

**Table 1 Maximal active tension and calcium-sensitivity of force generation of nemaline myopathy patients and of control subjects**

Biopsy	Maximal active tension (mN/mm <sup>2</sup> )				Calcium-sensitivity of force generation (pCa <sub>50</sub> )							
	Sarcomere length 2.1 µm				Sarcomere length 2.1 µm				Sarcomere length 2.6 µm			
	Vehicle	n	CK-2066260	n	Vehicle	n	CK-2066260	n	Vehicle	n	CK-2066260	n
<b>Nemaline myopathy patients with nebulin mutations</b>												
26-2   T33	25.6	6	24.9	6	5.80	6	5.87	6	5.93	4	5.91	4
258-2   T1069	17.6	4	18.0	4	5.92	5	6.15	5	5.94	2	6.15	2
974-1   T1033	9.8	4	9.3	4	5.74	5	6.17	5	-	-	-	-
988-1   T887	22.1	5	21.6	5	5.72	5	5.91	5	5.93	5	6.09	5
<b>Pediatric control subjects</b>												
212-1   T141	69.1	6	69.7	6	5.89	6	6.03	6	5.86	1	6.01	1
213-1   T142	71.0	3	71.0	3	5.86	3	5.92	3	-	-	-	-
218-1   T147	110.1	4	103.7	4	5.87	4	6.21	4	5.98	4	6.23	4
219-1   T148	115.0	4	115.0	4	5.90	3	6.02	3	6.05	1	6.11	1
<b>Adult control subjects</b>												
CTRL <sub>slow</sub>	85.8	4	84.9	4	5.84	4	5.86	4	-	-	-	-
CTRL <sub>fast</sub>	103.1	17	100.0	17	5.88	20	6.33	20	5.86	4	6.34	4

N.B. Due to the experimental design, at a sarcomere length of 2.6 µm some muscle preparations had to be excluded with respect to quality criteria. Therefore, the number of experiments per group is reduced at sarcomere length 2.6 µm.