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The Department of Vermont Health Access Medical Criteria

Subject: Huntington's Disease, Genetic Testing
Last Review: September 3, 2021*
Past Revisions: 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 and below Normal
27-35 Normal but potentially unstable
36-39 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 beneficiary's aid category. Prior Authorization (PA) is only valid if the beneficiary 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
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 beneficiaries:

- 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*, who is knowledgeable regarding genetic testing, and who provides medical care to the beneficiary 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 beneficiaries 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.
- 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 in order to provide reproductive and recurrence information.

And

- The results will directly impact the treatment plan of the beneficiary.
- Genetic counseling has been conducted.

Considerations: Providers requesting this test should provide pre- and post-test genetic counseling for the beneficiary 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.

References

Barboza, L. A., & Ghisi, N. C. (2018). Evaluating the current state of the art of Huntington disease research: A scientometric analysis. Brazilian Journal of Medical and Biological Research, 51(3), e6299. https://doi.org/10.1590/1414-431X20176299

Centers for Medicare and Medicaid Services. *Early and periodic screening, diagnostic, and treatment.* Retrieved August 22, 2019, from: https://www.medicaid.gov/medicaid/benefits/epsdt/index.html

Huntington's Disease Society of America. (2016). *Genetic testing protocol for Huntington's disease*. Retrieved from: http://hdsa.org/wp-content/uploads/2015/02/HDSA-Gen-Testing-Protocol-for-HD.pdf

Bachoud-Lévi, A.C., Ferreira J., Massart, R., Youssov, K., Rosser, A., Busse, M., … Burgunder, J-M. (2019). International guidelines for the treatment of Huntington's disease. *Frontiers in Neurology, 10.* https://doi.org/10.3389/fneur.2019.00710

Johnson, A.C. & Paulsen, J.S. (2014). *Understanding behavior in Huntington's disease.* Huntington's Disease Society of America. Retrieved from: http://hdsa.org/wp-content/uploads/2015/03/Understanding-Behavior.pdf

U.S. National Library of Medicine, National Institutes of Health, MedlinePlus. (2020). *Huntington disease.* Retrieved from: https://medlineplus.gov/genetics/condition/huntington-disease/

Mayo Clinic. (2014). *Huntington's disease.* Retrieved from: http://www.mayoclinic.org/diseases-conditions/huntingtons-disease/basics/tests-diagnosis/con-20030685

Myers, R. H. (2004). Huntington's disease genetics. *The Journal of the American Society for Experimental NeuroTherapeutics, 1*(2), 255-262. doi: 10.1602/neurorx.1.2.255

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