GUIDELINES
This policy does not certify benefits or authorization of benefits, which is designated by each individual policyholder contract. Paramount applies coding edits to all medical claims through coding logic software to evaluate the accuracy and adherence to accepted national standards. This guideline is solely for explaining correct procedure reporting and does not imply coverage and reimbursement.

DESCRIPTION
Cardiomyopathy is a chronic disease of the heart muscle (myocardium). The heart muscle becomes enlarged, thick or rigid resulting in a failure to pump blood effectively, irregular heartbeats (arrhythmias) and possibly heart failure. Cardiomyopathy can be acquired or inherited. Hypertrophic cardiomyopathy (HCM) is one of the main types of cardiomyopathy. Arrhythmogenic right ventricular dysplasia/cardiomyopathy (ARVD/C) is another type of cardiomyopathy characterized by fatty and fibro-fatty infiltration of the right ventricle with resultant myocardial cell atrophy, cell death and ventricular arrhythmias ranging from frequent premature ventricular contractions to ventricular tachycardia. Inherited arrhythmias, which may be life-threatening, often begin in childhood and adolescence.

Cardiac ion channelopathies are a group of diseases that develop due to defects in ion channels and can be caused by either genetic or acquired factors. Genetic testing may be used to detect mutations believed to be linked to inherited cardiomyopathies and channelopathies to assist with diagnosis, determine prognosis and identify susceptibility in at-risk asymptomatic family members.

Long QT Syndrome (LQTS) is a genetic cardiac channelopathy where the heart muscle takes more time than usual to recharge between beats. This shows as a prolonged QT interval on an electrocardiogram (EKG). This abnormal heartbeat pattern can lead to episodes of dizziness/fainting, cardiac arrest and sudden cardiac death (SCD) in affected individuals.

Catecholaminergic Polymorphic Ventricular Tachycardia (CPVT) is a genetic cardiac channelopathy characterized by an extremely fast and irregular heartbeat in response to exercise or emotional stress. CPVT may cause syncope (fainting), cardiac arrest, or sudden cardiac death (SCD).

Short QT Syndrome (SQTS) is an autosomal dominant channelopathy where the heart muscle takes less time than usual to recharge between beats. This shows as a shortened QT interval on an electrocardiogram (EKG). This abnormal heartbeat pattern may cause symptoms of dizziness/fainting and may increase the risk for adverse cardiac events. There is insufficient evidence to support a conclusion concerning the health outcomes or benefits associated with this procedure.

Brugada syndrome is characterized by cardiac conduction abnormalities that increase the risk of syncope, ventricular arrhythmia, and sudden cardiac death. The disorder primarily manifests during adulthood, although ages between 2 days and 85 years have been reported. Males are more likely to be affected than females (approximately an 8:1 ratio). There is insufficient evidence to support a conclusion concerning the health outcomes or benefits associated with this procedure.

POLICY
Genetic testing for cardiac conditions (81413, 81414, 81439, S3865, S3866) requires prior authorization.

Procedure code S3861 is non-covered.

HMO, PPO, Individual Marketplace, Elite, Advantage
Genetic testing may be excluded by contract. Please consult the member’s individual contract regarding coverage.

General Criteria for Genetic Tests
Paramount members may be eligible for genetic testing when ALL of the following criteria are met:

- Individual has not previously received genetic testing for the disorder. Note: In general, genetic testing for a particular disorder should be performed once per lifetime; however, there are rare instances in which
testing may be performed more than once in a lifetime (eg, previous testing methodology is inaccurate, a new discovery has added significant relevant mutations for a disease, significant changes in technology or treatments indicate that test results or outcomes may change as a result of repeat testing)

- Laboratory or clinical tests to definitively diagnose the genetic disorder are unavailable or results are equivocal
- Panels including, but may not limited to, multiple genes or multiple conditions, and in cases where a tiered approach/method is clinically available, may be covered ONLY for the number of genes or tests deemed medically necessary to establish a diagnosis
- Results of genetic testing will directly impact and change clinical management of the individual being tested who is a covered member
- Technical and clinical performance of the genetic test is supported by published peer-reviewed medical literature

The following genetic tests must meet the above General Criteria for Genetic Tests in addition to the individual criteria outlined below for each test.

Hypertrophic Cardiomyopathy (HCM)
Paramount members may be eligible for genetic testing for HCM when the following criteria are met:
- Individual to be tested has been evaluated (eg, electrocardiogram [ECG], echocardiography) and exhibits no clinical evidence of HCM; AND
- Has an affected first-degree blood relative (ie, parent, full-sibling, child) in whom a pathogenic or likely pathogenic HCM mutation has been identified. (Note: test for known familial mutation)

Arrhythmogenic Right Ventricular Cardiomyopathy/Dysplasia (ARVC/D)
Paramount members may be eligible for genetic testing for ARVC/D when the following criteria are met:
- Individual to be tested is asymptomatic and has an affected first-degree blood relative (ie, parent, full-sibling, child) or second-degree blood relative (ie, grandparents, grandchildren, aunts, uncles, nephews, nieces, half-siblings) in whom a pathogenic or likely pathogenic ARVC/D mutation has been identified (Note: test for known familial mutation)

Long QT Syndrome (LQTS)
Paramount members may be eligible for genetic testing for LQTS when the following criteria are met:
- Individual to be tested is asymptomatic and has an affected first-degree blood relative (ie, parent, full-sibling, child) in whom a pathogenic or likely pathogenic LQTS mutation has been identified (Note: test for known familial mutation);
- OR

Paramount does not cover genetic testing for LQTS with multi-gene panels that include other heritable cardiac conditions because it is considered experimental, investigational or unproven.

Paramount does not cover genetic screening for LQTS in the general population, because such screening is considered not medically necessary.

Catecholaminergic Polymorphic Ventricular Tachycardia (CPVT)
Paramount members may be eligible for genetic testing for CPVT when the following criteria are met:
- Individual to be tested is asymptomatic and has an affected first-degree blood relative (ie, parent, full-sibling, child) in whom a pathogenic or likely pathogenic CPVT mutation has been identified (Note: test for known familial mutation); OR
- Individual to be tested exhibits clinical features suggestive of CPVT including unexplained exercise- or catecholamine-induced polymorphic ventricular arrhythmias and syncope during physical activity or acute emotion occurring in a structurally normal heart

Non-Covered
Paramount members may NOT be eligible for genetic testing for cardiac conditions for ANY of the following:
- Short QT Syndrome (SQTS)
- Brugada syndrome

Paramount does not cover comprehensive or multi-condition multigene panels, with or without next-generation sequencing (NGS), including but may not be limited to:
Paramount does not cover genetic testing for atrial fibrillation because it is considered experimental, investigational or unproven.

Paramount does not cover genetic testing for hereditary cardiac conditions in the general population because such screening is considered not medically necessary.

**CODING/BILLING INFORMATION**

The appearance of a code in this section does not necessarily indicate coverage. Codes that are covered may have selection criteria that must be met. Payment for supplies may be included in payment for other services rendered.

**CPT CODES**

<table>
<thead>
<tr>
<th>Code</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>81280</td>
<td>Long QT syndrome gene analyses (e.g., KCNQ1, KCNH2, SCN5A, KCNE1, KCNE2, KCNJ2, CACNA1C, CAV3, SCN4B, AKAP, SNTA1, and ANK2); full sequence analysis <em>(Deleted effective 12/31/16)</em></td>
</tr>
<tr>
<td>81281</td>
<td>Long QT syndrome gene analyses (e.g., KCNQ1, KCNH2, SCN5A, KCNE1, KCNE2, KCNJ2, CACNA1C, CAV3, SCN4B, AKAP, SNTA1, and ANK2); known familial sequence variant <em>(Deleted effective 12/31/16)</em></td>
</tr>
<tr>
<td>81282</td>
<td>Long QT syndrome gene analyses (e.g., KCNQ1, KCNH2, SCN5A, KCNE1, KCNE2, KCNJ2, CACNA1C, CAV3, SCN4B, AKAP, SNTA1, and ANK2); duplication/deletion variants <em>(Deleted effective 12/31/16)</em></td>
</tr>
<tr>
<td>81403</td>
<td>Molecular pathology procedure, Level 4 (e.g., 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>
</tr>
<tr>
<td>81404</td>
<td>Molecular pathology procedure, Level 5 (e.g., analysis of 2-5 exons by DNA sequence analysis, mutation scanning or duplication/deletion variants of 6-10 exons, or characterization of a dynamic mutation disorder/triplet repeat by Southern blot analysis)</td>
</tr>
<tr>
<td>81405</td>
<td>Molecular pathology procedure, Level 6 (e.g., analysis of 6-10 exons by DNA sequence analysis, mutation scanning or duplication/deletion variants of 11-25 exons, regionally targeted cytogenomic array analysis)</td>
</tr>
<tr>
<td>81406</td>
<td>Molecular pathology procedure, Level 7 (e.g., analysis of 11-25 exons by DNA sequence analysis, mutation scanning or duplication/deletion variants of 26-50 exons, cytogenomic array analysis for neoplasia)</td>
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<tr>
<td>81407</td>
<td>Molecular pathology procedure, Level 8 (e.g., analysis of 26-50 exons by DNA sequence analysis, mutation scanning or duplication/deletion variants of &gt;50 exons, sequence analysis of multiple genes on one platform)</td>
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<tr>
<td>81408</td>
<td>Molecular pathology procedure, Level 9 (e.g., analysis of &gt;50 exons in a single gene by DNA sequence analysis)</td>
</tr>
<tr>
<td>81413</td>
<td>Cardiac ion channelopathies (e.g., Brugada syndrome, long QT syndrome, short QT syndrome, catecholaminergic polymorphic ventricular tachycardia); genomic sequence analysis panel, must include sequencing of at least 10 genes, including ANK2, CASQ2, CAV3, KCNE1, KCNE2, KCNH2, KCNJ2, KCNQ1, RYR2, and SCN5A</td>
</tr>
<tr>
<td>81414</td>
<td>Cardiac ion channelopathies (e.g., Brugada syndrome, long QT syndrome, short QT syndrome, catecholaminergic polymorphic ventricular tachycardia); duplication/deletion gene analysis panel, must include analysis of at least 2 genes, including KCNH2 and KCNQ1</td>
</tr>
<tr>
<td>81439</td>
<td>Inherited cardiomyopathy (e.g., hypertrophic cardiomyopathy, dilated cardiomyopathy, arrhythmogenic right ventricular cardiomyopathy) genomic sequence analysis panel, must include sequencing of at least 5 genes, including DSG2, MYBPC3, MYH7, PKP2, and TTN</td>
</tr>
</tbody>
</table>

**HCPCS CODES**

<table>
<thead>
<tr>
<th>Code</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>S3861</td>
<td>Genetic testing, sodium channel, voltage-gated, type V, alpha subunit (SCN5A) and variants for suspected Brugada Syndrome</td>
</tr>
<tr>
<td>S3865</td>
<td>Comprehensive gene sequence analysis for hypertrophic cardiomyopathy</td>
</tr>
<tr>
<td>S3866</td>
<td>Genetic analysis for a specific gene mutation for hypertrophic cardiomyopathy (HCM) in an individual with a known HCM</td>
</tr>
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</table>
mutation in the family

**TAWG REVIEW DATES:** 07/08/2009, 08/11/2010, 08/10/2011, 07/11/2012, 03/24/2017, 03/22/2018

**REVISION HISTORY EXPLANATION**

**01/01/11:** No changes

**07/12/12:** Revision to match U/CM Benefit Description and Limitations of Coverage

**09/09/14:** Removed deleted codes 83891, 83892, 83894, 83898, 83904, 83909, 83912, S3860, & S3862. Added new codes 81280, 81281, 81282, & 81406. Policy reviewed and updated to reflect most current clinical evidence per Medical Policy Steering Committee.

**03/24/17:** Changed title from Long QT Syndrome (LQTS) Genetic Testing to Genetic Testing for Cardiac Conditions. Deleted effective 12/31/16 CPT codes 81280-81282. Added effective 01/01/17 new CPT codes 81413, 81414, & 81439 requiring prior authorization. Added codes 81403-81405, 81407-81408, & S3861-S3866. Code S3861 is non-covered for all product lines. Codes 81403-81405, 81407-81408, & S3865, S3866 require prior authorization. Policy reviewed and updated to reflect most current clinical evidence per The Technology Assessment Working Group (TAWG).

**03/22/18:** Policy reviewed and updated to reflect most current clinical evidence per The Technology Assessment Working Group (TAWG).

**REFERENCES/RESOURCES**

Centers for Medicare and Medicaid Services, CMS Manual System and other CMS publications and services
Ohio Department of Medicaid [http://jfs.ohio.gov/](http://jfs.ohio.gov/)
Centers for Medicare and Medicaid Services, Healthcare Common Procedure Coding System, HCPCS Release and Code Sets
Industry Standard Review
Hayes, Inc.