on definitions of disease endpoints and therapeutic response for pemphigus. *J Am Acad Dermatol*. 2008;58(6):1043–6.