>Title: Genetic Testing for Cardiomyopathies

**Effective Date:** September 21, 2016

This policy was developed with input from specialists in cardiology and genetics and endorsed by the Medical Policy Committee.

**Important Information – Please Read Before Using This Policy**

These services may or may not be covered by all Medica plans. Please refer to the member’s plan document for specific coverage information. If there is a difference between this general information and the member’s plan document, the member’s plan document will be used to determine coverage. With respect to Medicare and Minnesota Health Care Programs, this policy will apply unless those programs require different coverage. Members may contact Medica Customer Service at the phone number listed on their member identification card to discuss their benefits more specifically. Providers with questions about this Medica utilization management policy may call the Medica Provider Service Center toll-free at 1-800-458-5512.

Medica utilization management policies are not medical advice. Members should consult with appropriate health care providers to obtain needed medical advice, care and treatment.

**Purpose**

To promote consistency between reviewers in utilization management decision-making by providing the criteria that generally determine the medical necessity of genetic testing for cardiomyopathies. The Coverage Issues box below outlines the process for addressing the needs of individuals who do not meet these criteria.

**Background**

Definitions:

A. **Cardiomyopathies** are a group of diseases that cause the heart muscle to become abnormally enlarged, thickened, and/or stiffened, diminishing the heart's ability to function and creating the potential for arrhythmias, heart failure and sudden cardiac death. The wall thickness, chamber size, contraction, relaxation, conduction and rhythm of the heart may all be affected. Some people with these conditions remain asymptomatic. However, the disorders can produce an irregular heart rhythm that may result in dizziness, palpitations, fainting, seizures, heart failure and sudden death. The World Health Organization (WHO) recognizes four classes of cardiomyopathy: hypertrophic cardiomyopathy, arrhythmogenic right ventricular cardiomyopathy, dilated cardiomyopathy, and restrictive cardiomyopathy. However, other rarer forms of cardiomyopathy have been identified as well. These cardiomyopathies may have overlapping features with any of the previous types described and include, but are not limited to, left ventricular non-compaction (LVNC) and mitochondrial cardiomyopathies. Cardiomyopathies may be acquired or inherited.

1. **Hypertrophic cardiomyopathy (HCM)** is an autosomal dominant disorder caused by mutation in one of the genes currently known to encode different components of the sarcomere, the basic contractile unit of the cardiac myocyte. HCM is characterized by left ventricular hypertrophy (LVH) in the absence of predisposing cardiac conditions or cardiovascular conditions and occurs in approximately one in 500 individuals world-wide. Genes for which clinical genetic testing is available, include but are not limited to: beta myosin heavy chain (MYH7); myosin binding protein C (MYBPC3); troponin T (TNNT2), troponin I (TNNI3); alpha tropomyosin (TPM1); actin (ACTC); regulator light chain (MYL2); and essential light chain (MYL3).

2. **Arrhythmogenic right ventricular cardiomyopathy (ARVC)** is an autosomal dominant disorder of the cardiac desmosome, the protein complexes that maintain cell-to-cell connections and provide mechanical attachments among adjacent cells. ARVC is characterized by progressive fibrofatty replacement of the myocardium that predisposes to ventricular tachycardia and sudden death in young individuals and athletes. It primarily affects the right ventricle. However, with time, it may also involve the left ventricle. The disease prevalence is estimated at 1:1000 to 1:2500, but may be higher.
MEDICAL NECESSITY CRITERIA

Indications

Genetic testing for HCM, ARVC and DCM, using single gene or multigene panels, is considered medically necessary.
necessary when documentation in the medical records indicates that all of the following criteria are met:

A. The member has one of the following:
   1. A close relative (1st or 2nd degree) with a known HCM, ARVC or DCM pathogenic mutation
   2. Clinically diagnosed HCM, ARVC or DCM for the purposes of identifying a pathogenic mutation that can be used for family-specific screening in at-risk blood relatives.

B. Medical records document all of the following:
   1. A detailed family history or creation of a pedigree
   2. Genetic counseling by a cardiologist/electrophysiologist, medical geneticist, or certified and licensed (where required) genetic counselor not associated with the laboratory performing the testing
   3. How the test results will lead to changes in treatment decisions (i.e., medical or surgical management, lifestyle modifications, surveillance and/or other protective measures) of the member.

C. The test is ordered by a cardiologist/electrophysiologist, medical geneticist, or board-certified and licensed (where required) genetic counselor not associated with the laboratory performing the testing.

COVERAGE ISSUES

1. Prior authorization is required for genetic testing for HCM, ARVC and DCM.
2. Genetic testing for all other inherited cardiomyopathies, including, but not limited to LVNC and RCM is investigative and therefore not covered.
3. Genetic testing is excluded and therefore not covered when performed in the absence of symptoms or high risk factors for a genetic disease or when knowledge of genetic status will not affect treatment decisions or screening for the disease. (See Medica’s coverage policy entitled: Genetic and Pharmacogenetic Testing).
4. Coverage may vary according to the terms of the member’s plan document.
5. For Medicare members, refer to the following, as applicable: https://www.cms.gov/medicare-coverage-database/search/advanced-search.aspx.
6. If the Medical Necessity and Coverage Criteria are met, Medica will authorize benefits within the limits in the member’s plan document.
7. If it appears that the Medical Necessity and Coverage Criteria are not met, the individual’s case will be reviewed by the medical director or an external reviewer. Practitioners are reminded of the appeals process in their Medica Provider Administrative Manual.

DOCUMENT HISTORY

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<thead>
<tr>
<th>Original Effective Date</th>
<th>February 2012</th>
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<tbody>
<tr>
<td>MPC Endorsement Date(s)</td>
<td>11/2011, 11/2012, 04/2013, 04/2014, 02/2015, 04/2016, 09/2016</td>
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<tr>
<td>Administrative Update(s)</td>
<td>05/01/2017</td>
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References:

Pre-04/2016 MPC


01/2016 MTAC:


04/2016 MPC:

09/2016 MPC:
No new references added.