STEMOD™ LQT1 Disease Model (KCNQ1 (KVLQT1, Kv7.1), c.922~1,032 del, p.308~344 del)

Cat. No.: MoCVD-ZXY006

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 (C.922 ~ 1,032 del; P.308 ~ 344 del) occur in the KCNQ1 gene. Generating pluripotent stem cells from dermal fibroblasts, then differentiate into cardiac myocytes. This disease cell model provides a platform to explore disease mechanisms in human genetic cardiac disorders.

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  c.922~1,032 del, p.308~344 del
Phenotype  Prolonged action potential duration (APD), reduced Iks
Source  Dermal fibroblasts
Cellular Assays  Whole-cell patch clamp techniques, immunostaining
### GENE INFORMATION

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