

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	Synonimy
Zespół wad wrodzonych	Isolated nose agenesis Agenezja nosa

Kod ORPHA	Kod OMIM	Kod ICD10
1134	-	Q30.1

Kod ICD11
LA70.0

*Źródło

orphanet