

Opis choroby *

Definicja

An extremely rare, major congenital malformation consisting of an absence of the nose ranging from hyporrhinia (absence of external nasal structures) to total arrhinia (absence of external nose, nasal airways, olfactory bulbs, or olfactory nerve) often causing respiratory distress and requiring surgical correction. Arrhinia can be bilateral or unilateral (hemiarrhinia). Associated anomalies include ocular features (hypertelorism, microphthalmia, eyelid coloboma), facial clefts, midline defects and microtia.

Dane

Klasyfikacja

Zespół wad wrodzonych
Isolated nose agenesis
Agenezja nosa

Kod ORPHA

1134

Kod OMIM

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Kod ICD10

Q30.1

Kod ICD11

LA70.0

*[Źródło](#)

orphanet