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**TREATMENT OF PEMPHIGUS AND PEMPHIGOIDS**

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**PEMPHIGUS:**

Autoimmune disease clinically characterized by bullae and erosions located on the skin and/or mucosa. Presents IgC autoantibodies circulating in peripheral blood directed against different proteins of the desmosomes, producing breaks in the intercellular junctions and the subsequent appearance of bullae.

Different varieties are distinguished within pemphigus, some of the most important being pemphigus vulgaris, pemphigus vegetans, pemphigus foliaceus, pemphigus erythematosus, drug-induced pemphigus and paraneoplastic pemphigus.

Pemphigus vulgaris is the most common, representing 85% of the total. Oral lesions tend to precede cutaneous ones by various months.

**Treatment:**

**Aim:** To stop outbreaks as soon as possible. Serious cases with widespread involvement of the skin and mucosa should be treated in hospital.

**Oral corticoids.** Before corticoids mortality from pemphigus was 90%.

They are the treatment of choice. Administered in doses of 1-3mg of oral prednisone per kilo of body weight per day, depending on the severity of the disease, for a period of 6 to 10 weeks. When the outbreak subsides the dosage is reduced gradually to a maintenance dosage of 10-40mg every 48 hours. If possible the corticoids are stopped or minimum dosage is maintained to avoid recurrence. These doses of corticoids produce many side effects (osteoporosis, peptic ulcer, hyperglycemia, hypertension, edema, euphoria, psychosis, myopathy, myasthenia gravis, predisposition to infection).

**Immunosuppressors** are used with the aim of reducing the corticoid dosage, or when the patient cannot tolerate corticoid treatment.

**Methotrexate, Cyclophosphamide** (Genoxal®) at doses of 100mg/day, **Azathioprine** (Imurel®) at doses of 1-2mg/kg/day are used. (determine thiopurine methyltransferase enzyme given that its action depends on efficacy and side effects).

Others: **Chlorambucil** (Leukeran®) doses of 0.1-0.15mg/kg/day, **Cyclosporin A** (Sandimmun®) high doses between 5 and 8mg/kg/day may control outbreaks of pemphigus if associated with prednisone (only for short periods for reasons of nephrotoxicity).

Also used are **Mofetil mycophenolate** (Cellcept®) at doses of 35-45mg/kg/day (0.5-1g twice daily), and gold salts.

**Plasmapheresis.** In serious cases and in the absence of response to corticotherapy.

**Pulse therapy.** Intravenous combination of cyclophosphamide and dexamethasone in high doses.

In **remnant oral** lesions: perilesional infiltration of triamcinolone acetonide or topical application of triamcinolone acetonide 0.1% or flucinolone acetonide 0.05% or clobetasol propionate in Orabase® in concentrations of 0.05% to 0.1%.

**General Measures:**

- Analgesics and antiseptics. Light diet and viscous lidocaine before meals with lesions that impede food intake.
- Patients who receive corticoids for prolonged periods require measures to prevent candida and osteoporosis.
- Skin lesions: topical care using astringents.
- Pemphigus is a serious disease of long evolution. Patients require periodic control.

**PEMPHIGOID**

Chronic mucocutaneous bullous disease, autoimmune and benign in character, with histological absence of acantholysis, and predominantly mucosal lesions.

Classically considered in two forms: bullous and cicatricial pemphigoid, also known as benign mucosal pemphigoid. Factors to be taken into account in treatment are: location, severity and speed of progression of the disease.

<b>HIGH RISK PATIENTS. OCULAR, GENITAL, ESOPHAGEAL AND NASOPHARYNGEAL LOCATION. RAPID PROGRESSION.</b>	<b>LOW-RISK PATIENTS. ORAL MUCOSA AND/OR SKIN ONLY</b>
Prednisone 1-1.5mg/kg/day and cyclophosphamide (1-2mg/kg/day), also intravenously. If cyclophosphamide is not tolerated, azathioprine is used (1-2mg/kg/day). Once under control: slowly lower the corticoid dose while maintaining the immunosuppressive regimen for a longer period.	Moderate or strong corticoids. Triamcinolone acetonide 0.1% or flucinolone acetonide 0.05% or clobetasol propionate in Orabase® in concentrations of 0.05% to 0.1% applied 3-4 times daily. With generalized gingival involvement, buccal splints can be used to improve medication contact with the lesions. Also as a mouthwash when lesions appear in various oral locations.
Another alternative: dapsone (50-200mg/day) for 12 weeks.	Tetracycline 1-2g/day and Nicotinamide 2-2.5g/day.
Immunoglobulin: intravenous (therapy resistant, ocular disease).	If no response, treat with dapsone (50-200mg/day). Hematologic control (hepatic and renal function) and neurological control.
Observations: Patients who receive prednisone and immunosuppressive treatment for prolonged periods require measures to prevent mycoses and osteoporosis.	Another alternative: prednisone (0.5mg/kg, with or without low doses of azathioprine (100mg/day). If severe oral disease is not controlled, high doses of prednisone are required, with or without immunosuppressor.