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Update in Management of ANCA-Associated Vasculitis

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Over 70% of patients with ANCA associated vasculitis have kidney involvement and the severity of kidney injury at diagnosis is a strong predictor for patient and kidney survival. Early diagnosis and institution of effective treatment provides the best opportunity for kidney recovery and is facilitated by PR3 and MPO ANCA testing, suspicion of a vasculitis diagnosis and confirmation by kidney biopsy. Induction therapy aims to remove features of disease activity – remission- and is ideally achieved within 3 months but may take longer. Kidney remission requires stability or improvement of GFR and reductions in proteinuria and hematuria. Remission induction regimens include glucocorticoids, typically IV methyl prednisolone 1500-3000mg, followed by a stepwise reducing dose of oral prednisone from 1mg/kg/day to < 10mg/day by 3 months accompanied by either cyclophosphamide or rituximab. The combination of rituximab with reduced dose cyclophosphamide has gained attraction for kidney vasculitis as being potentially more effective than either immune suppressive on their own and for being glucocorticoid sparing. Plasma exchange aimed at removing circulating ANCA improves chances of kidney recovery for those with high serum creatinine > 300umol/L (3.39mg.dl), and is also used for alveolar haemorrhage with hypoxia. Complement inhibition with avacopan has recently become available as an alternative to glucocorticoids with the potential to increase kidney recovery. Once remission is achieved, the majority of patients are at risk of relapse especially if PR3 or MPA ANCA remains positive at 6 months. Fixed interval rituximab is not the preferred relapse prevention agent which can be used without glucocorticoid. An oral immune suppressive, such as, azathioprine or mycophenolate mofetil, with or without glucocorticoid is an alternative.

Keywords: Vasculitis, Kidney, Treatment, Rituximab, Avacopan