

Opis choroby *

Definicja

Congenital smooth muscle hamartoma (CSMH) is a rare cutaneous hamartomatous lesion most often located on the lumbosacral area or proximal limbs (but rarely on atypical areas such as scalp, eyelid or foot) and characterized by a disorganized proliferation of smooth muscle fibres of arrector pili presenting usually as a localized skin-colored or hyperpigmented plaque (up to 10 cm in diameter) with prominent vellus hairs (most common classic form) or less commonly by multiple skin-colored papules that can coalesce to form irregularly shaped plaques. With time, hyperpigmentation and vellus hairs usually diminish and neither malignant transformation nor associated systemic involvement has been reported.

Dane

Klasyfikacja

Choroba

Kod ORPHA

263435

Kod OMIM

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Kod ICD10

C44.8

Kod ICD11

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*Źródło

orphanet