Gender influence on initial symptoms in Huntington disease

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Background: Huntington disease (HD) is an autosomal dominant neurodegenerative disorder caused by expansion of CAG triplets in the huntingtin gene. The number of CAG repeats directly correlates with the age of onset and severity of the disease. Epidemiological studies have not found significant difference between males and females in overall disease symptoms; however, there is a lack of data regarding the influence of gender on presenting symptoms. Aim of the study: To establish the correlation between gender and the presenting symptoms of HD. Methods: We assessed HD patients treated at the University Hospital Ostrava between 2014 and 2018. The age, gender, allelic composition and initial motor (chorea or parkinsonism) and neuropsychiatric symptoms (behavioral disturbances or cognitive impairment) were recorded. Chi-square test was used to compare initial symptoms in males and females. Results: Out of 29 included HD patients (19 women, mean age 50.5 years, range 21–82 years), motor symptoms were the first symptoms in the HD initial stage in 6 females (32%) and 7 males (70%), neuropsychiatric symptoms in 13 females (68%) and 3 (30%) males (p=0.048). Conclusion: Motor symptoms were significantly more frequent in the initial stage of HD in males whereas neuropsychiatric symptoms were significantly more frequent in females.