Huntington's Disease Society of America

COMPASSIONATE ALLOWANCE INFORMATION

DI 23022.923 Adult Onset Huntington Disease

Compassionate Allowance is a way to quickly identify diseases and other medical conditions that, by definition, meet Social Security's standards for disability benefits. The CAL initiative helps Social Security reduce waiting time to reach a disability determination for individuals with the most serious disabilities.

When completing a disability application for HD, always request compassionate allowance: ***Please flag my claim for CAL processing per DI 23022.923 because I have Huntington's Disease.***

ADULT ONSET HUNTINGTON DISEASE	
ALTERNATE NAMES	Huntington's chorea; Huntington's Disease
DESCRIPTION	**Huntington disease (HD)** is a hereditary neurodegenerative disorder that is characterized by progressively worsening motor, cognitive, behavioral, and psychiatric symptoms. HD is caused by a mutation of the Huntington gene called a "CAG repeat expansion". The mutation results in gradual neuronal degeneration in the basal ganglia of the brain, and progresses to involve other regions of the brain responsible for coordination of movements, thoughts, and emotions. Neuronal degeneration causes diffuse and severe brain atrophy that is comparable to late stage Alzheimer disease. Clinical presentation of HD may include changes in personality, behavior, cognition, speech, and coordination. Physical changes include random uncoordinated extremity movements (chorea), rigidity, leg stiffness, clumsiness, slowness of movement, tremors and muscle spasms. As the disease progresses, concentration on cognitive tasks becomes increasingly difficult, and an individual may have difficulty swallowing and feeding himself. Family history of HD is usually but not always positive.
DIAGNOSTIC TESTING, PHYSICAL FINDINGS, AND ICD-9-CM CODING	The diagnosis of HD is made by clinical history documenting changes in motor, behavioral and cognitive function; family history of HD; abnormal neurologic exam findings; abnormal neuropsychological test results; and HD gene test with abnormal results (40 or more CAG repeats). Brain imaging is optional, but if performed may show atrophy of the caudate nucleus or diffuse brain atrophy. ICD-9 code: 333.4

ONSET AND PROGRESSION	The average onset age is around 40, plus or minus 10 years; however, onset has been documented as young as age 5 (see Juvenile HD) and as old as age 90. Death usually occurs at about 15 to 20 years after onset of symptoms, and is due to complications of the disease.
TREATMENT	There is no cure or treatment to stop, slow or reverse the progression of HD. Claimant's medical source(s) may prescribe medications to manage symptoms. A psychiatrist or behavior management specialist may address behavior disorders. A speech language pathologist may evaluate communication and swallowing problems. A nutritionist may be consulted to address nutritional needs as the disease progresses. Assistive devices such as wheelchairs, helmets, and communication boards may be used for safety, and to improve quality of life.

SUGGESTED PROGRAMMATIC ASSESSMENT*

Suggested MER for evaluation:

- Claimant's medical source(s) records documenting progression of motor, cognitive, and psychiatric symptoms and signs; family history of HD; and abnormal neurological exam findings consistent with HD.
- Laboratory testing showing a CAG repeat in the HD gene (40 or more CAG repeats).
- Brain imaging may provide supporting evidence.
- Psychological or psychiatric reports including neurocognitive testing.

Suggested Listings for Evaluation:

DETERMINATION	LISTING	REMARKS
Meets Listing	11.17 or 12.02	Listing level neurological and/or cognitive findings must be documented; **diagnosis of HD or laboratory testing results alone do not meet listing severity.**

* Adjudicators may, at their discretion, use the Medical Evidence of Record or Listings suggested to evaluate the claim. However, the decision to allow or deny the claim rests with the adjudicator.

To Link to this section - Use this URL: http://policy.ssa.gov/poms.nsf/lnx/0423022923