Huntington's in Youth Closely Tied to Problems With School and Social Life

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Marta Figueiredo September 15, 2020

Behavioral changes due to early onset Huntington's disease and problems with school, and social and family life are common, particularly among pediatric patients, according to a case series study in Germany.

Socio-medical problems in these children were associated both with their own disease, and with the additional burden of parents with early onset Huntington's and/or an unstable family background.

The study, "Clinical Manifestation of Juvenile and Pediatric HD Patients: A Retrospective Case Series," was published in the journal *Brain Sciences*.

While Huntington's symptoms usually first appear in people between the ages of 30 and 50, disease onset can occur in those under 21 years old, and is identified as juvenile-onset disease.

However, researchers at the European Huntington Disease Network recently suggested that the term "pediatric" Huntington's be used for patients with disease onset before age 18, as the "juvenile-onset" definition can be blurred and is used in different ways.

"Although many important aspects about juvenile HD [Huntington's disease] have been described, no case reports about the recently defined pediatric [group] or comparing research of pediatric and juvenile HD in the boundary of typical characteristics in adult HD were described," the researchers wrote.

More research on the social-medical aspects of the disease is also needed, they said. A deeper understanding may help in developing better support systems and treatment approaches for both patients and their families.

Researchers at Ruhr-University Bochum retrospectively analyzed clinical, demographic, and socio-medical data on people with early onset Huntington's.

Of the 2,593 patients treated at their Huntington center in the last 25 years, 32 had disease onset before age 21 (1.23%), and 18 before the age of 18 (0.69%). This was consistent with the estimated frequency of juvenile-onset disease: 1%–9.6% of all Huntington's patients, the researchers noted.

The team divided these 32 patients into two groups according to their age at onset: before 18 years old (pediatric; 13 boys and five girls), and between 18 and 21 years old (juvenile; six males and eight females).

The mean age at symptom onset was 10.3 years for the pediatric group, and 19.3 years for the juvenile group. On average, about three years separated symptom onset and a Huntington's diagnosis in both groups.

Available family history data for 23 patients showed that most (69.6%) inherited the disease from their father. Clinical characteristics were missing from two patients with pediatric Huntington's and five in the juvenile group, and they were not considered in further analyses.

Compared with the juvenile group, more pediatric patients had muscle rigidity (69% vs. 56%), tremor (37.5% vs. 11.1%), and dystonia or involuntary muscle spasms (37.5% vs. 22.2%). In turn, chorea, or brief and abrupt involuntary movements, was more common among the juvenile group (55.6%) than in the pediatric group (12.5%).

Epilepsy was present in more than one-third (37.5%) of pediatric patients, and in 22.2% of older patients.

Aggression and irritability — behavioral and psychiatric issues — were frequently reported in both the pediatric (62.5%) and juvenile (33.3%) groups, as well as suicidal ideations or attempts, which occurred in about one-third of patients in both groups.

Children and adolescents with Huntington's also showed a high degree of school problems, which were frequently initial disease symptoms and accompanied cognitive decline. Serious problems concerning the social and family background, including attempted rape, robbery and family conflicts, were reported in 25% of this pediatric group.

Alcohol and drug abuse were also reported in two pediatric patients and one juvenile patient. One boy showed criminal behavior, possibly due to disease-related poor impulse control as well and "an additional impact of the social surrounding, or even further developments of the common puberty-age with a personal-development process," the researchers wrote.

"Beside[s] abnormalities in behavior from an early age due to HD pathology, children seem to have higher socio-medical problems related to additional burden caused by early affected parents, instable family backgrounds including drug abuse of a parent or multiple changes of partners," the researchers wrote.

These young patients were treated with dopaminergic medications for slowness of movement, benzodiazepines and Xenazine for dystonia, and cannabinoids, botulinum toxin injections, and deep brain stimulation for movement disorders. Clozapine was used to treat tremor and dopaminergic therapy-induced hallucinations, and zuclopenthixol for the treatment of severe aggressive behavior.

In many cases, treatment required individualized approaches.

Based on their experience, the research team emphasized that almost all of these children were more relieved by the diagnosis than burdened, as it finally explained their existing problems with school, sports, or friends.

“Our impression is, that children, as in other severe diagnoses like cancer, seem to cope with the diagnosis well in most cases whereas diagnosis is often more difficult for the parents,” the researchers wrote.

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