

Table 2. Physical Exam

Type	Physical Exam findings
SMA-1	<ul style="list-style-type: none">• Severe weakness, atrophy and hypotonia in trunk, neck and proximal>distal limbs, with legs more affected than arms• Bell-shaped thorax, diaphragmatic breathing pattern, bright eyed expression• Absent muscle stretch reflexes• Contractures common
SMA-2	<ul style="list-style-type: none">• Hypotonic, weak proximal>distal muscles, legs more affected than arms• Decreased or absent muscle stretch reflexes• Polyminimyoclonus (fine tremor of hands)• Tongue fasciculations• Contractures and progressive scoliosis develop over time• Normal sensation
SMA-3	<ul style="list-style-type: none">• Proximal weakness and hypotonia• Gait: Trendelenburg, hyperlordosis, may walk on toes as compensatory maneuver to promote knee extension• Positive Gower sign• Decreased or absent muscle stretch reflexes• Ankle contractures develop over time• Tongue fasciculations common
SMA-4	<ul style="list-style-type: none">• Mild proximal weakness affecting gait• Decreased muscle stretch reflexes• No respiratory involvement