

Recommendations from the EADV Vasculitis and Vasculopathy Task Force

Most vasculitides are treated with glucocorticoids or immunosuppressants when they progress to a certain stage. This therapy is based on evidence by clinical trials and consented recommendations (e.g. in ANCA-associated vasculitides), but sometimes only on individual experience or habit.

During the Covid pandemic the use of immunosuppressants should be even more restricted when there is not sufficient evidence for their efficacy (as in most cases of IgA vasculitis or of polyarteriitis nodosa cutanea), while they must not be stopped abruptly and prophylactically in ANCA associated and other severe systemic vasculitides.

IgA vasculitis (Henoch Schoenlein purpura)

The most common form of cutaneous vasculitis is IgA vasculitis (formerly called Henoch Schoenlein purpura). Although patients with IgA are often treated with systemic steroids, probably because of the rapid cessation of new lesions, the administration of systemic corticosteroids is not generally indicated, because most flares on the skin are self-limited and not dangerous. Glucocorticoids are only recommended when the formation of necrosis is imminent, heralded e.g. by occurrence of hemorrhagic blisters, because then they may prevent progression to – only slow healing - ulcers. And when they are given, they should be discontinued within one week. If signs of marked systemic involvement occur (arthralgia or severe abdominal pain), systemic glucocorticoids may shorten the duration of pain and other symptoms, but in times of Covid, their use should be restricted. In case of renal involvement. There is so far no randomised controlled trial that has safely demonstrated a therapeutic or prophylactic effect of corticosteroids, neither in childhood nor in adulthood. So consensus groups rather recommend ACE inhibitors or AT1 antagonists for persistent proteinuria of $>0.5-1$ g/day per 1.73 m². In cases of persistent or high proteinuria (>3.5 g/day) and rapid decrease of the glomerular filtration rate, immunosuppressive therapies are started analogous to IgA nephropathy, but during Covid pandemic we would recommend even more a cautious approach and individual decision making by an interdisciplinary boards.

Cutaneous polyarteriitis nodosa (cPAN)

CPAN is a cutaneous vasculitis, not infrequently diagnosed and treated by dermatologists. Immunosuppressants are sometimes used, but should be avoided, because of the non-life-threatening course of the disease. This recommendation weighs even stronger during the Covid pandemic.

It is helpful to consider that in some cases potential triggers can be tackled prior to any medications: as such eliciting drugs (e.g. minocycline) can be discontinued, hepatitis and hemolytic streptococci in pharynx can be treated. For symptomatic treatment of cPAN, compression therapy, NSAID and long-term dapsone or colchicine and perhaps methotrexate can be used without blatantly increased risk for severe course of Covid. Sometimes, when severe pain or a flare cannot be controlled, a short course of systemic glucocorticoids may be warranted.

ANCA-associated and other severe systemic vasculitides

Patients with ANCA associated vasculitis are often treated evidence-based with immunosuppressive medications which weaken their immune system and put them at a higher risk of contracting COVID-19. As is true for many other autoimmune diseases patients on immunosuppressants are not advised to stop their medications unless specifically instructed to do so by their physicians. Stopping these medications can result in a disease flare, which itself may increase the chances of picking up an infection. Yet, for each patient treatment should be individually re-evaluated.

Generally, patients should rather take extra precautions to minimize the risk of getting infected. In addition to the general preventive measures (social distancing, hygienic measures) they should stay at home as much as possible and stock up on necessary medications to a reasonable extent.

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