STEMOD™ LQT2 Disease Model (KCNH2, A614V (C821T))

Cat. No.: MoCVD-ZXY011

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 2, mutation (A614V, also called A614V) occur in the KCNH2 gene. Generating pluripotent stem cells from dermal fibroblasts, then differentiate into cardiac myocytes. This disease cell model is contributed to as a new platform to study pathophysiological mechanisms and drug testing.

SPECIFICATIONS

Organ System  Cardiovascular System
Disease  LQT2
Target Gene  KCNH2
Gene Function  KCNH2, also known as human Ether-a-go-go Related Gene and HERG, forms the α-subunit of the rapid-acting inward rectifying potassium (Ikr) channel.
Mutation  A614V (C821T)
Phenotype  Prolonged action potential duration (APD), reduced IKr
Source  Dermal fibroblasts
Cellular Assays  Whole-cell patch clamp techniques, immunostaining

GENE INFORMATION
Gene ID  3757
Uniprot ID  Q12809