LONG QT SYNDROME PANELS

I. Genetic testing for long QT syndrome (LQTS) via multigene panel (81403, 81406, 81407, 81413, 81414, 81479) is considered **medically necessary** when:

   A. The member is asymptomatic, **AND**

      1. The member has a confirmed prolonged QTc (greater than 460ms prepuberty, greater than 480 ms for adults) on resting ECG and/or provocative stress testing with exercise or during intravenous pharmacologic provocation testing (eg, with epinephrine), **OR**

      2. The member has a close relative with a clinical diagnosis of LQTS, whose genetic status is unknown, **OR**

   B. The member is symptomatic (for example: a history of syncope, cardiac arrest, and/or aborted sudden death), **AND**

      1. The member meets either of the following:

         a) A cardiologist has established a strong clinical suspicion for LQTS based on examination of the patient's clinical history, family history, and expressed electrographic phenotype, **OR**

         b) The member has a Schwartz score of 3.0 or more, **AND**

      2. Non-genetic causes of a prolonged QTc interval have been ruled out, such as QT-prolonging drugs, hypokalemia, structural heart disease, or certain neurologic conditions including subarachnoid bleed.

II. Genetic testing for long QT syndrome (LQTS) via multigene panel (81403, 81406, 81407, 81413, 81414, 81479) is considered **investigational** for all other indications.

NOTE: If a panel is performed, the appropriate panel code should be used

DEFINITIONS

1. **Close relatives** include first, second, and third degree blood relatives:
    a. **First-degree relatives** are parents, siblings, and children
    b. **Second-degree relatives** are grandparents, aunts, uncles, nieces, nephews, grandchildren, and half siblings
    c. **Third-degree relatives** are great grandparents, great aunts, great uncles, great grandchildren, and first cousins

REFERENCES

1. Ackerman MJ, Priori SG, Willems S, et al. 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. 2011;8(8):1308-1339. doi:10.1016/j.hrthm.2011.05.020
2. Schwartz PJ, Crotti L. QTc behavior during exercise and genetic testing for the long-QT syndrome. Circulation. 2011 Nov 15;124(20):2181-4. doi: 10.1161/CIRCULATIONAHA.111.062182. PMID: 22083145
3. Priori SG, Wilde AA, Horie M, et al. HRS/EHRA/APHRS expert consensus statement on the diagnosis and management of patients with inherited primary arrhythmia syndromes. Heart Rhythm. 2013;10(12):1932-1963.