

Coverage of any medical intervention discussed in a Prevea360 Health Plan medical policy is subject to the limitations and exclusions outlined in the member's benefit certificate or policy.

Genetic Testing for Neurologic Disorders

MP9497

Covered Service: Yes—when meets criteria below

Prior Authorization

Required: Yes—as shown below

Additional Information:

Pre- and post-test genetic counseling is required for any individual undergoing genetic testing for neurological disorders.

A first-degree relative is defined as an individual's parents, full siblings, and children.

A second-degree relative is defined as an individual's grandparents, grandchildren, aunts, uncles, nephews, nieces and half-siblings.

*Axonal neuropathy indicates etiology is related to diabetes, toxic medications, or thyroid disease.

Prevea360 Health Plan Medical Policy:

- 1.0 **Genetic Testing** for hereditary neurologic disorders **requires** prior authorization through the Health Services Division and is considered medically necessary and must meet **ALL** of the following criteria:
 - 1.1 The member displays clinical features, or is at direct risk of inheriting the mutation in question (pre-symptomatic); **AND**;
 - 1.2 The result of the test will directly impact the treatment being delivered to the member; **AND**;
 - 1.3 After history, physical examination, pedigree analysis, genetic counseling, and completion of conventional diagnostic studies a definitive diagnosis remains uncertain or identification of a genetic mutation will guide reproductive decision making.
- 2.0 Please reference the following links for specific criteria requirements for:
 - 2.1 **Huntington Disease** - [Genetic Testing for Huntington Disease MP9490](#)
 - 2.2 **Spinal Muscular Atrophy** - [Genetic Testing for Reproductive Carrier Screening and Prenatal Care MP9477](#)
- 3.0 **Alzheimer Disease (Early Onset) APP, PSEN1, PSEN2** genetic testing **requires** prior authorization through the Health Services Division and is considered medically necessary for the diagnosis or screening for Alzheimer disease if **1 or more** of the following are met:

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- 3.1 Early-onset familial Alzheimer disease, suspected, as indicated by **ALL** of the following:
 - 3.1.1 Dementia diagnosed in patient 60 years or younger and has **1 or more** of the following:
 - 3.1.1.1 Documented mutation of PSEN1, PSEN2, or APP gene in relative; **OR**;
 - 3.1.1.2 Family history of dementia; **OR**;
 - 3.1.1.3 Unknown family history of dementia.
 - 3.1.2 Reversible causes of dementia have been excluded by clinical examination, neuroimaging, and laboratory testing.
- 3.2 Predictive testing for at-risk asymptomatic adult if **BOTH** of the following are met:
 - 3.2.1 Disease causing mutation in PSEN1, PSEN2, or APP gene has been identified in affected first-degree or second-degree relative; **AND**;
 - 3.2.2 Testing is preceded by baseline neurologic examination.
- 4.0 **Ataxia – Telangiectasia ATM** genetic testing **requires** prior authorization through the Health Services Division and is considered medically necessary when **BOTH** of the following is present:
 - 4.1 Acquired causes of ataxia have been ruled out; **AND**;
 - 4.2 Diagnosis or screening for ataxia-telangiectasia, as indicated by **1 or more** of the following:
 - 4.2.1 ATM protein or ATM protein kinase activity equivocal or indeterminate; **OR**;
 - 4.2.2 Carrier testing for patient with family history of ataxia-telangiectasia; **OR**;
 - 4.2.3 Confirmed diagnosis, and need to establish disease-causing mutation; **OR**;
 - 4.2.4 Prenatal diagnosis, when disease-causing mutation in ATM gene has been identified in one or both prospective parents.
- 5.0 **CADASIL (Cerebral Autosomal Dominant Arteriopathy with Subcortical Infarcts and Leukoencephalopathy) NOTCH3** genetic testing **requires** prior authorization through the Health Services Division and is considered medically necessary for **1 or more** of the following indications:
 - 5.1 Asymptomatic individual 18 years or older, when disease-causing mutation has been confirmed in an affected relative; **OR**;

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- 5.2 Symptomatic individuals who have a family history consistent with an autosomal dominant pattern of inheritance of this condition (clinical signs and symptoms of CADASIL include stroke, cognitive defects and/or dementia, migraine, and psychiatric disturbances); **OR**;
- 5.3 Confirmation of diagnosis in patient with clinical features and neuroimaging findings suggestive of CADASIL, irrespective of age.
- 6.0 Charcot-Marie-Tooth Hereditary Neuropathy** – Genetic testing for the diagnosis of CMT Hereditary Neuropathy **requires** prior authorization through the Health Services Division and is considered medically necessary if **BOTH** of the following are met:
- 6.1 Clinical findings are suggestive of Charcot-Marie-Tooth (CMT) hereditary neuropathy, as indicated by **1 or more** of the following:
- 6.1.1 Diminished tendon reflexes; **OR**;
 - 6.1.2 Distal muscle atrophy; **OR**;
 - 6.1.3 Distal sensory loss; **OR**;
 - 6.1.4 Foot drop; **OR**;
 - 6.1.5 High-arched feet (pes cavus deformity) ; **OR**;
 - 6.1.6 Palpably enlarged nerves (e.g. ulnar nerve at olecranon groove, greater auricular nerve along lateral aspect of neck) ; **OR**;
 - 6.1.7 Progressive weakness of hands and feet; **OR**;
 - 6.1.8 Weak ankle dorsiflexion; **AND**;
- 6.2 The patient has had abnormal electromyography and nerve conduction studies, and consistent with axonal neuropathy or demyelinating neuropathy and **EITHER** of the following:
- 6.2.1 The requested genes are targeted to the specific phenotype;
 - 6.2.2 There is a family history of the condition and **one** of the following:
 - 6.2.2.1 Testing has not been performed in another affected family member, and the requested test is targeted to genes associated with the suspected inheritance pattern;
 - 6.2.2.2 The requested test is targeted to the known familial mutation(s).
- 7.0 Familial Dysautonomia IKBKAP** genetic testing **requires** prior authorization through the Health Services Division and is considered medically necessary for the diagnosis or screening for familial dysautonomia, as indicated by **1 or more** of the following:
- 7.1 Carrier testing for **1 or more** of the following:
- 7.1.1 Individual of Ashkenazi Jewish ancestry and of reproductive age;

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- 7.1.2 Individual with first- or second-degree family history of familial dysautonomia.
- 7.2 Clinical findings suggestive of familial dysautonomia, including **ALL** of the following:
 - 7.2.1 Absence of axon flare response after intradermal histamine injection; **AND**
 - 7.2.2 Absence of overflow tears with emotional crying; **AND**
 - 7.2.3 Decreased or absent deep tendon reflexes; **AND**
 - 7.2.4 Decreased taste and absence of fungiform papillae of tongue on visual inspection; **AND**
 - 7.2.5 Hypotonia in infancy; **AND**
 - 7.2.6 Pupillary hypersensitivity to parasympathomimetic agents (e.g. topical methacholine or pilocarpine).
- 8.0 **Friedreich Ataxia (FXN)** genetic testing **requires** prior authorization through the Health Services Division and is considered medically necessary for the diagnosis or carrier testing for Friedreich ataxia when **1 or more** of the following are present:
 - 8.1 Carrier testing in at-risk relative when disease-causing FXN gene mutation has been identified in family; **OR**;
 - 8.2 Carrier testing in reproductive partner of known FXN mutation carrier; **OR**;
 - 8.3 Confirmation of diagnosis in patient with clinical findings suggestive of Friedreich ataxia (e.g. fatigue, aggressive scoliosis, loss of coordination in arms and legs).
- 9.0 **Duchene and Becker Muscular Dystrophy (DMD Gene)** genetic testing **requires** prior authorization through the Health Services Division and is considered medically necessary when **ANY** one of the following criteria is met:
 - 9.1 Carrier screening when the individual to be tested is an asymptomatic female and has an affected blood relative in whom a disease-causing DMD or BMD mutation has been identified; **OR**;
 - 9.2 Carrier screening when the individual to be tested is an asymptomatic female with a male relative who has been clinically diagnosed with Duchenne or Beck Muscular Dystrophy, who is either deceased or unavailable for genetic testing; **OR**;
 - 9.3 Individual to be tested exhibits characteristic features of DMD or BMD (e.g. progressive symmetric muscular weakness (proximal greater than distal) often with enlargement of calf muscles, wheelchair dependency before 13 years of age for DMD and after 16 years of age for BMD); and individual has elevated serum creatine kinase (CK) concentration. * In males with DMD, serum CK

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levels are >10 times normal and in BMD at 5 times normal. Some female carriers of DMD or BMD have levels 2 to 10 times normal.

10.0 **Nemaline Myopathy ACTA1, CFL2, KBTBD13, KLHL40, KLHL41, LMOD3, NEB, TNNT1, TPM2 and TPM3** genetic testing **requires** prior authorization through the Health Services Division and is considered medically necessary for diagnosis or screening for nemaline myopathy when **1 or more** of the following criteria are met:

10.1 Carrier testing for **1 or more** of the following:

10.1.1 For NEB gene mutations: individual of Ashkenazi Jewish ancestry and of reproductive age; **OR**;

10.1.2 For TNNT1 gene mutations: individual of Old Order Amish ancestry and of reproductive age; **OR**;

10.1.3 Individual with family history of nemaline myopathy.

10.2 Clinical findings are suggestive of nemaline myopathy and equivocal findings on muscle biopsy; **OR**;

10.3 Need to establish disease-causing mutation in patient with confirmed diagnosis; **OR**;

10.4 Screening of parent of affected individual with ACTA1, KBTBD13, TPM2 or TPM3 gene mutation and no known family history.

11.0 **Seizure Disorders, Hereditary SCN1A** genetic testing **requires** prior authorization through the Health Services Division and is considered medically necessary for the diagnosis of SCN1A-related seizure disorder or screening of at-risk relatives, as indicated by **either** of the following:

11.1 Confirmation of SCN1A-related seizure disorder in individual with clinical suspicion of **1 or more** of the following:

11.1.1 Dravet syndrome;

11.1.2 Intractable childhood epilepsy with generalized tonic-clonic seizures;

11.1.3 Severe infantile multifocal epilepsy.

11.2 Evaluation of asymptomatic parent of proband with pathogenic SCN1A mutation and no known history of other affected family members.

12.0 **Spinocerebellar Ataxia (SCA) ATXN1, ATXN2, ATXN3, ATXN7 and CACNA1A** genetic testing **requires** prior authorization through the Health Services Division and is considered medically necessary to aid in the diagnosis of SCA when **both** of the following criteria are met:

12.1 Individual to be tested exhibits signs and symptoms of SCA such as progressive gait and limb incoordination, imbalance, dysarthria and disturbances of eye movements; **AND**;

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- 12.2 Non-genetic causes of ataxia have been excluded (e.g. alcoholism, multiple sclerosis, primary or metastatic tumors or paraneoplastic diseases associated with occult carcinoma of the ovary, breast or lung, vascular disease, vitamin deficiencies).
- 13.0 Spinal Muscular Atrophy SMN1 and SMN2 genetic testing requires prior authorization through the Health Services Division and is considered medically necessary when **ANY** of the following criteria are met:
- 13.1 Carrier screening in at-risk individuals, as indicated by **1 or more** of the following:
- 13.1.1 Parent of sibling of reproductive age of deceased child with suspected spinal muscular atrophy without molecular genetic testing confirmation; **OR**;
 - 13.1.2 Parent or sibling of reproductive age of one or more children with spinal muscular atrophy confirmed by molecular genetic testing.
- 13.2 Confirm or establish diagnosis in individual suspected of having spinal muscular atrophy.
- 14.0 **Non-Covered Tests:** The following tests are considered experimental and investigational and therefore are not medically necessary:
- 14.1 Alzheimer Disease (Late Onset) – APOE Genotyping
 - 14.2 Autism Spectrum Disorders – Multi-Gene Panels
 - 14.3 Familial Frontotemporal Dementia - C9orf72, GRN and MAPT Genes
 - 14.4 Parkinson Disease - ATP13A2, GBA, LRRK2, MAPT, PARK2, PARK7, PINK1 and SNCA Genes
 - 14.5 Epilepsy/Seizure Disorders (Hereditary) – Multi-Gene Panels
 - 14.6 Sensory-Motor Neuropathy – Multi-Gene Panels
 - 14.7 Hereditary Spastic Paraplegia – Multi -Gene Panels
 - 14.8 Whole genome/Exome Sequencing for Neuro Disorders (or multigene panels that are not targeted to a specific condition)
 - 14.9 Amyotrophic Lateral Sclerosis (ALS) – SOD1 Gene all other genes;
 - 14.10 Oculopharyngeal Muscular Dystrophy (OPMD) – PABPN1
- 15.0 All other indications not listed above are considered investigational and experimental and therefore are not medically necessary.

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CPT/HCPCS Codes Related to MP9497

* The list of codes (and their descriptors, if any) is provided for informational purposes only and may not be all inclusive or current. Listing of a code in this medical policy does not imply that the service described by the code is a covered or non-covered service. Benefit coverage for any service is determined by the member's policy of health coverage with Prevea360 Health Plan. Inclusion of a code above does not imply any right to reimbursement or guarantee claim payment. Other medical policies may also apply.

81161	DMD (dystrophin) (e.g., Duchenne/Becker muscular dystrophy) deletion analysis, and duplication analysis, if performed
81229	Cytogenomic constitutional (genome-wide) microarray analysis; interrogation of genomic regions for copy number and single nucleotide polymorphism (SNP) variants for chromosomal abnormalities
81243	FMR1 (fragile X mental retardation 1) (e.g., fragile X mental retardation) gene analysis; evaluation to detect abnormal (e.g., expanded) alleles
81260	IKBKAP (inhibitor of kappa light polypeptide gene enhancer in B-cells, kinase complex-associated protein) (e.g., familial dysautonomia) gene analysis, common variants (e.g., 2507+6T>C, R696P)
81302	MECP2 (methyl CpG binding protein 2) (e.g., Rett syndrome) gene analysis; full sequence analysis
81304	MECP2 (methyl CpG binding protein 2) (e.g., Rett syndrome) gene analysis; duplication/deletion variants
81321	PTEN (phosphatase and tensin homolog) (e.g., Cowden syndrome, PTEN hamartoma tumor syndrome) gene analysis; full sequence analysis
81324	PMP22 (peripheral myelin protein 22) (e.g., Charcot-Marie-Tooth, hereditary neuropathy with liability to pressure palsies) gene analysis; duplication/deletion analysis
81325	PMP22 (peripheral myelin protein 22) (e.g., Charcot-Marie-Tooth, hereditary neuropathy with liability to pressure palsies) gene analysis; full sequence analysis
81326	PMP22 (peripheral myelin protein 22) (e.g., Charcot-Marie-Tooth, hereditary neuropathy with liability to pressure palsies) gene analysis; known familial variant
81400	Molecular pathology procedure level 1
81401	Molecular pathology procedure level 2
81403	Molecular pathology procedure level 4

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81404	Molecular pathology procedure level 5
81405	Molecular pathology procedure level 6
81406	Molecular pathology procedure level 7
81407	Molecular pathology procedure level 8
81408	Molecular pathology procedure level 9
81415	Exome (e.g., unexplained constitutional or heritable disorder or syndrome); sequence analysis
81416	Exome (e.g., unexplained constitutional or heritable disorder or syndrome); sequence analysis, each comparator exome (e.g., parents, siblings)
81417	Exome (e.g., unexplained constitutional or heritable disorder or syndrome); re-evaluation of previously obtained exome sequence (e.g., updated knowledge or unrelated condition/syndrome)
81425	Genome (e.g., unexplained constitutional or heritable disorder or syndrome); sequence analysis
81426	Genome (e.g., unexplained constitutional or heritable disorder or syndrome); sequence analysis, each comparator genome (e.g., parents, siblings)
81427	Genome (e.g., unexplained constitutional or heritable disorder or syndrome); re-evaluation of previously obtained genome sequence (e.g., updated knowledge or unrelated condition/syndrome)
81479	Unlisted molecular pathology procedure
S3800	Genetic testing for amyotrophic lateral sclerosis (ALS)

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