

**Policy #: 082**

**Original policy date: 3/01/09**

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**Revised date: 8/3/09**

**Title:**

**Genetic Testing for Congenital Long QT Syndrome**

**Description:**

Congenital Long QT Syndrome refers to a genetic abnormality, present at birth, which affects the conduction system of the heart. A patient with Congenital Long QT Syndrome is at-risk for a particular type of heart rhythm disturbance known as “Torsades de Pointes”, which is similar to ventricular tachycardia (heart rate greater than 100 beats per minutes) and can rapidly lead to sudden death from cardiac arrest. This condition is considered to be a frequent cause of sudden death in young people, particularly athletes during exertion and others during stressful periods. Patients with the condition can reduce their risk of sudden death through medication and changes in life style.

The condition can be diagnosed several ways. Physical signs of the condition include fainting, syncope (sudden loss of consciousness), and cardiac arrest. In addition to these physical symptoms, congenital long QT syndrome can be diagnosed by EKG. The segment of the EKG known as the QT interval is longer than expected in these patients. Sometimes the diagnosis is not clear from the EKG, and another approach is needed.

It has been shown that there are three genetic abnormalities that cause 90% of long QT syndrome cases. Genetic testing can detect these anomalies and these patients can be treated to reduce their risk. In those cases where the diagnosis is obvious from the EKG, genetic testing has not been demonstrated to change the treatment of the patient, and is therefore not medically necessary.

**When services are covered for Commercial products (including Medicare HMO Blue, Medicare PPO Blue and Blue Medicare PFFS Plus Rx products)**

We cover genetic testing in patients with suspected congenital long QT syndrome for the following indications: Individuals who do not meet the clinical criteria (visible manifestation of the three genetic abnormalities that cause 90% of long QT syndrome cases), but who have:

- a close relative (i.e., first-, second-, or third-degree relative) with a known LQTS mutation; or
- a close relative diagnosed with LQTS by clinical means whose genetic status is unavailable; or
- signs and/or symptoms indicating a moderate-to-high pretest probability of LQTS.

Determining the pretest probability of LQTS is not standardized. An example of a patient with a moderate to high pretest probability of LQTS is a patient with a Schwartz score of 2-3.\*

\*Schwartz score is the calculated heart rate corrected QT interval (QTc) from the 12-lead ECG. In addition, the score considers findings from both a clinical and family history. A Schwartz score equal to or exceeding 4 indicates high probability or definite LQTS (D-LQTS), whereas lower scores are associated with either possible-LQTS (P-LQTS), also called borderline LQTS, or low-probability LQTS.

**When services are not covered for Commercial products ( including Medicare HMO Blue, Medicare PPO Blue and Blue Medicare PFFS Plus Rx)**

We do not cover genetic testing for Long QT Syndrome to determine prognosis and/or direct therapy in patients with known Long QT Syndrome.

**Individual consideration**

All our medical policies are written for the majority of people with a given condition. Each policy is based on medical science. For many of our medical policies, each individual’s unique clinical circumstances may be considered in light of current scientific literature. For consideration of an individual patient, physicians may send relevant clinical information to:

**For services already billed**

Blue Cross Blue Shield of Massachusetts  
Provider Appeals  
PO Box 986065  
Boston, MA 02298

**Prior to performance of service**

Blue Cross Blue Shield of Massachusetts  
Case Creation/Medical Policy  
One Enterprise Drive  
Quincy, MA 02171  
Tel: 1-800-327-6716  
Fax: 1-888-641-5330

**Managed care guidelines**

- Any specialist visit requires a referral for **Medicare HMO Blue**.
- For all other Managed Care plans, any specialist visit requires a referral, except for visits performed by OB/GYN specialists.
- Authorization is required for an inpatient admission.

**Indemnity and PPO guidelines**

All authorization requirements are determined by the individual’s subscriber certificate, however:

- Authorizations are required for all inpatient services.
- Authorizations are not required for most outpatient services as determined by the individual’s subscriber certificate.
- Referrals to a specialist are not required.

**Coding information**

*Procedure codes are from current CPT, HCPCS Level II, Revenue Code, and/or ICD-9-CM manuals, as recommended by the American Medical Association, Centers for Medicare and Medicaid Services and American Hospital Associations. Blue Cross Blue Shield Association national codes may be developed when appropriate.*

*The following code is included below for informational purposes. Inclusion or exclusion of a code does not constitute or imply member coverage or provider reimbursement. Please refer to the member’s contract benefits in effect at the time of service to determine coverage or non-coverage as it applies to an individual member.*

There is no specific CPT code for this test. Multiple codes describing genetic analysis would likely be used.

CPT genetic testing code modifier specific to this syndrome:

Modifier 8C - Long QT syndrome

- HCPCS National code 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 (*New procedure code effective 10/01/08*)

- HCPCS National code S3861; Genetic testing, sodium channel, voltage-gated, type V, alpha subunit (SCN5A) and variants for suspected Brugada Syndrome (*New procedure code effective 10/01/08*)
- HCPCS National code 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 (*New procedure code effective 10/01/08*)

**NOTE:** The above codes will deny, leaving no patient balance if submitted with a diagnosis other than the covered conditions.

See footnote 2 for medically necessary diagnoses for all Products, including Medicare HMO Blue, Medicare PPO Blue, and Medicare PFFS PlusRx Products.

#### **Policy update history**

03/09, New policy created from review of literature stating that genetic testing for Long QT syndrome is a screening technique for individuals who meet the stated criteria. 3/09 Updated references 21-27. No change in policy statement. Reviewed 4/09 MPG – Cardiology, no changes in coverage were made.

#### **Footnotes:**

<sup>1</sup>Based on Blue Cross Blue Shield Association medical policy # 2.04.43, Genetic Testing for Long QT Syndrome.

<sup>2</sup> ICD-9 CM Diagnosis code, **426.82** - Long QT syndrome (code effective 1/1/06)

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