Systemic glucocorticosteroids

We **suggest** using systemic glucocorticosteroids *only* as rescue therapy for acute flares in AE patients.

systemic glucocorticosteroids: general licence for adults and children; starting dose 0.5mg/kg per day; dosage maximum: 1 mg/kg per day

We **recommend against** the long-term use of systemic glucocorticosteroids in AE patients.

Mechanisms of action and efficacy

Glucocorticoids are a class of steroid hormones that bind to the glucocorticoid receptor. The activated glucocorticoid receptor complex upregulates the expression of anti-inflammatory proteins and suppresses the expression of pro-inflammatory proteins, leading to broad anti-inflammatory property.(1)

There are only few studies in adult and paediatric AE patients, despite the regular use of systemic glucocorticosteroids in clinical practice. In studies conducted on children and adults, systemic glucocorticosteroids do not induce long-term remission and swift rebound is common. Systemic glucocorticosteroids have significantly inferior efficacy than ciclosporin.(2, 3)

**Dosage: acute flare, short term, long term**

- Acute flare: Starting dose is usually 0.5 mg/kg bodyweight per day. Treatment should be discontinued or tapered as soon as possible.
- Short-term and long-term: no relevant dosing
- We recommend combining systemic glucocorticosteroids, as any systemic treatment, with emollients and, whenever needed, topical anti-inflammatory treatment in AE patients.

**Safety**

Systemic glucocorticosteroids have a wide therapeutic index. Toxicity is related to the mean dose, cumulative dose and duration of use. At high doses and with long-term use (typically >0.5mg/kg/day) important side effects include skin atrophy, weight gain, sleep disturbance, mood changes, hyperglycaemia or new onset diabetes, peptic ulcers/gastritis, osteoporosis, and increased susceptibility to infections.(4) In particular with long-term use, patients can also develop adrenal suppression and together with a high risk of rebound flares when tapering the treatment dose,
cessation can be challenging. Systemic glucocorticosteroids must therefore be avoided as a long-term treatment in adults and children. Even a fairly high dose can simply be stopped without tapering when used for no longer than three weeks.(5)

**Monitoring**

No standard set of variables are recommended when used for acute rescue therapy, but many patient individual needs for monitoring may apply.

**Combination with other treatments**

There are none of the other treatments in AE that are contraindicated when using systemic glucocorticosteroids.

**Special considerations**

Treatment of acute flares of AE with oral glucocorticosteroids is moderately effective.(2, 3) Systemic glucocorticosteroids have an unfavourable risk/benefit ratio for the long-term treatment of adult and paediatric AE.
References