

## Opis choroby \*

### Definicja

Rigid spine syndrome (RSS) is a slowly progressive childhood-onset congenital muscular dystrophy (see this term) characterized by contractures of the spinal extensor muscles associated with abnormal posture (limitation of neck and trunk flexure), progressive scoliosis of the spine, early marked cervico-axial muscle weakness with relatively preserved strength and function of the extremities and progressive respiratory insufficiency.

### Dane

#### Klasyfikacja

Choroba

#### Synonimy

Rigid spine congenital muscular dystrophy

Rigid spine congenital muscular dystrophy

#### Kod ORPHA

97244

#### Kod OMIM

602771

#### Kod ICD10

G71.2

#### Kod ICD11

8C70.6

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#### \*Źródło

orphanet