

# Heterotopia guzkowa okołokomorowa

**Kod Orpha: 98892 Kod OMIM: 615544**

## Opis choroby \*

### Definicja

Periventricular nodular heterotopia (PNH) is a brain malformation, due to abnormal neuronal migration, in which a subset of neurons fails to migrate into the developing cerebral cortex and remains as nodules that line the ventricular surface. Classical PNH is a rare X-linked dominant disorder far more frequent in females who present normal intelligence to borderline intellectual deficit, epilepsy of variable severity and extra-central nervous system signs, especially cardiovascular defects or coagulopathy. The disorder is generally associated with prenatal lethality in males.

### Dane

#### Klasyfikacja

Podtyp kliniczny

#### Synonimy

PVNH

PVNH

#### Kod ORPHA

98892

#### Kod OMIM

615544

#### Kod ICD10

Q04.8

#### Kod ICD11

LA05.5Y

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[\\*Źródło](#)

orphanet

## Rozszerzony opis choroby

Brak opisu rozszerzonego dla tej choroby. Opracowanie w toku.