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# Rigid spine syndrome

INSERM

## Source

INSERM. (1999). *Orphanet: an online rare disease and orphan drug data base. Rigid spine syndrome. ORPHA:97244*

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.