STEMOD™ LQT1 Disease Model (KCNQ1 (KVLQT1, Kv7.1), G345E (G1034A))

Cat. No.: MoCVD-ZXY004

This product is for research use only and is not intended for diagnostic use.

PRODUCT INFORMATION

Product Overview

Long-QT syndromes are heritable diseases associated with prolongation of the QT interval on an electrocardiogram and a high risk of sudden cardiac death due to ventricular tachyarrhythmia. This disease cell model belongs to long-QT syndrome type 1, mutation (G345E, also called G1034A) occur in the KCNQ1 gene. Generating pluripotent stem cells from dermal fibroblasts, then differentiate into cardiac myocytes. This disease cell model is contributed to as a new platform for drug testing.

SPECIFICATIONS

Organ System          Cardiovascular System
Disease               LQT1
Target Gene           KCNQ1 (KVLQT1, Kv7.1)
Gene Function         KCNQ1 encodes the repolarizing potassium channel mediating the delayed rectifier Iks current, pore-forming α subunit of the channel generating the slow component of the delayed rectifier potassium current Iks
Mutation              G345E (G1034A)
Phenotype             Prolonged action potential duration (APD)
Source                Dermal fibroblasts
Cellular Assays       Whole-cell patch clamp techniques, immunostaining
<table>
<thead>
<tr>
<th><strong>Gene Information</strong></th>
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<td><strong>Gene ID</strong></td>
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<td><strong>Uniprot ID</strong></td>
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