

Table 1. Characteristics of SMA by Disease Type

Type	Age of Onset	Function	Survival
SMA-0 (aka type 1a, congenital SMA)	Prenatal	Respiratory failure at birth, severe hypotonia, never sit	Death within weeks
SMA-1 (aka Werdnig-Hoffman disease, 'non-sitters,' severe SMA) (~45% of cases)	0-6 months	Never sit, weak suck, poor feeding, weak cry, progressive severe hypotonia and weakness affecting limbs and respiratory muscles	< 2 years without ventilator support
SMA-2 (aka Dubowitz disease, 'sitters,' intermediate SMA) (~20% of cases)	6-18 months	Can sit unassisted, but unable to stand Initial progressive symptoms, then relative stabilization of weakness with slow progression over years	>25 years
SMA-3 (aka Kugelberg-Welander disease, 'ambulators,' mild SMA) (~30% of cases)	> 18 months	Ambulatory for some period of time, with proximal > distal weakness affecting legs > arms Relatively stable with slow progression over years	Adult
SMA-4 (aka adult SMA) (< 5% of cases)	> 30 years	Ambulatory with mild weakness developing in adulthood	Adult