Observing Huntington's Disease: the European Huntington's Disease Network's REGISTRY.

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Abstract

BACKGROUND:

Huntington's disease (HD) is a rare triplet repeat (CAG) disorder. Advanced, multi-centre, multi-national research frameworks are needed to study simultaneously multiple complementary aspects of HD. This includes the natural history of HD, its management and the collection of clinical information and biosamples for research.

METHODS:

We report on cross-sectional data of the first 1766 participants in REGISTRY, the European Huntington's Disease Network's (EHDN), multi-lingual, multi-national prospective observational study of HD in Europe. Data collection (demographics, phenotype, genotype, medication, comorbidities, biosamples) followed a standard protocol.

RESULTS:

Phenotype, and the HD genotype, of manifest HD participants across different European regions was similar. Motor onset was most common (48%) with a non-motor onset in more than a third of participants. Motor signs increased, and cognitive abilities and functional capacity declined as the disease burden (CAGn-35.5) X age) increased. A life-time history of behavioural symptoms was common, but the behavioural score was not related to disease burden. One fifth of participants had severe psychiatric problems, e.g. suicidal ideation and attempts, and/or irritability/aggression, with psychosis being less common. Participants on anti-dyskinetic medication had a higher motor and lower cognitive score, were older, and more prone to physical trauma. A higher motor and a lower cognitive score predicted more advanced disease.

CONCLUSIONS:

The unparalleled collection of clinical data and biomaterials within the EHDN's REGISTRY can expedite the search for disease modifiers (genetic and environmental) of age at onset and disease progression that could be harnessed for the development of novel treatments.