Local Coverage Article:

Links in PDF documents are not guaranteed to work. To follow a web link, please use the MCD Website.

Contractor Information

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<th>Contractor Name</th>
<th>Contract Type</th>
<th>Contract Number</th>
<th>Jurisdiction</th>
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Article Information

General Information

<table>
<thead>
<tr>
<th>Article ID</th>
<th>Original Article Effective Date</th>
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<tr>
<td>A55181</td>
<td>10/24/2016</td>
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<tr>
<th>Article Title</th>
<th>Revision Effective Date</th>
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Revision Ending Date N/A

Retirement Date N/A
**Article Guidance**

**Article Text:**

Hereditary hemorrhagic telangiectasia (HHT) is an autosomal dominant disorder with variable clinical presentation. Most common manifestations are epistaxis, recurrent gastrointestinal bleeding, iron deficiency anemia, and mucocutaneous telangectasias. Arteriovenous malformations (AVMs), of various organs (lung, liver, brain) are common.

The two most prevalent forms of HHT, Type 1 and Type 2, are caused by mutations in the endoglin (ENG) or the ACVRL1 gene respectively. Although identification of these gene mutations can confirm the diagnosis of HHT, these tests are not necessary in many cases. HHT is generally established using well vetted consensus criteria (most often the International Curacao Criteria (ICC)). The ICC uses the clinical characteristics of epistaxis, cutaneous or mucosal telangectasias, visceral AVMs, and a first-degree relative with HHT to judge likelihood of a given patient having HHT. A 'definite' diagnosis is established when a patient has 3 or 4 of these criteria. Genetic testing for ENG/ACVRL1 is not warranted. A patient with 0 to 1 criteria is 'unlikely' to have HHT, and similarly would not be a candidate for genetic testing. Patients with 2 or 3 Curacao criteria are defined as “suspected” of HHT and are candidates for ENG/ACVRL1 testing.

Since screening of patients without signs or symptoms of HHT, who have a first-degree relative with HHT, is not a Medicare benefit, the MolDX Team has determined ENG and/or ACVRL1 genetic testing and panels of tests that include ENG/ACVRL1 are statutorily excluded services.

**EXCEPTIONS:** For patients with “suspected” HHT in which diagnosis confirmation would demonstrate an improved outcome, approval will be made on a case-by-case basis through the appeal process.

For tests that include ENG and ACVRL1 registered and assigned a single Identifier, submit CPT code 81479.

To receive an ENG and/or ACVRL1 gene test denial, please submit the following claim information:

- Select appropriate CPT code for test
  - 81403-ENG, known familial variant
  - 81405-ENG, Duplication/deletion
  - 81406-ENG, full gene sequence
  - 81479-ACVRL1
  - 81479-ACVRL1 and ENG

- An Advance Beneficiary Notice (ABN) is not required for statutorily excluded services
For a voluntary issued ABN, append with GX modifier.

To indicate a valid ABN is on file for a known statutorily excluded service, append with a GY modifier.

Select the appropriate diagnosis for the patient.

For CPT non-NOC codes, Labs may either use the SV101-7 or SV202-7 (preferred) or the NTE field to submit this required information.

- Enter the appropriate DEX Z-Code™ identifier adjacent to the CPT code in the comment/narrative field for the following Part B claim field/types:
  - Loop 2400 or SV101-7 for the 5010A1 837P
  - Item 19 for paper claim
- Enter the appropriate DEX Z-Code™ identifier adjacent to the CPT code in the comment/narrative field for the following Part A claim field/types:
  - Line SV202-7 for 837I electronic claim
  - Block 80 for the UB04 claim form

Reference: Sec. 1862 (1)(A) Statutory Exclusion covers diagnostic testing “except for items and services that are not reasonable and necessary for the diagnosis or treatment of illness or injury or to improve the functioning of a malformed body member,...”

Coding Information

Bill Type Codes:

Contractors may specify Bill Types to help providers identify those Bill Types typically used to report this service. Absence of a Bill Type does not guarantee that the article does not apply to that Bill Type. Complete absence of all Bill Types indicates that coverage is not influenced by Bill Type and the article should be assumed to apply equally to all claims.

N/A

Revenue Codes:

Contractors may specify Revenue Codes to help providers identify those Revenue Codes typically used to report this service. In most instances Revenue Codes are purely advisory. Unless specified in the article, services reported under other Revenue Codes are equally subject to this coverage determination. Complete absence of all Revenue Codes indicates that coverage is not influenced by Revenue Code and the article should be assumed to apply equally to all Revenue Codes.

N/A

CPT/HCPCS Codes

Group 1 Paragraph: N/A

Group 1 Codes:

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<td>Group 1 CPT/HCPCS Code</td>
<td>Group 1 CPT/HCPCS Code Description</td>
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<td>MOLECULAR PATHOLOGY PROCEDURE, LEVEL 4 (EG, ANALYSIS OF SINGLE EXON BY DNA SEQUENCE ANALYSIS, ANALYSIS OF &gt;10 AMPLICONS USING MULTIPLEX PCR IN 2 OR MORE INDEPENDENT REACTIONS, MUTATION SCANNING OR DUPLICATION/DELETION VARIANTS OF 2-5 EXONS)</td>
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ICD-10 Codes that are Covered N/A
ICD-10 Codes that are Not Covered N/A

**Revision History Information**

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<tr>
<td>10/24/2016</td>
<td>R1</td>
<td>Added Part A claim filing information.</td>
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Related Local Coverage Document(s) N/A

Related National Coverage Document(s) N/A

Statutory Requirements URL(s) N/A

Rules and Regulations URL(s) N/A

CMS Manual Explanations URL(s) N/A

Other URL(s) N/A

Public Version(s) Updated on 05/02/2018 with effective dates 10/24/2016 - N/A Updated on 08/12/2016 with effective dates 10/24/2016 - N/A

**Keywords**

- MolDX
- ENG
- ACVRL1
- 81403
- 81405
- 81406
- 81479

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