

**Supplementary Table 1 Clinical motor phenotypes**

Clinical motor phenotype	Clinical features		Clinical site of onset (~% frequency)	Number of all cases (~%)	Age at onset (mean, years)	M:F	Prognosis Median in yrs ( 10-yr survival rate, %)
	UMN	LMN					
'Typical' phenotype:							
Classical ALS	+	+	UL (34) LL (33)	70-75	56.3-62.8 (mean range)	1.65:1 1:0.71	2.8 (6.3)
			Bulbar (20)		61.4	0.98:1	2.0 (2.1-3.4)
			Respiratory (1-5) Axial, head drop (rare)		(as above)	6.00:1 --	1.4^ (0) --
'Atypical' phenotypes:							
Primary lateral sclerosis (PLS)	+++	+/-*	LL (81) Bulbar (13) UL (6)	2-5	45-52 (mean range)	0.98:1	13.1 (71.1)
Progressive muscular atrophy (PMA)	+/-**	+++	UL (47) LL (30) Shoulder girdle (23)	5	61.1	2.04:1	3.3 (11.5) [Similar to classical forms when generalised (>50% regions) ]
Restricted variants							
- Flail arm variant - Flail leg variant	+/-	+++	UL (100) LL (100) [Restricted for ≥12 months]	5-6 3-3.5	57.3 55.0	4.00:1 1.03:1	5.0 (15.8) 3.0-5.7 (5.3-12.8)
Isolated bulbar palsy (IBP)	++	+	Bulbar (100) [Restricted for ≥6 months]	1-4	65-68 (mean range)	1:3.00	3.4

**Supplementary Table 1:** Frequency, demographics and survival of the typical and atypical phenotypes. +++, primary feature; +/-, possible but not typical; + or ++, typical to the indicated degree. \* No 'active' LMN degeneration, but minor denervation may be present on EMG ; \*\* clinical UMN signs can develop within 18months for PMA. UMN, upper motor neurone; LMN, lower motor neurone; UL, upper limb; LL, lower limb; M, male; F, female. N.B: Reported frequencies differ slightly across populations [1-4].^ mean survival given here.