**Letter of Medical Necessity for the Dilated Cardiomyopathy (DCM)/Left Ventricular Non-compaction (LVNC) Panel**

**Patient Information**

**Date:**

**Patient Name:**

**Patient DOB:**

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

**Policy Number:**

**Group Number:**

**ICD10 Codes:**

**Test Information**

**Test Name:** Dilated Cardiomyopathy (DCM)/Left Ventricular Non-compaction (LVNC) Panel

**CPT Codes:** 81405x1, 81406x1, 81407x2

**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 Dilated Cardiomyopathy (DCM)/Left Ventricular Non-compaction (LVNC) 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 DCM/LVNC 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.

DCM and LVNC are diseases of the heart muscle that have overlapping clinical presentations. Most individuals present with one or more of the following: heart failure with symptoms of congestion (edema, orthopnea or paroxysmal dyspnea); reduced cardiac output, resulting in fatigue or dyspnea on exertion, arrhythmias and/or conduction system disease; thromboembolic disease or stroke, mainly from left ventricular mural thrombus.1,2,3 However, there is a broad range of clinical severity, from asymptomatic disease to progressive deterioration of cardiac function and even sudden cardiac death.

Diagnosis of DCM and LVNC can most often be established with noninvasive cardiac imaging, including echocardiography and/or cardiac magnetic resonance imaging (cardiac MRI). DCM is characterized by left ventricular enlargement and systolic dysfunction, resulting in a reduction in the myocardial force of contraction. LVNC is characterized by abnormal trabeculations in the left ventricle, most frequently at the apex. While DCM and LVNC can often be differentiated by cardiac imaging, differentiating between the two disorders can be challenging in some cases.4 Additionally, imaging results may be absent, subtle, or non-specific. Therefore, molecular diagnosis with genetic testing aids in diagnosis, management and establishing recurrence risk for family members. Dilated cardiomyopathy can be inherited in an autosomal dominant, autosomal recessive, X-linked, or mitochondrial manner.

Multiple national and international medical societies have published guidelines that recommend genetic testing for DCM and other cardiomyopathies:

* In 2018, the Heart Failure Society of America (HFSA) published a guideline in conjunction with the American College of Medical Genetics and Genomics (ACMG) that recommends genetic testing for cardiomyopathies using multi-gene testing panels. The recommendation cites studies demonstrating the cost-effectiveness of genetic testing, the importance of results in determining specific interventions that can improve survival and reduce morbidity, and the benefits of cascade screening for family members.10 Genetic testing for LVNC is recommended when it occurs in conjunction with DCM or other cardiomyopathies.10
* The Heart Rhythm Society / European Heart Rhythm Association (HRS/EHRA) Expert Consensus Statement on the State of Genetic Testing for the Channelopathies and Cardiomyopathies states that comprehensive or targeted DCM genetic testing is recommended, and comprehensive or targeted genetic testing can be useful for patients satisfying task force diagnostic criteria LVNC.11

**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 cardiomyopathy and surveillance for progression is critical and is strongly influenced by knowledge of the underlying genetic cause.1,2

Management for dilated cardiomyopathy is summarized in specific consensus documents from the American College of Cardiology Foundation / American Heart Association (ACCF/AHA), the European Society of Cardiology (ESC), the Heart Failure Society of America (HFSA).12-14 Patients with DCM can be offered pharmacological treatments such as beta blockers and L-type calcium channel blockers, as well as prevention of primary manifestations such as ICD therapy for patients at increased risk for sudden cardiac death.1 Additionally, knowledge of the specific genetic cause may impact decisions about the timing of treatment.5 Cardiac transplantation for progressive disease and additional lifestyle recommendations such as contraindications for pregnancy and pregnancy management to optimize outcomes for both the mother and child must also be considered.1,3 DCM/LVNC can also be a presenting feature of other inherited disorders, such as Danon disease, Carvajal syndrome, mitochondrial myopathy, or Emery-Dreifuss muscular dystrophy.2,6,7,8,9 These disorders, which may be subtle or difficult to diagnose without genetic testing, require further medical management, screening, and diagnosis.

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

**Summary**

The Dilated Cardiomyopathy (DCM)/Left Ventricular Non-compaction (LVNC) 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:

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2. Callis et al. (2010) Evolving molecular diagnostics for familial cardiomyopathies: at the heart of it all. *Expert Review Of Molecular Diagnostics* 10 (3):329-51 (PMID: 20370590)
3. Yancy et al. (2013) 2013 ACCF/AHA guideline for the management of heart failure: a report of the American College of Cardiology Foundation/American Heart Association Task Force on Practice Guidelines. *J. Am. Coll. Cardiol.* 62 (16):e147-239 (PMID: 23747642)
4. Tarando et al. (2017) Left ventricular non-compaction and idiopathic dilated cardiomyopathy: the significant diagnostic value of longitudinal strain. *Int J Cardiovasc Imaging* 33 (1):83-95 (PMID: 27659478)
5. Japp et al. (2016) The Diagnosis and Evaluation of Dilated Cardiomyopathy. *J. Am. Coll. Cardiol.* 67 (25):2996-3010 (PMID: 27339497)
6. Bennett et al. (2016) The Current Approach to Diagnosis and Management of Left Ventricular Noncompaction Cardiomyopathy: Review of the Literature. *Cardiology Research and Practice* 2016:5172308 (PMID: 26881173)
7. Rooms et al. (2015) Non-compaction cardiomyopathy: a genetically and clinically heterogeneous disorder. Acta Cardiol 70 (6):625-31 (PMID: 26717209)
8. Yang et al. (2005) Circulation 112 (11):1612-7 (PMID: 16144992)
9. D'souza et al. (2014) Danon disease: clinical features, evaluation, and management. *Circulation. Heart Failure* 7 (5):843-9 (PMID: 25228319)
10. Hershberger et al. (2018) Genetic Evaluation of Cardiomyopathy-A Heart Failure Society of America Practice Guideline*. J. Card. Fail*. 24 (5):281-302 (PMID: 29567486)
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12. Gersh et al. (2011) 2011 ACCF/AHA guideline for the diagnosis and treatment of hypertrophic cardiomyopathy: a report of the American College of Cardiology Foundation/American Heart Association Task Force on Practice Guidelines. J. Thorac. Cardiovasc. Surg. 142 (6):e153-203 (PMID: 22093723)
13. Yancy et al. (2016) ACC/AHA/HFSA Focused Update on New Pharmacological Therapy for Heart Failure: An Update of the 2013 ACCF/AHA Guideline for the Management of Heart Failure: A Report of the American College of Cardiology/American Heart Association Task Force on Clinical Practice Guidelines and the Heart Failure Society of America. Circulation 134 (13):e282-93 (PMID: 27208050)
14. Ponikowski et al. (2016) ESC Guidelines for the diagnosis and treatment of acute and chronic heart failure: The Task Force for the diagnosis and treatment of acute and chronic heart failure of the European Society of Cardiology (ESC). Developed with the special contribution of the Heart Failure Association (HFA) of the ESC. Eur. J. Heart Fail. 18 (8):891-975 (PMID: 27207191)