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Update on management of Vasculitis

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The ANCA-associated vasculitides are a group of rare disorders characterised by necrotizing inflammation of small blood vessels, frequently complicated by crescentic glomerulonephritis, and which untreated are associated with high levels of morbidity and mortality.

A series of well-designed randomised controlled trials during the last two decades has refined immunosuppressive regimens in AAV, which typically comprise a period of remission-induction treatment with high-dose corticosteroids and either cyclophosphamide or rituximab, to gain rapid disease control, followed by an extended period of remission-maintenance treatment that aims to prevent relapse while minimising drug toxicity.

Recent clinical studies have addressed the role of MMF and plasmapheresis for remission induction, the potential for combination drug-approaches, and the use of rituximab for remission-maintenance.

Advances in understanding of the pathophysiology of AAV have also identified a number of potential new therapeutic approaches, including inhibition of B cell survival factors, complement blockade and targeted neutrophil inhibition that are currently being investigated in pre—clinical or early phase clinical studies.

These novel approaches may provide further opportunities to refine treatment strategies in AAV, and to overcome the problems posed by refractory disease, relapsing disease, and treatment-related toxicities.