Abstract: Motor symptoms in Huntington's Disease (HD) are commonly assessed by the Unified Huntington's Disease Rating Scale-Total Motor Score (UHDRS-TMS). However, the UHDRS-TMS is limited by interrater variability, its categorical nature, and insensitivity in premanifest subjects. More objective and quantitative measures of motor phenotype may complement the use of the UHDRS-TMS as outcome measure and increase the power and sensitivity of clinical trials. Deficits in tongue protrusion are well acknowledged in HD and constitute a subitem of the UHDRS-TMS. We, therefore, investigated whether objective and quantitative assessment of tongue protrusion forces (TPF) provides measures that (1) correlate to the severity of motor phenotype detected in the UHDRS-TMS in symptomatic HD, (2) detect a motor phenotype in premanifest HD gene-carriers, and (3) exhibit a correlation to the genotype as assessed by a disease burden score (based on CAG-repeat length and age). Using a precalibrated force transducer, the ability of premanifest gene carriers (n = 15) and subjects with symptomatic HD (n = 20) to generate and maintain isometric TPF at three target force levels (0.25, 0.5, and 1.0 N) was assessed and compared with age-matched controls (n = 20) in a cross-sectional study. Measures of variability of TPF and tongue contact time distinguished controls, premanifest, and...
symptomatic HD groups and correlated to the UHDRS-TMS and disease burden score, suggesting a strong genotype-phenotype correlation. Group distinction was most reliable at the lowest target force level. We conclude that assessment of TPF may be a useful objective and quantitative marker of motor dysfunction in premanifest and symptomatic HD.