

UPMC Health Plan POLICY AND PROCEDURE MANUAL

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SUBJECT: Genetic Testing for Long QT Syndrome
INDEX TITLE: Medical Management
ORIGINAL DATE: June 2009

This policy applies to the following lines of business: (Check those that apply.)

Commercial:					
HMO ()		POS ()		PPO ()	
Fully Insured ()		Self-funded/ASO ()		HSA ()	
Medicare Select ()		Medicare Supplement ()			
DPW-MA:					
Health Choices ()			Voluntary ()		All ()
CMS-MA:					
OH ()		WV ()		PA ()	All (X)
HMO (X)	PPO(X)	Specialty Needs Plan (X)	Part D ()	PFFS ()	All ()
PID-CHIP:					
Free ()		Sub ()		Full ()	All (X)
APPLICABLE TO:					
Community Care ()			Work Partners ()		

I. POLICY

It is the policy of UPMC health plan to cover Genetic Testing for Long QT Syndrome when it is medically necessary and covered under the member's benefit plan. UPMC Health Plan recognizes the need for genetic testing which improves the diagnosis of Long QT Syndrome with resulting changes in patient management that improves health outcomes.

All denials are based on medical necessity and appropriateness as determined by a UPMC Health Plan Medical Director (Medical Director).

II. DEFINITIONS

N/A

III. PURPOSE

The purpose of this policy is to establish criteria for coverage of Genetic Testing for Long QT Syndrome.

IV. SCOPE

This policy applies to various UPMC Health Plan departments as indicated by the Benefit and Reimbursement Committee. These include but are not limited to Medical Management, Benefit Configuration and Claims Departments.

Proprietary and Confidential Information of UPMC Health Plan

V. PROCEDURE

A. Medical Description / Background

Hereditary Long QT Syndrome (LQTS) is a disorder of the heart's electrical system. The QT interval on an EKG represents the time it takes for an electrical impulse to travel through the ventricles of the heart and then recharge. Long QT syndrome is characterized by a lengthening of the QT interval. The normal QT interval is about 0.44 seconds although this can vary among individuals and by sex. LQTS disorders are also referred to as channelopathies as it is a defect in the cardiac ion channels (abnormalities in the sodium and potassium channels) which causes the delay in the time required for the electrical system of the heart to recharge after a heartbeat. This defect predisposes individuals to cardiac events and arrhythmias including: torsades de pointes (a form of ventricular tachycardia with a long QT interval), syncope, ventricular fibrillation and cardiac arrest. Many individuals with LQTS are asymptomatic. Others may exhibit syncope (fainting) and have abnormal cardiac rate and/or rhythm. Some may demonstrate prolongation of the QT interval only during physical exercise or intense emotion. By detecting genetic mutations in certain individuals, the test results can help to guide treatment options and may prevent sudden cardiac death.

Currently there are four recognized LQTS syndromes depending on the genes responsible and the features associated with the condition:

- Romano Ward syndrome (RWS) which is the most common form and accounts for approximately 85% of all LQTS cases. In this syndrome abnormalities are confined to the heart. A syncopal event is the most common symptom.
- Jervell and Lange-Nielson syndrome is associated with congenital sensorineural deafness. This is a more rare and severe syndrome. The QT interval is more markedly prolonged, usually longer than 500 msec which corresponds with a high risk for malignant ventricular arrhythmias and sudden death. Over half of all children who are untreated die before 15 years of age.
- Anderson-Tawil syndrome is associated with multiple organ involvement including dysmorphic body features (low set ears, small mandible, fifth digit clinodactyly, short stature) and periodic paralysis.
- Timothy syndrome is a rare multisystem disorder involving disorders of the skeleton, nervous system and heart. It is reported that none of the individuals diagnosed with this syndrome have had an affected parent.

The most common LQTS genotype mutations fall into 3 subtypes – LQT1, LQT2 and LQT3. Each subtype has their own set of specific triggers associated with cardiac events such as the LQT1 (the most common type of LQTS) trigger appears to be exercise/stress versus rest/sleep. Therefore patients with LQT1 may be advised to minimize exercise. Also the frequency of lethal events appears affected by the specific genotype.

Although there is no gold standard for diagnosis of this disorder, the Schwartz point score system (utilizing EKG findings, family history and clinical exam) can be used. However, studies indicate that genetic testing will identify more individuals with a LQTS mutation than those diagnosed by clinical methods. This is significant because underdiagnosing of LQTS may result in catastrophic outcomes.

The FAMILION test (Clinical Data, Inc) detects genetic mutations that can cause cardiac channelopathies including LQTS. The test helps diagnose the disease, guides treatment options and can determine whether family members are at risk. Genetic counseling prior to genetic testing is prudent, considering the impact of positive results on the member and the family.

Treatment of LQTS consists of lifestyle modification, Beta blocker therapy and/or an implantable cardiac defibrillator for individuals with previous cardiac arrest who have reasonable expectation of survival with a good functional status. Management would also include avoidance of drugs that are known to prolong the QT interval.

Long QT syndrome therefore can also be drug induced. Drugs that can induce this syndrome include tricyclic antidepressants, phenothiazines and certain antivirals and antifungals. If a drug is the cause - it is discontinued but until drug clearance is complete, patients may still require treatment.

B. Specific Indications

For members with suspected LQTS as evidenced by any of the following:

1. QTc interval > 470 msec in males and > 480 msec in females
2. Documented history of Torsades de pointes
3. Presence of T- wave alternans **AND** notched T waves in 3 leads
4. A first degree relative with a confirmed clinical diagnosis of LQTS
5. Members with a QTc > 440 msec **AND** an episode of aborted sudden death without another cause (such as cardiomyopathy or MI)
6. Unexplained syncope and either
 - A QTc > 450 msec for males or > 460 msec for females **OR**
 - A known family member (1st or 2nd degree relative) with genetically identified LQTS

C. Limitations

1. Members with a known cause for **acquired** LQTS such as drug induced, intracranial bleed or acute MI
2. Genetic screening for LQTS in the general population
3. Genetic screening to determine prognosis and/or direct therapy in patients with known LQTS
4. Family testing of members with genetically-proven LQTS

D. Information Required for Review

In order for medical necessity to be established, adequate information must be furnished by the treating physician. Necessary information includes, but is not limited to, a physician's letter of medical necessity which includes supporting documentation such as patient symptoms, QT interval per EKG and family history with clinical documentation validating diagnosis.

E. Review Process

1. The Medical Management staff assigned to review obtains the clinical information to determine if there is adequate clinical information. If the case does not meet the established criteria, it is referred to a UPMC Health Plan Medical Director.
2. If referred, the Medical Director determines if the requested service is medically necessary and appropriate.
3. The Medical Management staff completes the review process and communicates the review decision according to the Timeliness of UM Decisions policy for the member's benefit plan.

F. Variations

N/A

G. References

1. Clinical Data, Inc., PGx Health Division, *The Familion Test*, 1999-2008
2. Blue Cross and Blue Shield Association, Technology Evaluation Center, *Genetic Testing for Long QT Syndrome*, 2008
3. Aetna, Clinical Policy Bulletin, *Genetic Testing*, 04/07/2009
4. Cigna, Medical Coverage Policy #0193, *Genetic Testing for Long QT Syndrome*, 12/15/2008
5. American Heart Association, *Long Q-T Syndrome*, last updated 09/21/2007
6. Journal of the American College of Cardiology, 2006; 49:247-346, *ACC/AHA/ESC 2006 Guidelines for Management of Patients with Ventricular Arrhythmias and the Prevention of Sudden Cardiac Death*
7. Highmark, Medical Policy L-34, *Genetic Testing*, 11/03/2008
8. BlueCross BlueShield of North Carolina, Corporate Medical Policy MED 1172, *Genetic Testing for Long QT Syndrome*, 10/2008

Disclaimer:

UPMC Health Plan medical payment and prior authorization policies do not constitute medical advice and are not intended to govern or otherwise influence the practice of medicine. The policies constitute only the reimbursement and coverage guidelines of UPMC Health Plan and its affiliated managed care entities. Coverage for services varies for individual members in accordance with the terms and conditions of applicable Certificates of Coverage, Summary Plan Descriptions, or contracts with governing regulatory agencies.

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