

**LETTER OF MEDICAL NECESSITY**

**Ataxia Common Repeat Expansion Evaluation**

**Instructions for Healthcare Provider:**

1. This letter template is being provided as a tool for clinicians to assist communication with payers.
2. Include specific patient information in the letter for this tool to be effective. The areas that must be edited/deleted are indicated in gray on the template
3. Print the template on the physician’s letterhead, **NOT** Athena letterhead. There should be no Athena branding on the letter.

[LMN: Ataxia Common Repeat Expansion (10.18.2021)]

<Date>

ATTN: <Medical Director/ Physician Name>, MD

 <Institution/Insurance Company>

  <Street Address>

 <City>**,** <State> <zip code>

RE: <Patient Name>

DOB: <MM/DD/YYYY>

Member ID: <Insurance ID Number>

Group #: <Enter Group #>

Dear Doctor <Medical Director/ Physician Name>:

I am writing this letter on behalf of my patient <Patient Name> to request coverage for the Ataxia, Common Repeat Expansion Evaluation, which analyzes genes for repeat expansion mutations associated with the 8 most common spinocerebellar ataxias (SCAs 1, 2, 3, 6, 7, 8, 10, and 17). This letter documents the medical necessity for *Ataxia, Common Repeat Expansion Evaluation*, in light of my patient’s medical history. Results from the test will be used to guide appropriate medical care for my patient.

I have determined that this test is medically necessary because of the following aspects of this patient’s history:

<Patient name> is a <age>-year old <gender> with a suspected diagnosis of hereditary spinocerebellar ataxia. Symptoms and clinical findings are consistent with this diagnosis:

1. <Symptom #1 with ICD-10 code>

2. <Symptom #2 with ICD-10 code>

<Symptoms should support diagnosis or risk of genetic disease. Relevant information may include results of prior testing, such as an MRI scan and tests for hypothyroidism, and results of a physical examination and patient consultation.>

<Note family or other personal history if relevant. Consider including information on both neurological and non-neurological problems, such as movement disorders, spasticity, peripheral neuropathy, intellectual impairment, etc.>

**Rationale for Testing**

Hereditary ataxias compose a group of diseases characterized by incoordination of speech and movement.1 Obtaining a specific diagnosis is complex because genetic causes are highly heterogeneous and clinical symptoms frequently overlap among these diseases.1-3 Hereditary ataxias are broadly classified by mode of inheritance, predominantly as AD or AR ataxias, but confirming a diagnosis for a specific type of ataxia is difficult without a molecular diagnosis obtained through genetic testing.1

The presence of a family history warrants genetic testing for specific genes (eg, genes associated with AD ataxias) to identify or confirm the pathogenic variant.4 When an AD cerebellar ataxia is suggested based on family history, the European Federation of the Neurological Societies (EFNS) recommends testing for spinocerebellar ataxia (SCA) types 1, 2, 3, 6, 7, and 17 (and DRPLA in Asian patients).5 Similarly, Ataxia UK, a patient support organization that developed guidelines in consultation with >30 health professionals, recommends initial testing for the most common ataxias that includes SCA 1, 2, 3, 6, and 7 (and optionally SCAs 12 and 17).6 SCAs 1, 2, 3, 6, and 7 represent approximately 60% of the AD ataxias.2 Thus, initial testing with a panel that tests for these ataxias, such as the 8-gene *Ataxia, Common Repeat Expansion Evaluation*, would be the initial step in establishing a diagnosis based on guideline recommendations.

Identifying the specific genetic alteration is important because the SCA subtype helps inform a patient’s prognosis. For instance, some types (eg, SCA1) are associated with more rapid disease progression, while others (eg, SCA6) have a more stable disease course.1,7 In addition, certain subtypes of SCA are associated with distinctive clinical features, such as cognitive decline (SCA17), neuromuscular disorders (SCAs 2, 3, and 10), and vision problems (SCAs 6 and 7).1,2,8,9

Therefore, identification of a repeat expansion can provide resolution of the diagnostic odyssey and help patients and healthcare providers prepare for appropriate interventions, such as treatment, genetic counseling, life and family planning, and enrollment in support groups and research activities.4,10 For example, treatment with valproic acid (an anticonvulsant) or varenicline (a smoking cessation aid) may potentially improve symptoms of SCA3 such as gait and stance.5,11 Prognostic information can inform coordination of allied healthcare interventions (eg, physical and occupational therapy), which has demonstrated efficacy in the management of cerebellar ataxia and is highly individualized depending on symptoms.5,12 Therefore, a diagnosis acquired through genetic testing brings awareness of specific disease characteristics that are likely to occur, allowing for appropriate planning for disease management.

In summary, initial testing with Ataxia, Common Repeat Expansion Evaluation is guideline supported and improves the clinical management of patients with common SCA repeat expansions. Therefore, I am requesting that <Patient Name> be approved for the Ataxia, Common Repeat Expansion Evaluation (Test Code 6901; see CPT codes below\*) offered by Athena Diagnostics®.

I hope you will support this letter of medical necessity for <Patient Name>. Please feel free to contact me at <Physician Phone> if you have additional questions.

Sincerely,

<Physician Name>, MD

NPI #: <Physician NPI#>

Contact information:

< Address>

<City>**,** <State>, <zip code>

Contact Phone: <phone number>

\*CPT codes: 81177(1), 81178(1), 81179(1), 81180(1), 81181(1), 81182(1), 81184(1), 81344(1). The CPT codes provided are based on AMA guidelines and are for informational purposes only. CPT coding is the sole responsibility of the billing party. Please direct any questions regarding coding to the payer being billed.

**References**

**1.** Jayadev S, Bird TD. Hereditary ataxias: overview. *Genet Med*. 2013;15(9):673-683. doi:10.1038/gim.2013.28

**2.** Sandford E, Burmeister M. Genes and genetic testing in hereditary ataxias. *Genes (Basel)*. 2014;5(3):586-603. doi:10.3390/genes5030586

**3.** Sailer A, Houlden H. Recent advances in the genetics of cerebellar ataxias. *Curr Neurol Neurosci Rep*. 2012;12(3):227-236. doi:10.1007/s11910-012-0267-6

**4.** Ashizawa T, Xia G. Ataxia. *Continuum (Minneap Minn)*. 2016;22(4 Movement Disorders):1208-1226. doi:10.1212/CON.0000000000000362

**5.** van de Warrenburg BP, van Gaalen J, Boesch S, et al. EFNS/ENS Consensus on the diagnosis and management of chronic ataxias in adulthood. *Eur J Neurol*. 2014;21(4):552-562. doi:10.1111/ene.12341

**6.** de Silva R, Greenfield J, Cook A, et al. Guidelines on the diagnosis and management of the progressive ataxias. *Orphanet J Rare Dis*. 2019;14(1):51. doi:10.1186/s13023-019-1013-9

**7.** Tezenas du Montcel S, Charles P, Goizet C, et al. Factors influencing disease progression in autosomal dominant cerebellar ataxia and spastic paraplegia. *Arch Neurol*. 2012;69(4):500-508. doi:10.1001/archneurol.2011.2713

**8.** Shakkottai VG, Fogel BL. Clinical neurogenetics: autosomal dominant spinocerebellar ataxia. *Neurol Clin*. 2013;31(4):987-1007. doi:10.1016/j.ncl.2013.04.006

**9.** Wallace SE, Bird TD. Molecular genetic testing for hereditary ataxia: what every neurologist should know. *Neurol Clin Pract*. 2018;8(1):27-32. doi:10.1212/CPJ.0000000000000421

**10.** Orengo JP, Murdock DR. Genetic testing in neuromuscular disorders. *Pract Neurol*. 2019;July/August:35-41.

**11.** Zesiewicz TA, Wilmot G, Kuo SH, et al. Comprehensive systematic review summary: Treatment of cerebellar motor dysfunction and ataxia: Report of the Guideline Development, Dissemination, and Implementation Subcommittee of the American Academy of Neurology. *Neurology*. 2018;90(10):464-471. doi:10.1212/WNL.0000000000005055

**12.** Fonteyn EM, Keus SH, Verstappen CC, et al. The effectiveness of allied health care in patients with ataxia: a systematic review. *J Neurol*. 2014;261(2):251-258. doi:10.1007/s00415-013-6910-6