Genetic Testing for Inherited Peripheral Neuropathies

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**Definitions**

| Charcot-Marie-Tooth neuropathy | A major type of hereditary polyneuropathy characterized by decreased motor nerve conduction velocity, distal sensory loss and weakness, abnormal deep tendon reflexes and skeletal abnormalities. CMT is genetically and clinically heterogenous, with more than 40 genes associated with the disease and the clinical phenotype ranging from minimal neurological findings to the classic pes cavus and "stork legs" and a severe polyneuropathy with respiratory failure. CMT is one of the most common inherited neuromuscular disorder affecting approximately 30 per 100,000 individuals. |
| Hereditary neuropathy with liability to pressure palsies (HNPP) | A slowly progressive but benign neuromuscular disorder caused by deletions of the PMP22 gene and resulting in inadequate production of PMP22 (i.e., peripheral myelin protein 22). HNPP increases susceptibility to nerve injury from stretch, pressure or repetitive use and is characterized by recurrent pressure neuropathies including carpal tunnel syndrome and peroneal palsy. |

**Related Medical Guidelines**

[Genetic Counseling and Testing](#)

**Guideline**

Genetic testing to confirm a diagnosis for inherited peripheral neuropathy and for all other indications is considered investigational and not medically necessary.
Applicable Procedure Codes

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<th>Code</th>
<th>Description</th>
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<td>81324</td>
<td>PMP22 (peripheral myelin protein 22) (eg, Charcot-Marie-Tooth, hereditary neuropathy with liability to pressure palsies) gene analysis; duplication/deletion analysis</td>
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<td>81325</td>
<td>PMP22 (peripheral myelin protein 22) (eg, Charcot-Marie-Tooth, hereditary neuropathy with liability to pressure palsies) gene analysis; full sequence analysis</td>
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<td>81326</td>
<td>PMP22 (peripheral myelin protein 22) (eg, Charcot-Marie-Tooth, hereditary neuropathy with liability to pressure palsies) gene analysis; known familial variant</td>
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References


