Long QT Syndrome Panels

I. Genetic testing for long QT syndrome (LQTS) via multigene panel 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,[1] **OR**

   B. The member is symptomatic (e.g., 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 member'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 is considered **investigational** for all other indications.

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

[1]If a pathogenic or likely pathogenic variant has been identified in an explanatory gene in the affected family member, refer to the *General Criteria for Known Familial Variant Analysis for a Genetic Condition* within the *General Approach to Laboratory Testing.*

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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

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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. Wilde AAM, Semsarian C, Márquez MF, et al. European Heart Rhythm Association/Heart Rhythm Society/Asia Pacific Heart Rhythm Society/Latin American Heart Rhythm Society expert consensus statement on the state of genetic testing for cardiac diseases. [published correction appears in Europace. 2022 Aug 30]. Europace. 2022;24(8):1307-1367. doi:10.1093/europace/euac030

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