**Letter of Medical Necessity for Catecholaminergic Polymorphic Ventricular Tachycardia (CPVT) Panel**

**Patient Information**

**Date:**

**Patient Name:**

**Patient DOB:**

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

**Policy Number:**

**Group Number:**

**ICD10 Codes:**

**Test Information**

**Test Name:** Catecholaminergic Polymorphic Ventricular Tachycardia (CPVT) Panel

**CPT Codes:** 81403x1, 81405x1, 81408x1

**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 Catecholaminergic Polymorphic Ventricular Tachycardia (CPVT) Panel to 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 CPVT 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.

CPVT is a potentially fatal cardiac arrhythmia in individuals with a structurally normal heart. In patients with CPVT, the stress-induced release of catecholamines causes a dysfunction of calcium-ion channels in myocytes that induces ventricular arrhythmias.1 Spontaneous recovery from the arrhythmia is possible, but the ventricular tachycardia can progress to ventricular fibrillation and sudden death.1 Symptoms, including syncope, dizziness, arrhythmia and sudden cardiac arrest/death, typically begin in the first decade of life and may be triggered by physical activity or intense emotion, although some individuals experience cardiac events following normal wakeful activities.2 Diagnosis can prove difficult due to normal echocardiogram and electrocardiogram in a resting state. Cardiac testing must be performed under stress-inducing conditions in order to accurately evaluate a possible diagnosis. Although the incidence of CPVT within the population is not precisely known, it is estimated to be 1:10,000.3

The diagnosis of arrhythmias, including CPVT, may 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 CPVT and other arrhythmias:

* The HRS/EHRA Expert Consensus Statement on the State of Genetic Testing for the Channelopathies and Cardiomyopathies states that comprehensive or targeted CPT genetic testing is recommended for patients with clinical suspicion of CPVT.6

**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. Treatment for arrhythmia and surveillance for progression is critical and is strongly influenced by knowledge of the underlying genetic cause.1,2,4,5 Molecular genetic testing is critical to aid patient management in a cost-effective way and to minimize morbidity and mortality.

Management of CPVT is summarized in a specific consensus document from the Heart Rhythm Association (HRS) and the European Heart Rhythm Association (EHRA) and summarized in the recent version of the European Society of Cardiology (ESC) guidelines on ventricular arrhythmias.4,5 Recommendations include beta blockers and ICD placement but also treatment with Flecainide and left cardiac sympathetic denervation (LCSD), which are unique to this condition.1

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

**Summary**

The Catecholaminergic Polymorphic Ventricular Tachycardia (CPVT) Panel at 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. Napolitano C, Priori SG, Bloise R. Catecholaminergic Polymorphic Ventricular Tachycardia. 2004 Oct 14 [Updated 2014 Mar 6]. In: Pagon RA, Adam MP, Ardinger HH, et al., editors. GeneReviews® [Internet]. Seattle (WA): University of Washington, Seattle; 1993-2015. Available from: <http://www.ncbi.nlm.nih.gov/books/NBK1289/>
2. Roston et al. (2018) The clinical and genetic spectrum of catecholaminergic polymorphic ventricular tachycardia: findings from an international multicentre registry. *Europace* 20 (3):541-547 (PMID: 28158428
3. Liu et al. (2008) Catecholaminergic polymorphic ventricular tachycardia. *Prog Cardiovasc Dis* 51 (1):23-30 (PMID: 18634915)
4. 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)
5. 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)
6. 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)