



ADMINISTRATIVE POLICY AND PROCEDURE

Policy #:	1402	
Subject:	Genetic Testing for Long QT Syndrome	
Section:	Care Management	
Effective Date:	01/22/2012	
Revision Date(s):	10/13, 10/14	
Review Date(s):	10/15, 10/16	
Responsible Parties:	Patryce Toye, MD	
Responsible Department(s):	Utilization Management	
Regulatory References:	MD Delmarva 2016: 7.2 NCQA 2016: UM 2C	
Approved:		
	Carol Attia, RN AVP, Care Management	Patryce A. Toye, MD Senior Medical Director

Purpose: This policy describes the medical necessity and indications used by MFC in reviewing requests for Genetic Testing for Long QT Syndrome (LQTS)

Scope: MedStar Family Choice, MD; MedStar Family Choice, District of Columbia Healthy Families and Alliance.

Policy: MedStar Family Choice considers genetic testing for Long QT Syndrome (LQTS) medically necessary for ANY of the following indications when:

- 1) The results will directly impact clinical decision-making and/or clinical outcome for the individual, and
- 2) The testing method is considered scientifically valid for identification of the genetic mutation

Indications:

1. Persons with a prolonged QT interval on resting electrocardiogram, (i.e., corrected QT [QTc] of >470 msec in males and >480 msec in females) without an identifiable external cause for QTc prolongation (such as heart failure, bradycardia, electrolyte imbalance, certain medications [some antibiotics, antidepressants, antihistamines, diuretics, heart medications, cholesterol lowering, diabetic, antivirals, and/or antifungals])

2. **Person who has a first degree relative (siblings, parents, children) with a known LQT mutation or long QT syndrome**
3. **Person who has a first degree relative with a prolonged QTc (see above) who has experienced sudden death, or near death, had not been genetically tested and a genetic syndrome is suspected.**

Background: Hereditary Long QT Syndrome (LQTS) is a disorder of the heart's electrical system. It predisposes the individual to life threatening cardiac events and arrhythmias including: torsades de pointes, ventricular tachycardia, syncopal episodes, ventricular fibrillation and cardiac arrest. It can present in childhood. LQTS may also be caused by acquired factors, most commonly by use of certain drugs that will cause prolongation of the QT interval. Drug induced prolongation of the QT interval should be excluded before considering gene testing.

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

1. **Romano Ward Syndrome (RWS) is the most common mutation (85%) and abnormalities are confined to the heart. T wave changes may coexist. The most common symptom is syncope. It is inherited as an autosomal dominant.**
2. **Jervell and Lange-Nielson Syndrome is associated with congenital sensorineural deafness. The QT interval is usually more markedly prolonged, and associated with ventricular arrhythmias and sudden death. It is inherited as an autosomal recessive.**
3. **Anderson-Tawil Syndrome is associated with multiple organ involvement such as abnormalities with digits, ears, stature et.al. Periodic paralysis may also be a finding.**
4. **Timothy Syndrome is a rare multisystem disorder of the skeleton, heart, and nervous system. None of these person's parents are affected.**

Currently the Familion test (Transgenomic, INC) is the test of choice in detecting LQTS and guides treatment options. Genetic Counseling is encouraged before ordering the test and after the results are known to assist with the complex decision making that may follow the testing.

Procedure:

1. **Requests for authorization for testing should be submitted to MSFC office in Maryland or the District of Columbia during normal business hours**
2. **Requests can be mailed or faxed.**
3. **Submit supporting documentation with the request including Medical records outlining symptoms, EKG, Holter monitor report, other cardiac studies, pedigree information, medication history or any other information deemed appropriate by the requesting practitioner.**
4. **Requests for indications other than those listed will be reviewed by a Medical Director. In addition to the information outlined above, supporting information for such requests might also include information from the medical literature or other information.**

Summary of Changes:	10/16: <ul style="list-style-type: none">• No changes 10/15: <ul style="list-style-type: none">• No changes
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