

Table 5: Limb-girdle muscular dystrophy: Autosomal dominant

<u>Disease</u>	<u>Onset</u>	<u>CK Level</u>	<u>Progression</u>	<u>Muscular Findings</u>	<u>Extramuscular Findings</u>
LGMD D1 DNAJB6-related	Childhood to late Adulthood	Normal to moderately High	Slow	Lower limb>upper limb; Proximal>distal or distal >proximal; Cramps; Rimmed Vacuoles	Dysphagia (20%)
LGMD D2 TNP03-related	Young child-to adulthood	Normal to mildly high	Rapid in younger	Early proximal weakness; Legs>Arms contractures; Spinal deformities; Rimmed vacuoles	Respiratory insufficiency in younger onset
LGMD D3 HNRNPDL-related	Adolescence to Adulthood	Normal to very high	Slow	Progressive finger and toe flexion limitation; Cramps; Rimmed vacuoles	Cataracts