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## Cicatricial pemphigoid

### What is cicatricial pemphigoid?

Cicatricial pemphigoid is an autoimmune disease that is characterised by blistering lesions on mucous membranes. It is also called "benign mucous membrane pemphigoid". Areas commonly involved are the oral mucosa (lining of the mouth) and conjunctiva (mucous membrane that coats the inner surface of the eyelids and the outer surface of the eye). Other areas that may be affected include the nostrils, oesophagus, trachea and genitals. Sometimes the skin may also be involved where blistering lesions can be found on the face, neck and scalp.

### Who gets cicatricial pemphigoid?

Cicatricial pemphigoid is predominantly a disease of the elderly with a peak incidence at around 70 years. However, childhood cases have been reported. It appears to be twice as common in women than men.

### What are the signs and symptoms of cicatricial pemphigoid?

Site	Features
Eye	<ul style="list-style-type: none"> <li>• Sensation of grittiness or pain</li> <li>• Conjunctivitis</li> <li>• Lesions form, erode and heal to leave scar tissue</li> <li>• May lead to impaired vision or blindness</li> </ul>
Mouth	<ul style="list-style-type: none"> <li>• Blisters form first on the gums near the teeth</li> <li>• Palate, tongue, lips, buccal mucosa, floor of the mouth and throat may be affected</li> <li>• Painful and make it difficult to eat</li> <li>• Lesions occurring in the throat (oesophagus, trachea and larynx) can become life-threatening</li> </ul>
Skin	<ul style="list-style-type: none"> <li>• Blisters on the skin develop in 25–30% of patients</li> <li>• May be itchy</li> <li>• Bleeding may occur if traumatised</li> </ul>
Nose	<ul style="list-style-type: none"> <li>• Nose bleeds after blowing the nose</li> <li>• Crusting causing discomfort</li> </ul>
Genitals	<ul style="list-style-type: none"> <li>• Painful blisters and erosions on the clitoris, labia, shaft of the penis, perianal area</li> </ul>

### Cicatricial pemphigoid



## What causes cicatricial pemphigoid?

Cicatricial pemphigoid is an autoimmune blistering disease, which basically means that an individual's immune system starts reacting against his or her own tissue. In this particular instance autoantibodies react with proteins found in mucous membranes and skin tissue resulting in blistering lesions. The binding site appears to be within the anchoring filaments that help the epidermis (outside layer of skin) stick to the dermis (inner layer of skin).

## Treatment of cicatricial pemphigoid

The primary aim of treatment is to stop blister formation, promote healing and prevent scarring. Cicatricial pemphigoid is a particularly difficult disease to treat as it can affect so many different parts of the body. Usually a team of medical specialists are required for overall care.

Medical treatment is very much dictated by the organs involved.

Organ	Specific treatments
Mouth, nose and skin	<ul style="list-style-type: none"> <li>• <a href="#">Topical steroid</a> creams/ointments</li> <li>• Intralesional steroid injections</li> <li>• Occasional short burst of <a href="#">oral corticosteroids</a> in severe cases</li> </ul>
Eye, oesophagus, trachea and larynx	<ul style="list-style-type: none"> <li>• <a href="#">Dapsone</a> treatment of choice for pure ocular cicatricial pemphigoid</li> <li>• Oral steroids</li> <li>• Immunosuppressants such as <a href="#">cyclophosphamide</a> and <a href="#">azathioprine</a></li> </ul>

Appropriate wound care is particularly important as this should promote healing and minimise scarring.

Cicatricial pemphigoid is a chronic, progressive disease that responds poorly to treatment. Spontaneous remissions are rare. Some patients may experience long remissions with intermittent exacerbations. Patients should identify factors that may cause exacerbations of the disease and avoid these precipitants.

### Related information

**References:**

**On DermNet NZ:**

- [Blistering diseases](#)
- [Bullous pemphigoid](#)
- [Cicatricial pemphigoid](#) - emedicine dermatology, the online textbook

**Other websites:**

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DermNet does not provide an on-line consultation service.  
If you have any concerns with your skin or its treatment, see a [dermatologist](#) for advice.

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