



**GENETIC TESTING FOR
LONG QT SYNDROME
HS-148**



Harmony Behavioral Health, Inc.

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Genetic Long QT Syndrome

Policy Number: HS-148

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DISCLAIMER

The Clinical Coverage Guideline is intended to supplement certain standard WellCare benefit plans. The terms of a member's particular Benefit Plan, Evidence of Coverage, Certificate of Coverage, etc., may differ significantly from this Coverage Position. For example, a member's benefit plan may contain specific exclusions related to the topic addressed in this Clinical Coverage Guideline. When a conflict exists between the two documents, the Member's Benefit Plan always supersedes the information contained in the Clinical Coverage Guideline. Additionally, Clinical Coverage Guidelines relate exclusively to the administration of health benefit plans and are NOT recommendations for treatment, nor should they be used as treatment guidelines. The application of the Clinical Coverage Guideline is subject to the benefit determinations set forth by the Centers for Medicare and Medicaid Services (CMS) National and Local Coverage Determinations and state-specific Medicaid mandates, if any.

APPLICATION STATEMENT

The application of the Clinical Coverage Guideline is subject to the benefit determinations set forth by the Centers for Medicare and Medicaid Services (CMS) National and Local Coverage Determinations and state-specific Medicaid mandates, if any.

BACKGROUND

Long QT syndrome (LQTS) is a disorder of the heart's electrical system. It is characterized by prolongation of the QT interval. The QT interval is the section on the electrocardiogram (ECG) that represents the time it takes for the electrical system to fire an impulse through the ventricles and then recharge. The electrical activity of the heart is produced by the flow of ions in and out of the cells of the heart. LQTS is a defect in the ion channels, which causes a delay in the time it takes for the electrical system to recharge after each heartbeat. LQTS disorders are considered channelopathies, or diseases that affect cardiac ion channels. This condition predisposes the individual to cardiac events and arrhythmias including: torsades de pointes ventricular tachycardia, syncopal episodes, ventricular fibrillation and cardiac arrest. LQTS may also be caused by acquired factors, most commonly by use of certain drugs that will cause prolongation of the QT interval.

LQTS is diagnosed by considering the clinical features, the family history and the ECG findings of the patient. Unexplained syncope or sudden cardiac death in a child or young adult should raise suspicion of the possibility of LQTS. Electrophysiological testing has not been demonstrated to be helpful in making this diagnosis. The clinical features are a result of the torsades, and may range from minor symptoms such as dizziness, to seizure, syncope and sudden death. Congenital LQTS will usually manifest before the age of 40 years, generally in childhood and adolescence. The age is usually dependent on the genotype. The prolongation of the QT interval is a risk factor independent of patient's age, history of myocardial infarction, heart rate and history of drug use. Patients with QT interval corrected by heart rate (QTc) of greater than 440 milliseconds are at two to three times higher risk for sudden cardiac death than those with QTc interval of under 440 milliseconds. The mortality rate of untreated patients with LQTS is in the range of 1% to 2% per year. Sudden cardiac death may be precipitated by a triggering event, such as physical exercise, swimming, sleep deprivation, auditory stimuli, and sudden intense sympathetic stimuli. In an effort to enhance diagnostic reliability, an elaborate point score system has been proposed that incorporates QTc duration, as well as other hallmarks such as syncope and a family history of this condition.

There are several forms of LQTS, depending on the genes responsible and the features associated with the condition. Most forms of LQTS are carried in an autosomal dominant manner, with the exception being Jervell and Lange-Nielsen syndrome (JLNS), which is inherited in an autosomal recessive manner. Articles in the medical literature may use the terminology LQT1, LQT2, etc., to refer to the locus name of genes involved in LQTS, or the phenotype.

Familion[®] Test

The Familion[®] test (Clinical Data, Inc., Newton, MA) is a patented genetic test that is intended to provide analysis of five major cardiac ion channel genes. According to the Clinical Data website, the analysis includes sequence determination and variant detection. It is noted at the website that the genes analyzed by this test include: KCNQ1, KCNH2, SCN5A, KCNE1, and KCNE2. Testing for LQTS may be performed with one of following configurations:

- Comprehensive cardiac ion channel analysis: This will provide analysis for variants in all five genes and is appropriate when there is a high index of suspicion of disease such as stress-induced syncope, prolonged QT interval, family history of sudden cardiac death and unexplained ventricular arrhythmia.
- Family specific analysis: This test provides analysis of one or more mutations found in an index case using either one of the above test configurations or confirmed results from another laboratory and is appropriate for testing blood relatives.

POSITION STATEMENT

Genetic testing for long QT syndrome **is considered medically necessary** in the following circumstances:

- Members with a prolonged QT interval on resting electrocardiogram (a corrected QT interval (QTc) of 470 msec in adult males and 480 msec in adult females*) without an identifiable external cause for QTc prolongation (such as heart failure, bradycardia, electrolyte imbalances, certain medications and other medical conditions); **OR**,
- Members with first-degree relatives (siblings, parents, or offspring) with a defined LQT mutation, or long QT syndrome resulting in sudden death of a first or second degree relative.

* NOTE: A corrected QT interval (QTc) of 450 msec is used in male children, and 460 msec in female children.

Genetic testing for long QT syndrome **is considered NOT medically necessary** and NOT a covered benefit in the following circumstances:

- The above criteria are not met; **AND**,
- General population testing; **AND**,
- Prenatal testing

CODING

CPT® Codes and units billed specific to the Familion Long QT Syndrome Test

- 83891** Molecular diagnostics; isolation or extraction of highly purified nucleic acid, each nucleic acid type (DNA or RNA) 1 unit is billed
- 83898** Molecular diagnostics; amplification, target, each nucleic acid sequence 108 units are billed
- 83904** Molecular diagnostics; mutation identification by sequencing, single segment, each segment 108 units are billed
- 83909** Molecular diagnostics; separation and identification by high resolution technique (e.g., capillary electrophoresis), each nucleic acid preparation 108 units are billed
- 83912** Molecular diagnostics; interpretation and report 1 unit is billed.

ICD-9-CM Procedure Codes - Not applicable

HCPCS Level II Codes

- S3860*** Genetic testing, comprehensive cardiac ion channel analysis, for variants in 5 major cardiac ion channel genes for individuals with high index of suspicion for familial Long QT Syndrome (LQTS) or related syndromes
- S3861*** Genetic testing, sodium channel, voltage-gated, type V, alpha subunit (SCN5A) and variants for suspected Brugada syndrome
- S3862*** Genetic testing, family-specific ion channel analysis, for blood-relatives of individuals (index case) who have previously tested positive for a genetic variant of a cardiac ion channel syndrome using either one of the above test configurations or confirmed results from another laboratory
- S3865*** Comprehensive gene sequence analysis for hypertrophic cardiomyopathy
- S3866*** Genetic analysis for a specific gene mutation for hypertrophic cardiomyopathy (HCM) in an individual with a known HCM mutation in the family.

*Note: S-Codes are NON COVERED FOR MEDICARE – Refer to HCPCS Level II Temporary National Codes For Medicare, bill the appropriate CPT code listed above

Covered ICD-9-CM Diagnoses Codes

- 426.82** Long QT syndrome
V17.41 Family history of sudden cardiac death (SCD)
V17.49 Family history of other cardiovascular diseases

*Current Procedural Terminology (CPT®) ©2012 American Medical Association: Chicago, IL.

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Government Agencies, Professional and Medical Organizations

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HISTORY AND REVISIONS

Date	Action
1/5/2012	<ul style="list-style-type: none">• Approved by MPC. Reformatted references. No changes.
12/1/2011	<ul style="list-style-type: none">• New template design approved by MPC.