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
162516

Kod OMIM
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Kod ICD10
Q30.8

Kod ICD11
LA70.Y

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*Źródło

orphanet