

## Opis choroby \*

### Definicja

A rare otorhinolaryngological malformation characterized by narrowing of the pyriform aperture (i. e. < 8 to 10 mm in a full-term infant) due to an overgrowth of the nasal process of the maxilla, resulting in potentially lethal nasal airway obstruction in the newborn. Depending on the degree of obstruction, clinical signs and symptoms include inspiratory stridor, respiratory distress, cyanosis, sternal retraction, ribcage asymmetry, and feeding difficulties.

### Dane

#### Klasyfikacja

Zespół wad wrodzonych

#### Synonimy

Isolated apertura pyriformis stenosis

Izolowana hipoplazja otworu gruszkowego nosa

Izolowane zwężenie otworu gruszkowego nosa

Isolated nasal pyriform aperture hypoplasia

#### Kod ORPHA

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#### Kod OMIM

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#### Kod ICD10

Q30.8

#### Kod ICD11

LA70.Y

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#### \*Źródło

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