

The Department of Vermont Health Access Medical Criteria

Subject: Huntington's Disease, Genetic Testing

Last Review: February 10, 2023*

Past Revisions: September 3, 2021, July 6, 2017, April 25, 2016, February 20, 2015

***Please note: Most current content changes will be highlighted in yellow.**

Description of Service or Procedure

Huntington disease (HD) is a brain disorder that affects a person's ability to think, talk, and move. It presents itself in three groups of symptom features: movement (chorea), psychiatric/behavioral and cognitive disorders. It is an inherited autosomal dominant condition meaning that each child of an affected parent, regardless of gender, has a 50% chance of inheriting the disease-causing gene. Some early clinical symptoms include dementia, mood swings, depression, lack of coordination, irritability, trouble driving, or learning new things. They may also have difficulty remembering a fact or making a decision. As the disease progresses, concentrating and performing task becomes increasingly more difficult. The symptoms and severity of the disease vary between individuals as well as the age of onset.

The typical onset of HD symptoms is between ages 30-50. However, onset of symptoms has been seen in persons as young as 5 years or as old as 90 years. HD is associated with an expansion of the tandem repeat of the DNA nucleotides sequence cytosine, adenine, and guanine (CAG) in the Huntington (HTT) gene. Analysis of the CAG repeat sequence via genetic testing is useful in three clinical situations:

- 1) Confirmation of a suspected diagnosis of HD
- 2) Predictive testing in an asymptomatic individual known to be at-risk for carrying the gene
- 3) Prenatal testing.

CAG REPEAT SIZE INTERPRETATION

26 or less is normal

27-35 is normal but potentially unstable

36-39 is abnormal with variable penetrance; unstable

40 and above Huntington Disease

Disclaimer

Coverage is limited to that outlined in Medicaid Rule or Health Care Administrative Rules that pertains to the member's aid category. Prior Authorization (PA) is only valid if the member is eligible for the applicable item or service on the date of service.



Medicaid Rule

Medicaid and Health Care Administrative Rules can be found at <https://humanservices.vermont.gov/rules-policies/health-care-rules/health-care-administrative-rules-hcar/adopted-rules>

- 7102.2 Prior Authorization Determination
- 7405 Laboratory and Radiology Services
- 4.101 Medical Necessity for Covered Services
- 4.104 Medicaid Non-Covered Services

Coverage Position

A genetic test for Huntington's disease may be covered for members:

- When the test is prescribed by a licensed medical provider, enrolled in the Vermont Medicaid program, operating within their scope of practice as described on the Vermont's Office of Professional Regulation's website*, Statute, or rule who is knowledgeable regarding genetic testing, and who provides medical care to the member AND
- When the clinical criteria below are met.

* Vermont's Office of Professional Regulation's website: <https://sos.vermont.gov/opr/>

Coverage Criteria

Genetic testing for Huntington's disease may be covered for members based on the following:

- Adults:
 - To confirm or rule out the diagnosis when symptoms strongly suggest the diagnosis of Huntington's disease but there is no known family history.
 - Testing in asymptomatic relatives of a person with a genetically confirmed diagnosis of HD
- Children:
 - When symptoms are suggestive, and Huntington's disease is suspected based on family history and clinical phenotype. Testing of a child with the clinical phenotype, in the absence of a family history, should be done after ruling out other causes. It is not appropriate to presymptomatically test children < 18 years of age, when a parent has been diagnosed with adult-onset Huntington's disease.
- Presymptomatic testing for an individual at risk to provide reproductive and recurrence information.
- Prenatal testing in fetuses from families in which there is a history of HD and
 - The results will directly impact the treatment plan of the member.
 - Genetic counseling has been conducted.

Exclusions: Genetic testing for HD for routine screening.

Considerations: Providers requesting this test should provide pre- and post-test genetic counseling for the member and family, if applicable.

Early and Periodic Screening, Diagnostic and Treatment (EPSDT) exception: Vermont Medicaid will provide comprehensive services and furnish all Medicaid coverable, appropriate, and medically necessary services needed to correct and ameliorate health conditions for Medicaid members under age 21.

Please note, Vermont Medicaid Clinical Criteria is reviewed based on available literature, evidence-based guidelines/standards, Medicaid rule and policy, and Medicare coverage determinations that may be appropriate to incorporate when applicable.

Clinical criteria for repeat service or procedure

Once per lifetime.

Type of service or procedure covered

Genetic testing for Huntington's Disease.

References

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