

### **Inherited: Neonatal Progeroid Syndrome (Wiedemann-Rautenstrauch syndrome)**

Neonatal progeroid syndrome (NPS), also known as Wiedemann- Rautenstrauch Syndrome, is a rare autosomal recessive disorder characterized by accelerated aging and lipodystrophy from birth. Affected children have extreme intrauterine growth retardation, poor postnatal weight gain, and have a characteristic progeroid face (triangular, old-looking face with a beak shaped or pinched nose), pseudohydrocephalus, wide fontanelles, prominent veins (especially of the scalp) and sparse scalp hair. They also have a distinct pattern of lipodystrophy involving subcutaneous fat loss from the face, extremities, paravertebral and lateral gluteal regions. These features are apparent at birth and therefore this syndrome needs to be differentiated from congenital generalized lipodystrophy. Metabolic abnormalities related to insulin resistance are however uncommon in this condition.

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