BACKGROUND

Members of the superfamily of voltage-gated cation channels open and close in response to changes in the voltage across the cell membrane to generate electrical currents that underlie neuronal signaling and skeletal and cardiac muscle contraction. The voltage-gated openings of these channels are controlled by intrinsic voltage sensors, including several positively charged residues in the fourth transmembrane (S4) segment of the channel, which move in response to changes in the membrane voltage. The voltage sensor movements trigger opening of the channel gate through a coupling mechanism that is not well-understood, and which may vary between specific channels and channel complexes. A fundamental understanding of how voltage sensors move and how this movement couples to channel opening is important to understand how these channels function during physiological and pathophysiological conditions and how mutation of these channels and their associated accessory subunits causes disease. One channel whose gating properties are of particular interest is the I_{Ks} potassium channel, a slowly activating channel that is essential to normal cardiac function. That the unique biophysical properties of the I_{Ks} channel are important for normal cardiac physiology is evidenced by multiple pathophysiological clinical phenotypes associated with mutations that change the biophysical and regulatory properties of the I_{Ks} channel, such as long QT syndrome, short QT syndrome, and familial atrial fibrillation.[1] The I_{Ks} channel consists of an α-subunit (KCNQ1, also known as Kv7.1) and an accessory β-subunit (KCNE1). KCNQ1 belongs to the canonical voltage-gated potassium channel family, forming a homotetrameric channel with a central pore domain and four peripheral voltage-sensing domains. KCNE1 is a small (129-amino acid) single-pass transmembrane protein. Although four KCNQ1 subunits assemble to form functional tetrameric voltage-gated channels, the biophysical and regulatory properties of the KCNQ1 channel when expressed alone are completely distinct from I_{Ks} currents: KCNQ1 homomeric channels activate and deactivate rapidly and begin to open at voltages more negative than those that activate I_{Ks} channels. Coassembly of KCNE1 with KCNQ1 drastically slows the activation kinetics, shifts the voltage dependence of activation, slows deactivation, and increases single-channel conductance, thereby reproducing the critical properties of the native I_{Ks} channel.[2]

The *KCNQ1* gene has a total of 17 exons, spans 404 kb of chromosome sequence and is located on chromosome 11p15.5. *KCNQ1* codes for the pore-forming alpha subunit of the voltage-gated K+ channel (KvLQT1) that is highly expressed in the heart. This channel plays an important role in controlling repolarization of the ventricles, where its primary function is to limit action potential prolongation during sympathetic stimulation.

Mutations in *KCNQ1* have been described to lead to cardiac long QT syndrome, Jervell and Lange-Nielsen syndrome, which are associated with cardiac conduction abnormalities and hearing loss. *KCNQ1* is also expressed to lesser extent in the pancreas, placenta, lung, liver, kidney, brain, and adipose tissue. In addition, *KCNQ1* is expressed *in vitro* in insulin-secreting cell lines. Insulin secretion from pancreatic β cells is regulated by complex interplay between K_{ATP} channels and K_{v}- channels and voltage-dependent Ca^{++} channels. Ionic mechanisms at K_{ATP} and K_{v}- channels are primarily important in triggering and maintaining glucose-stimulated insulin secretion and inhibition of this potassium channel has been shown to significantly increase insulin secretion. It was shown that that the variation within the *KCNQ1* locus confers a significant risk to type II diabetes (T2D).[3]

References:
1. Lundby, A. et al: Heart Rhythm 7:708-13, 2010
2. Osteen, J.D. et al: Proc. Natl. Acad. Sci. USA 107:22710–5, 2010
3. Been, J.F. et al: BMC Med. Genet. 12:18, 2011

TECHNICAL INFORMATION

Source:
KCNQ1 Antibody is a mouse monoclonal antibody raised against recombinant human KCNQ1 fragments expressed in *E. coli*.

Specificity and Sensitivity:
This antibody detects endogenous KCNQ1 proteins without cross-reactivity with other family members.

Storage Buffer: PBS and 30% glycerol

Storage:
Store at -20°C for at least one year. Store at 4°C for frequent use. Avoid repeated freeze-thaw cycles.

APPLICATIONS

Application:	*Dilution:
WB	1:1000
IP	n/d
IHC	1:50-200
ICC	1:50-200
FACS	1:50-200

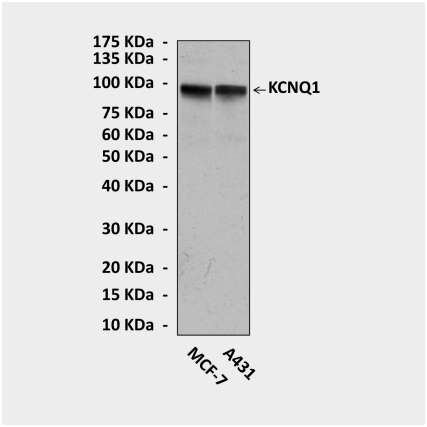
**Optimal dilutions must be determined by end user.*

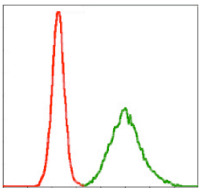
KCNQ1 Antibody

Cat. No. CP10438

Applications:	**WB, FACS**
Detected MW:	**95 kDa**
Species & Reactivity:	**Human, Mouse, Rat**
Isotype:	**Mouse IgG1**

QUALITY CONTROL DATA





Top: Western Blot detection of KCNQ1 proteins in MCF-7 and A431 cell lysates using KCNQ1 Antibody. **Bottom:** This antibody specifically reacts with KCNQ1 proteins in MCF7 cells detected by FACS (KCNQ1 Antibody: Green; Mouse IgG control: Red).