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Immunosuppression to Prevent CKD in Children with IgA Vasculitis

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Immunoglobulin A (IgA) vasculitis, formerly called Henoch-Schönlein purpura (HSP) is the most common form of IgA-mediated systemic vasculitis in children. It mainly affects the skin, joints, gastrointestinal tract and kidney. The overall prognosis of the disorder is favorable, but the long-term prognosis depends on the degree of renal involvement. An older age at onset, purpura persistent for more than > 4 weeks, severe abdominal symptoms, decreased factor XIII activity and the occurrence of relapses are associated with renal involvement in children with IgA vasculitis. Although the pathogenesis of IgA vasculitis is not fully understood, genetic predisposition, abnormal glycosylation of IgA1, complement activation, various cytokines, autoantibodies and abnormal coagulation are involved. IgA vasculitis nephritis (IgAVN) is a mesangial proliferative glomerulonephritis characterized by IgA deposition with varying degrees of mesangial hypercellularity, segmental sclerosis and crescents. Corticosteroid therapy does not prevent the development of nephritis in IgA vasculitis. A previous well conducted randomized, double-blind, placebo-controlled trial showed no benefit of prednisone (4-week treatment) in preventing the development of nephritis in IgA vasculitis, but observed more rapid resolution of nephritis. Treatment of severe IgAVN remains controversial due to a paucity of randomized controlled trials, but various immunosuppressive interventions including methylprednisolone pulse therapy, azathioprine, cyclosporin A, mycophenolate mofetil and plasmapheresis have been suggested to be efficacious. The necessity of early treatment in severe IgAVN has been emphasized to prevent chronic kidney disease (CKD) in children with IgA vasculitis