**Letter of Medical Necessity for Long QT Syndrome (LQTS) Sequencing and Deletion/Duplication Panel**

**Patient Information**

**Date:**

**Patient Name:**

**Patient DOB:**

**Insurance Company Name, Address, City, State:**

**Policy Number:**

**Group Number:**

**ICD10 Codes:**

**Test Information**

**Test Name:** Long QT Syndrome (LQTS) Sequencing and Deletion/Duplication Panel

**CPT Codes:** 81403x1, 81406x2

**Laboratory:**

GeneDx, Inc.

(NPI#1487632998 / TAXID#205446298 / CLIA#21D0969951)

207 Perry Parkway

Gaithersburg, MD 20877

Telephone: (301) 519-2100

Fax: (201) 421-2010

This letter is in regards to my patient, [FIRST NAME LAST NAME], to request full coverage for the Long QT Syndrome (LQTS) Sequencing and Deletion/Duplication Panelto be performed by GeneDx. It is my professional determination that testing is medically necessary and will have a direct impact on this patient’s treatment and management.

**Patient Clinical and Family History**

This testing is requested due to this patient’s personal medical history, which includes the following clinical findings:

* Add Phenotype
* Add Phenotype
* Add Phenotype

The patient’s family history is negative for related conditions / unknown / remarkable for the following related clinical features:

The patient has previously had the following uninformative genetic and other testing:

* Add test
* Add test
* Add test

**Clinical Evidence and Guidelines for Testing**

The Long QT Sequencing and Deletion/Duplication Panel includes germline analysis of genes involved in conditions that include severe cardiovascular manifestations, including sudden cardiac arrest and sudden cardiac death. Panel testing includes both sequencing and deletion/duplication analysis of multiple genes simultaneously.

Long QT syndrome (LQTS) is due to abnormal cardiac ion channel function and is characterized by prolongation of the QT interval on electrocardiogram (ECG). LQTS is associated with increased risk for syncope, ventricular arrhythmia, and sudden cardiac death in young adults with normal heart structure. Sudden death is the first and final symptom in 10-15% of individuals.1 LQTS has an estimated prevalence of 1 in 2000 individuals, occurs in all ethnicities, and results in approximately 4000 deaths annually in the US.1,2,3

The diagnosis of LQTS is based on clinical history, ECG findings, genetic testing, and family history. Typically, the disorder manifests in patients younger than 40 years of age and may present as early as infancy. Patients often have a history of syncope or palpitations in the absence of any other causes, such as medications, structural heart abnormalities, myocardial ischemia, or electrolyte imbalances. In some patients, syncope may be mistakenly diagnosed as seizures. LQTS may be present even in the absence of any clinical symptoms. In some patients, sudden cardiac death occurs without any preceding symptoms and without an identifiable cause at autopsy. Inherited LQTS may underlie up to 10-15% of sudden infant death syndrome (SIDS) cases.4  Approximately seventy-five percent of cases of LQTS are due to known genetic causes.1

The diagnosis of arrhythmias, including long QT, can often be established by noninvasive electrophysiological studies, including electrocardiogram, cardiac stress test, Holter and other event monitoring. However, when imaging results are absent, subtle, or non-specific, molecular diagnosis with genetic testing aids in diagnosis, management and establishing recurrence risk for family members.

National and international medical societies have published guidelines that recommend genetic testing for long QT syndrome and other arrhythmias:

* The HRS/EHRA Expert Consensus Statement states that genetic testing is recommended with patients with clinical suspicion of LQTS or asymptomatic patients with primary QT prolongation.7

**Patient Clinical Utility and Medical Management Implications**

The results of this testing will guide appropriate medical management for this patient, including surveillance, preventive measures, and medical and surgical treatment. Management for arrhythmias is summarized in specific consensus documents from the American College of Cardiology / American Heart Association (ACC/AHA), the Heart Rhythm Association (HRS) and the European Heart Rhythm Association (EHRA), and in the European Society of Cardiology (ESC) guidelines on ventricular arrhythmias.5-8 Treatment for arrhythmia, and surveillance for progression, is critical and is strongly influenced by knowledge of the underlying genetic cause.1,5,6 Molecular genetic testing is critical to aid patient management in a cost-effective way and to minimize morbidity and mortality.1,5,6 Lifestyle modifications are also important to prevent or reduce exposure to triggers that can precipitate cardiac events in individuals with LQTS. 1,5,6

Specifically for this patient, the results of this test will also {ADD ADDITIONAL INFORMATION}

**Summary**

The Long QT Syndrome (LQTS) Sequencing and Deletion/Duplication Panelat GeneDx is a highly sensitive and cost-effective genetic test. I am requesting coverage for this medically necessary test in order to establish appropriate medical management for this patient. Without testing, treatment would be suboptimal, subjecting this patient to increased morbidity and potentially early mortality.

Thank you for your review and consideration. If you have questions, or if I can be of further assistance, please do not hesitate to call me at (XXX) XXX-XXXX.

Sincerely,

Signature

Ordering Provider’s Name

References:

1. Alders M, Bikker H, Christiaans I. Long QT Syndrome. 2003 Feb 20 [Updated 2018 Feb 8]. In: Adam MP, Ardinger HH, Pagon RA, et al., editors. GeneReviews® [Internet]. Seattle (WA): University of Washington, Seattle; 1993-2018. Available from: https://www.ncbi.nlm.nih.gov/books/NBK1129/Lehnart et al. (2007)
2. Inherited arrhythmias: a National Heart, Lung, and Blood Institute and Office of Rare Diseases workshop consensus report about the diagnosis, phenotyping, molecular mechanisms, and therapeutic approaches for primary cardiomyopathies of gene mutations affecting ion channel function. *Circulation* 116 (20):2325-45 (PMID: 17998470)
3. Vincent, et al. (1998) The molecular genetics of the long QT syndrome: genes causing fainting and sudden death. *Annual Review Of Medicine* 49 :263-74 (PMID: 9509262)
4. Arnestad et al. (2007) Prevalence of long-QT syndrome gene variants in sudden infant death syndrome. *Circulation* 115 (3):361-7 (PMID: 17210839)
5. Priori et al. (2013) Executive summary: HRS/EHRA/APHRS expert consensus statement on the diagnosis and management of patients with inherited primary arrhythmia syndromes. *Europace* 15 (10):1389-406 (PMID: 23994779)
6. Priori et al. (2015) ESC Guidelines for the management of patients with ventricular arrhythmias and the prevention of sudden cardiac death: The Task Force for the Management of Patients with Ventricular Arrhythmias and the Prevention of Sudden Cardiac Death of the European Society of Cardiology (ESC)Endorsed by: Association for European Paediatric and Congenital Cardiology (AEPC). *Europace* 17 (11):1601-87 (PMID: 26318695)
7. Ackerman et al. (2011) HRS/EHRA expert consensus statement on the state of genetic testing for the channelopathies and cardiomyopathies this document was developed as a partnership between the Heart Rhythm Society (HRS) and the European Heart Rhythm Association (EHRA). Heart Rhythm : The Official Journal Of The Heart Rhythm Society 8 (8):1308-39 (PMID: 21787999)
8. Epstein et al. (2013) 2012 ACCF/AHA/HRS focused update incorporated into the ACCF/AHA/HRS 2008 guidelines for device-based therapy of cardiac rhythm abnormalities: a report of the American College of Cardiology Foundation/American Heart Association Task Force on Practice