syndrome

Rapidly progressive glomerulonephritis

haemorrhage

Acute neurologic involvement	Acute neuropsychiatric (psychosis, aseptic meningitis, cerebral vasculitis, and neuropathies) First line treatment: high dose GCs plus CYC. Refractory cases: GCs plus RTX is recommended. Maintenance therapy: azathioprine 2mg/kg/d or MMF 3g/d or cyclosporin 500mg/day and oral prednisolone.
Systemic vasculitis	Intravenous corticosteroids, cyclophosphamide, rituximab, intravenous immunoglobulin, and/or plasmanheresis

Acute profound

a thrombotic
thrombocytopenia
purpura (TTP)-like

Pulses of intravenous methylprednisolone

Life-threatening bleeding or infection: IVIG ± GCs

Refractory cases: Rituximab and/or IVIG , Plasmapheresis, Eltrombopag, Eculizumab, Splenectomy

CYC 750 mg to 1 g/m²/month, or MMF (from 2 to 3 g/day for 6 months), or Tacrolimus 0.1-0.15

mg/kg/day + pulses of methylprednisone 1 g/day for 3 days with gradual tapering.

Refractory cases: Rituximab

Maintenance treatment: MME (from 2 to 3 m/day), available of the 2 to 3 m/day.

Maintenance treatment: MMF (from 2 to 3 g/day), azathioprine (from 2 to 3 mg/kg/day) and prednisone at a tapering dose.

ACE-I: as antiproteinuric agents

Pulmonary IV Glucocorticoid + immunosur

IV Glucocorticoid + immunosuppressive (as Cyclophosphamide) or Rituximab ± plasmapheresis.
refractory cases: IVIG.

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