

## Opis choroby \*

### Definicja

Ichthyosis-alopecia-eclabion-ectropion-intellectual disability syndrome is an ectodermal dysplasia syndrome characterized by severe generalized lamellar ichthyosis at birth with alopecia, eclabium, ectropion and intellectual disability. Although similar to Sjögren-Larsson syndrome, this syndrome lacks the presence of neurologic or macular changes. There have been no further descriptions in the literature since 1987.

### Dane

<b>Klasyfikacja</b>	<b>Synonimy</b>
Choroba	Jagell-Holmgren-Hofer syndrome Zespół Jagella, Holmgrena i Hofera

<b>Kod ORPHA</b>	<b>Kod OMIM</b>	<b>Kod ICD10</b>
2269	242510	Q82.4

**Kod ICD11**  
LD27.0Y

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### \*Źródło

orphanet