**Letter of Medical Necessity for the Arrhythmogenic Right Ventricular Dysplasia/Cardiomyopathy (ARVC) Panel**

**Patient Information**

**Date:**

**Patient Name:**

**Patient DOB:**

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

**Policy Number:**

**Group Number:**

**ICD10 Codes:**

**Test Information**

**Test Name:** Arrhythmogenic Right Ventricular Dysplasia/Cardiomyopathy (ARVC) Panel

**CPT Codes:** 81406x5

**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 Arrhythmogenic Right Ventricular Dysplasia/Cardiomyopathy (ARVC) 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 Arrhythmogenic Right Ventricular Dysplasia/Cardiomyopathy (ARVC) 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.

ARVC is a disorder of the intracellular desmosomal junctions of cardiomyocytes, which are responsible for providing and maintaining cell-to-cell adhesion.1 Cardiomyocyte death and progressive fibro-fatty replacement of the right ventricular myocardium are pathognomonic hallmarks of ARVC, which predispose to ventricular tachyarrhythmia and sudden cardiac death (SCD). Clinical findings are limited to the heart in most individuals, although there are some genetic syndromes associated with ARVC and other clinical findings. The disease prevalence is estimated at 1:1000 to 1:2500, but may be higher in certain populations because of non-diagnosed or misdiagnosed cases.

Patients with ARVC typically develop symptoms between the second and fifth decade of life (mean age at diagnosis 31 years), but the age of onset is widely variable.1,2 The most common presenting symptoms of ARVC are heart palpitations, syncope, and SCD. Sometimes, SCD is the first presenting symptom, particularly in young people and athletes. Many patients with ARVC are asymptomatic and diagnosed only after a routine electrocardiogram (ECG). Therefore, disease presentation and severity is also variable.

Diagnostic criteria for ARVC were established by McKenna et al. in 1994 and revised in 2010.3,4 Diagnosis using these criteria is based on genetic, electrocardiographic, structural, and functional findings. The diagnosis can 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. Genetic predispositions to arrhythmias 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 ARVC 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 ARVC 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.8
* 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 ARVC genetic testing can be useful for patients satisfying task force diagnostic criteria for ARVC and LVNC.5,6,9

**Patient Clinical Utility and Medical Management Implications**

This testing is requested because the results 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.1,5,6,7 Molecular genetic testing is critical to aid patient management in a cost-effective way and to minimize morbidity and mortality.

Management of ARVC is summarized in specific consensus documents from the American College of Cardiology / American Heart Association (ACC/AHA) and the European Society of Cardiology (ESC) guidelines.6,10 The recommendations for clinical management and prevention of sudden cardiac death in patients with ARVC define subgroups based on clinical history that correlate to physical exercise management, follow-up imaging studies, and ICD indications.6,7,10

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

**Summary**

The Arrhythmogenic Right Ventricular Dysplasia/Cardiomyopathy (ARVC) 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. Nava A, Bauce B, Basso C, Muriago M, et al.. Clinical profile and long-term follow-up of 37 families with arrhythmogenic right ventricular cardiomyopathy. J Am Coll Cardiol. 2000; 36: 2226-33. (PMID: 11127465)
3. McKenna WJ, Thiene G, Nava A, et al. Diagnosis of arrhythmogenic right ventricular dysplasia / cardiomyopathy. Task Force of the Working Group Myocardial and Pericardial Disease of the European Society of Cardiology and of the Scientific Council on Cardiomyopathies of the International Society and Federation of Cardiology. Br Heart J. 1994: 71: 215-8 (PMID: 8142187)
4. Marcus FI et al. Diagnosis of arrhythmogenic right ventricular cardiomyopathy/dysplasia: Proposed modification of the Task Force Criteria. Eur Heart J. 31:806-814, 2010 (PMID: 20172912)
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) 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. Brugada & Fernandez-Armenta (2012) E-Journal of Cardiology Practice. 10(25) Available from: https://www.escardio.org/Journals/E-Journal-of-Cardiology-Practice/Volume-10/Arrhythmogenic-right-ventricular-dyplasia (Accessed 4/14/2017).
8. 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).
9. 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)
10. 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 Guidelines and the Heart Rhythm Society. Journal Of The American College Of Cardiology. J. Am. Coll. Cardiol. 61 (3):e6-75 (PMID: 23265327)