

Opis choroby *

Definicja

A rare orofacial granulomatosis characterized by the triad of recurrent or persistent orofacial edema (facial and lip edemas), fissured tongue, and relapsing, unilateral or bilateral peripheral facial nerve paralysis. Most cases present with partial symptoms. Typical age of onset is in childhood or adolescence. Histological examination shows non-caseating epithelioid cell granulomas and lymphedema.

Dane

Klasyfikacja

Zespół wad wrodzonych

Kod ORPHA

2483

Kod OMIM

155900

Kod ICD10

G51.2

Kod ICD11

8B88.Y

*Źródło

orphanet