Erythromelalgia

Condition for which Ig is not supported.

Specific Conditions	• Erythromelalgia
Level of Evidence	Insufficient data (Category 4a)

Erythromelalgia is an uncommon pain disorder characterised by episodic extremity redness and burning pain relieved by cooling the affected area (Caldito et al. 2023a, 2023b). Its causes may be classified as genetic, secondary or idiopathic. Most cases are caused by mutations in the SCN9A gene (a sodium channel gene). Secondary causes are mainly due to myeloproliferative disorders (polycythaemia rubra vera and essential thrombocythaemia being the most common), but associations with connective tissue diseases and diabetes have been suggested. There have been several case reports of IVIg being used to treat erythromelalgia but these likely reflect the effect of IVIg on an unrecognised immune disorder as the driver of the erythromelalgia symptoms (Kuroda et al. 2014, Moody et al. 2012).

Currently, the management of genetic and idiopathic erythromelalgia is symptomatic and there is no evidence to support the use of IVIg for this condition. Similarly, the treatment of secondary erythromelalgia is directed at the underlying cause and there is no evidence for the use of IVIg in these situations (Caldito et al. 2023a, 2023b).

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