**Supplemental Materials**

**A novel subcutaneous intrathecal catheter system for repeated outpatient dosing of nusinersen: preliminary observations of safety and tolerability**

The weight-adjusted SMA Force Index (SFI) is a novel parameter designed to capture the change in motor power distribution characteristic of SMA. As the disease progresses, proximal and extensor muscle groups weaken while intrinsic muscle strength (e.g. that which allows for digital pinching) is preserved or even increases over time. The SFI incorporates 4 (among 12) dynamometry maneuvers (measured in Newtons, N) with the strongest Pearson correlations to patient age (Table S1), which include shoulder abduction (SA; r = -0.91, r2 = 0.83, p = 0.0006), elbow extension (EE; r = -0.83, r2 = 0.68, p = 0.0061), hand grip (G; r = -0.87, r2 = 0.76, p = 0.0023), and three-point pinch (P; r = 0.94, r2 = 0.88, p = 0.0002). It is calculated from these four measures as follows: **SFI = [(SA + EE + G)/P]0.5**. Indexed to body weight, the SFI correlates strongly with age and natural disease course (r = -0.94, 95% CI -0.74 to -0.99, r2 = 0.89, p = 0.0001).

|  |
| --- |
| **Table S1.** Dynometry correlations with age among six SMA subjects with 3 copies of *SMN2* |
| **Maneuver** | **r** | **95% CI** | **R squared** | **P (two-tailed)** |
| Pinch | 0.94 | 0.7314 to 0.9874 | 0.88 | ***0.0002*** |
| Elbow Flexors | 0.32 | -0.4351 to 0.8124 | 0.10 | 0.3980 |
| Knee Flexors | 0.32 | -0.4378 to 0.8113 | 0.10 | 0.4026 |
| Hip Abductors | 0.28 | -0.4755 to 0.7944 | 0.08 | 0.4728 |
| Wrist Extensors | 0.18 | -0.5525 to 0.7524 | 0.03 | 0.6499 |
| Foot Dorsiflexors | -0.17 | -0.7493 to 0.5574 | 0.03 | 0.6629 |
| Hip Flexors | -0.28 | -0.7972 to 0.4694 | 0.08 | 0.4608 |
| Neck Flexors | -0.33 | -0.8151 to 0.4288 | 0.11 | 0.3872 |
| Knee Extensors | -0.52 | -0.8789 to 0.2254 | 0.27 | 0.1550 |
| Elbow Extensors | -0.83 | -0.9622 to -0.3573 | 0.68 | ***0.0061*** |
| Grip | -0.87 | -0.9722 to -0.4853 | 0.76 | ***0.0023*** |
| Shoulder Abductors | -0.91 | -0.9813 to -0.6243 | 0.83 | ***0.0006*** |