Susac syndrome Condition for which IVIg use is in exceptional circumstances only

Specific Conditions	Susac syndrome
Level of Evidence	Insufficient data (Category 4a)

Susac syndrome is a rare, microangiopathic disorder characterised by encephalopathy, hearing loss and retinal artery branch occlusions. Case reports show benefit of intravenous immunoglobulin (IVIg) therapy in combination with corticosteroids, with or without other immunosuppressive agents.

Dose: 1–2 g/kg/month for one year, providing documented clinical improvement.

Dosing above 1 g/kg per day is contraindicated for some IVIg products.

Note: Effectiveness of IVIg therapy may be difficult to determine due to the fluctuating course of disease.

Refer to the current product information sheet for further information.

The aim should be to use the lowest dose possible that achieves the appropriate clinical outcome for each patient.

Bibliography

Aubart-Cohen, F, Klein, I, Alexandra, J, et al 2007, 'Long-term outcome in Susac syndrome', *Medicine (Baltimore)*, vol. 86, no. 2, pp. 93–102.

Fox, R, Costello, F, Judkins, A, et al 2006, 'Treatment of Susac syndrome with gamma globulin and corticosteroids', *Journal of the Neurological Sciences*, vol. 251, no. 1–2, pp. 17–22.

Generated on: 31 March 2019