

Supplementary references

- Abbott, G.W. (2016). Regulation of human cardiac potassium channels by full-length KCNE3 and KCNE4. *Sci. Rep.* *6*, 38412.
- Aguilar, M.B., Pérez-Reyes, L.I., López, Z., de la Cotera, E.P.H., Falcón, A., Ayala, C., Galván, M., Salvador, C., and Escobar, L.I. (2010). Peptide sr11a from Conus spurius is a novel peptide blocker for Kv1 potassium channels. *Peptides* *31*, 1287–1291.
- Akhtar, S., McIntosh, P., Bryan-Sisneros, A., Barratt, L., Robertson, B., and Dolly, J.O. (1999). A functional spliced-variant of beta 2 subunit of Kv1 channels in C6 glioma cells and reactive astrocytes from rat lesioned cerebellum. *Biochemistry* *38*, 16984–16992.
- Akhtar, S., Shamotienko, O., Papakosta, M., Ali, F., and Dolly, J.O. (2002). Characteristics of brain Kv1 channels tailored to mimic native counterparts by tandem linkage of alpha subunits: implications for K⁺ channelopathies. *J. Biol. Chem.* *277*, 16376–16382.
- Al-Sabi, A., Kaza, S.K., Dolly, J.O., and Wang, J. (2013). Pharmacological characteristics of Kv1.1- and Kv1.2-containing channels are influenced by the stoichiometry and positioning of their α subunits. *Biochem. J.* *454*, 101–108.
- Aréchiga-Figueroa, I.A., Delgado-Ramírez, M., Morán-Zendejas, R., and Rodríguez-Menchaca, A.A. (2015). Modulation of Kv2.1 channels inactivation by curcumin. *Pharmacol. Rep. PR* *67*, 1273–1279.
- Bähring, R., Vardanyan, V., and Pongs, O. (2004). Differential modulation of Kv1 channel-mediated currents by co-expression of Kvbeta3 subunit in a mammalian cell-line. *Mol. Membr. Biol.* *21*, 19–25.
- Baranauskas, G., Tkatch, T., Nagata, K., Yeh, J.Z., and Surmeier, D.J. (2003). Kv3.4 subunits enhance the repolarizing efficiency of Kv3.1 channels in fast-spiking neurons. *Nat. Neurosci.* *6*, 258–266.
- Bardien-Kruger, S., Wulff, H., Arief, Z., Brink, P., Chandy, K.G., and Corfield, V. (2002). Characterisation of the human voltage-gated potassium channel gene, KCNA7, a candidate gene for inherited cardiac disorders, and its exclusion as cause of progressive familial heart block I (PFHBI). *Eur. J. Hum. Genet. EJHG* *10*, 36–43.
- Beck, E.J., Bowlby, M., An, W.F., Rhodes, K.J., and Covarrubias, M. (2002). Remodelling inactivation gating of Kv4 channels by KChIP1, a small-molecular-weight calcium-binding protein. *J. Physiol.* *538*, 691–706.
- Bentzen, B.H., Schmitt, N., Calloe, K., Dalby Brown, W., Grunnet, M., and Olesen, S.-P. (2006). The acrylamide (S)-1 differentially affects Kv7 (KCNQ) potassium channels. *Neuropharmacology* *51*, 1068–1077.
- Bertoli, A., Moran, O., and Conti, F. (1994). Activation and deactivation properties of rat brain K⁺ channels of the Shaker-related subfamily. *Eur. Biophys. J. EBj* *23*, 379–384.
- Blaine, J.T., and Ribera, A.B. (1998). Heteromultimeric potassium channels formed by members of the Kv2 subfamily. *J. Neurosci. Off. J. Soc. Neurosci.* *18*, 9585–9593.
- Blom, S.M., Schmitt, N., and Jensen, H.S. (2009). The acrylamide (S)-2 as a positive and negative modulator of Kv7 channels expressed in *Xenopus laevis* oocytes. *PloS One* *4*, e8251.
- Blom, S.M., Rottländer, M., Kehler, J., Bundgaard, C., Schmitt, N., and Jensen, H.S. (2014). From pan-reactive KV7 channel opener to subtype selective opener/inhibitor by addition of a methyl group. *PloS One* *9*, e100209.

- Brown, M.R., El-Hassar, L., Zhang, Y., Alvaro, G., Large, C.H., and Kaczmarek, L.K. (2016). Physiological modulators of Kv3.1 channels adjust firing patterns of auditory brain stem neurons. *J. Neurophysiol.* *116*, 106–121.
- Chen, S.-H., Fu, S.-J., Huang, J.-J., and Tang, C.-Y. (2016). The episodic ataxia type 1 mutation I262T alters voltage-dependent gating and disrupts protein biosynthesis of human Kv1.1 potassium channels. *Sci. Rep.* *6*, 19378.
- Choveau, F.S., Hernandez, C.C., Bierbower, S.M., and Shapiro, M.S. (2012a). Pore determinants of KCNQ3 K⁺ current expression. *Biophys. J.* *102*, 2489–2498.
- Choveau, F.S., Bierbower, S.M., and Shapiro, M.S. (2012b). Pore helix-S6 interactions are critical in governing current amplitudes of KCNQ3 K⁺ channels. *Biophys. J.* *102*, 2499–2509.
- Chuang, C.-C., Jow, G.-M., Lin, H.-M., Weng, Y.-H., Hu, J.-H., Peng, Y.-J., Chiu, Y.-C., Chiu, M.-M., and Jeng, C.-J. (2014). The punctate localization of rat Eag1 K⁺ channels is conferred by the proximal post-CNBHD region. *BMC Neurosci.* *15*, 23.
- Clancy, S.M., Chen, B., Bertaso, F., Mamet, J., and Jegla, T. (2009). KCNE1 and KCNE3 beta-subunits regulate membrane surface expression of Kv12.2 K(+) channels in vitro and form a tripartite complex in vivo. *PLoS One* *4*, e6330.
- D'Adamo, M.C., Gallenmüller, C., Servettini, I., Hartl, E., Tucker, S.J., Arning, L., Biskup, S., Grottesi, A., Guglielmi, L., Imbrici, P., et al. (2014). Novel phenotype associated with a mutation in the KCNA1(Kv1.1) gene. *Front. Physiol.* *5*, 525.
- Diochot, S., Schweitz, H., Béress, L., and Lazdunski, M. (1998). Sea anemone peptides with a specific blocking activity against the fast inactivating potassium channel Kv3.4. *J. Biol. Chem.* *273*, 6744–6749.
- Dong, W.-H., Chen, J.-C., He, Y.-L., Xu, J.-J., and Mei, Y.-A. (2013). Resveratrol inhibits K(v)2.2 currents through the estrogen receptor GPR30-mediated PKC pathway. *Am. J. Physiol. Cell Physiol.* *305*, C547–557.
- Elinder, F., Madeja, M., and Arhem, P. (1996). Surface Charges of K channels. Effects of strontium on five cloned channels expressed in Xenopus oocytes. *J. Gen. Physiol.* *108*, 325–332.
- Ferber, M., Al-Sabi, A., Stocker, M., Olivera, B.M., and Terlau, H. (2004). Identification of a mammalian target of kappaM-conotoxin RIIK. *Toxicon Off. J. Int. Soc. Toxinology* *43*, 915–921.
- Fernandez, F.R., Morales, E., Rashid, A.J., Dunn, R.J., and Turner, R.W. (2003). Inactivation of Kv3.3 potassium channels in heterologous expression systems. *J. Biol. Chem.* *278*, 40890–40898.
- Finol-Urdaneta, R.K., Strüver, N., and Terlau, H. (2006). Molecular and Functional Differences between Heart mKv1.7 Channel Isoforms. *J. Gen. Physiol.* *128*, 133–145.
- Gamper, N., Stockand, J.D., and Shapiro, M.S. (2003). Subunit-specific modulation of KCNQ potassium channels by Src tyrosine kinase. *J. Neurosci. Off. J. Soc. Neurosci.* *23*, 84–95.
- García-Fernández, R., Peigneur, S., Pons, T., Alvarez, C., González, L., Chávez, M.A., and Tytgat, J. (2016). The Kunitz-Type Protein ShPI-1 Inhibits Serine Proteases and Voltage-Gated Potassium Channels. *Toxins* *8*, 110.
- Gilling, M., Rasmussen, H.B., Calloe, K., Sequeira, A.F., Baretto, M., Oliveira, G., Almeida, J., Lauritsen, M.B., Ullmann, R., Boonen, S.E., et al. (2013). Dysfunction of the Heteromeric KV7.3/KV7.5 Potassium Channel is Associated with Autism Spectrum Disorders. *Front. Genet.* *4*, 54.

- Gómez-Hernandez, J.M., Lorra, C., Pardo, L.A., Stühmer, W., Pongs, O., Heinemann, S.H., and Elliott, A.A. (1997). Molecular basis for different pore properties of potassium channels from the rat brain Kv1 gene family. *Pflugers Arch.* **434**, 661–668.
- Guighard, G., Bellocq, C., Grelet, E., and Escande, D. (2003). Human Kv1.6 current displays a C-type-like inactivation when re-expressed in cos-7 cells. *Biochem. Biophys. Res. Commun.* **311**, 83–89.
- Hatton, W.J., Mason, H.S., Carl, A., Doherty, P., Latten, M.J., Kenyon, J.L., Sanders, K.M., and Horowitz, B. (2001). Functional and molecular expression of a voltage-dependent K(+) channel (Kv1.1) in interstitial cells of Cajal. *J. Physiol.* **533**, 315–327.
- Heinemann, S.H., Rettig, J., Graack, H.R., and Pongs, O. (1996). Functional characterization of Kv channel beta-subunits from rat brain. *J. Physiol.* **493**, 625–633.
- Hernández-Pineda, R., Chow, A., Amarillo, Y., Moreno, H., Saganich, M., Vega-Saenz de Miera, E.C., Hernández-Cruz, A., and Rudy, B. (1999). Kv3.1-Kv3.2 channels underlie a high-voltage-activating component of the delayed rectifier K⁺ current in projecting neurons from the globus pallidus. *J. Neurophysiol.* **82**, 1512–1528.
- Honoré, E., Attali, B., Romey, G., Lesage, F., Barhanin, J., and Lazdunski, M. (1992). Different types of K⁺ channel current are generated by different levels of a single mRNA. *EMBO J.* **11**, 2465–2471.
- Hou, P., Zhang, R., Liu, Y., Feng, J., Wang, W., Wu, Y., and Ding, J. (2014). Physiological role of Kv1.3 channel in T lymphocyte cell investigated quantitatively by kinetic modeling. *PloS One* **9**, e89975.
- Hsu, P.-H., Miaw, S.-C., Chuang, C.-C., Chang, P.-Y., Fu, S.-J., Jow, G.-M., Chiu, M.-M., and Jeng, C.-J. (2012). 14-3-3θ is a binding partner of rat Eag1 potassium channels. *PloS One* **7**, e41203.
- Jentsch, T.J. (2000). Neuronal KCNQ potassium channels: physiology and role in disease. *Nat. Rev. Neurosci.* **1**, 21–30.
- Jeong, I., Choi, B.H., and Hahn, S.J. (2011). Rosiglitazone inhibits Kv4.3 potassium channels by open-channel block and acceleration of closed-state inactivation. *Br. J. Pharmacol.* **163**, 510–520.
- Jeong, I., Choi, B.H., Yoon, S.H., and Hahn, S.J. (2012). Carvedilol blocks the cloned cardiac Kv1.5 channels in a β-adrenergic receptor-independent manner. *Biochem. Pharmacol.* **83**, 497–505.
- Jerng, H.H., Shahidullah, M., and Covarrubias, M. (1999). Inactivation gating of Kv4 potassium channels: molecular interactions involving the inner vestibule of the pore. *J. Gen. Physiol.* **113**, 641–660.
- Jerng, H.H., Qian, Y., and Pfaffinger, P.J. (2004). Modulation of Kv4.2 channel expression and gating by dipeptidyl peptidase 10 (DPP10). *Biophys. J.* **87**, 2380–2396.
- Ju, M., and Wray, D. (2002). Molecular identification and characterisation of the human eag2 potassium channel. *FEBS Lett.* **524**, 204–210.
- Kalman, K., Nguyen, A., Tseng-Crank, J., Dukes, I.D., Chandy, G., Hustad, C.M., Copeland, N.G., Jenkins, N.A., Mohrenweiser, H., Brandriff, B., et al. (1998). Genomic organization, chromosomal localization, tissue distribution, and biophysical characterization of a novel mammalian Shaker-related voltage-gated potassium channel, Kv1.7. *J. Biol. Chem.* **273**, 5851–5857.
- Kanda, V.A., Lewis, A., Xu, X., and Abbott, G.W. (2011). KCNE1 and KCNE2 provide a checkpoint governing voltage-gated potassium channel α-subunit composition. *Biophys. J.* **101**, 1364–1375.
- Kanemasa, T., Gan, L., Perney, T.M., Wang, L.Y., and Kaczmarek, L.K. (1995). Electrophysiological and pharmacological characterization of a mammalian Shaw channel expressed in NIH 3T3 fibroblasts. *J. Neurophysiol.* **74**, 207–217.

- Kazmierczak, M., Zhang, X., Chen, B., Mulkey, D.K., Shi, Y., Wagner, P.G., Pivaroff-Ward, K., Sasic, J.K., Bayliss, D.A., and Jegla, T. (2013). External pH modulates EAG superfamily K⁺ channels through EAG-specific acidic residues in the voltage sensor. *J. Gen. Physiol.* **141**, 721–735.
- Kerschensteiner, D., Monje, F., and Stocker, M. (2003). Structural determinants of the regulation of the voltage-gated potassium channel Kv2.1 by the modulatory α -subunit Kv9.3. *J. Biol. Chem.* **278**, 18154–18161.
- Kerschensteiner, D., Soto, F., and Stocker, M. (2005). Fluorescence measurements reveal stoichiometry of K⁺ channels formed by modulatory and delayed rectifier alpha-subunits. *Proc. Natl. Acad. Sci. U. S. A.* **102**, 6160–6165.
- Kihira, Y., Hermanstyne, T.O., and Misonou, H. (2010). Formation of heteromeric Kv2 channels in mammalian brain neurons. *J. Biol. Chem.* **285**, 15048–15055.
- Kim, K.S., Duignan, K.M., Hawryluk, J.M., Soh, H., and Tzingounis, A.V. (2016). The Voltage Activation of Cortical KCNQ Channels Depends on Global PIP2 Levels. *Biophys. J.* **110**, 1089–1098.
- Kinoshita, M., Matsuoka, Y., Suzuki, T., Mirrieles, J., and Yang, J. (2012). Sigma-1 receptor alters the kinetics of Kv1.3 voltage gated potassium channels but not the sensitivity to receptor ligands. *Brain Res.* **1452**, 1–9.
- Kuryshev, Y.A., Wible, B.A., Gudz, T.I., Ramirez, A.N., and Brown, A.M. (2001). KChAP/Kvbeta1.2 interactions and their effects on cardiac Kv channel expression. *Am. J. Physiol. Cell Physiol.* **281**, C290–299.
- Lang, R., Lee, G., Liu, W., Tian, S., Rafi, H., Orias, M., Segal, A.S., and Desir, G.V. (2000). KCNA10: a novel ion channel functionally related to both voltage-gated potassium and CNG cation channels. *Am. J. Physiol. Renal Physiol.* **278**, F1013–1021.
- Lee, H.M., Hahn, S.J., and Choi, B.H. (2016). Blockade of Kv1.5 channels by the antidepressant drug sertraline. *Korean J. Physiol. Pharmacol. Off. J. Korean Physiol. Soc. Korean Soc. Pharmacol.* **20**, 193–200.
- Lee, J.-H., Choi, S.-H., Shin, T.-J., Lee, B.-H., Hwang, S.-H., Kim, H.-C., and Nah, S.-Y. (2011). Effect of dextromethorphan on human K(v)1.3 channel activity: involvement of C-type inactivation. *Eur. J. Pharmacol.* **651**, 122–127.
- Lewis, A., McCrossan, Z.A., and Abbott, G.W. (2004). MinK, MiRP1, and MiRP2 diversify Kv3.1 and Kv3.2 potassium channel gating. *J. Biol. Chem.* **279**, 7884–7892.
- Li, Y., Gamper, N., and Shapiro, M.S. (2004). Single-channel analysis of KCNQ K⁺ channels reveals the mechanism of augmentation by a cysteine-modifying reagent. *J. Neurosci. Off. J. Soc. Neurosci.* **24**, 5079–5090.
- MacDonald, P.E., Salapatek, A.M.F., and Wheeler, M.B. (2003). Temperature and redox state dependence of native Kv2.1 currents in rat pancreatic beta-cells. *J. Physiol.* **546**, 647–653.
- Macica, C.M., von Hehn, C.A.A., Wang, L.-Y., Ho, C.-S., Yokoyama, S., Joho, R.H., and Kaczmarek, L.K. (2003). Modulation of the kv3.1b potassium channel isoform adjusts the fidelity of the firing pattern of auditory neurons. *J. Neurosci. Off. J. Soc. Neurosci.* **23**, 1133–1141.
- Malin, S.A., and Nerbonne, J.M. (2002). Delayed rectifier K⁺ currents, IK, are encoded by Kv2 alpha-subunits and regulate tonic firing in mammalian sympathetic neurons. *J. Neurosci. Off. J. Soc. Neurosci.* **22**, 10094–10105.
- Mauerhöfer, M., and Bauer, C.K. (2016). Effects of Temperature on Heteromeric Kv11.1a/1b and Kv11.3 Channels. *Biophys. J.* **111**, 504–523.

- Mestre, T.A., Manole, A., MacDonald, H., Riazi, S., Kraeva, N., Hanna, M.G., Lang, A.E., Männikkö, R., and Yoon, G. (2016). A novel KCNA1 mutation in a family with episodic ataxia and malignant hyperthermia. *Neurogenetics* *17*, 245–249.
- Meyer, R., and Heinemann, S.H. (1997). Temperature and pressure dependence of Shaker K⁺ channel N- and C-type inactivation. *Eur. Biophys. J. EB* *26*, 433–445.
- Miceli, F., Cilio, M.R., Taglialatela, M., and Bezanilla, F. (2009). Gating currents from neuronal K(V)7.4 channels: general features and correlation with the ionic conductance. *Channels Austin Tex* *3*, 274–283.
- Miyake, A., Mochizuki, S., Yokoi, H., Kohda, M., and Furuichi, K. (1999). New ether-à-go-go K(+) channel family members localized in human telencephalon. *J. Biol. Chem.* *274*, 25018–25025.
- Mock, A.F., Richardson, J.L., Hsieh, J.-Y., Rinetti, G., and Papazian, D.M. (2010). Functional effects of spinocerebellar ataxia type 13 mutations are conserved in zebrafish Kv3.3 channels. *BMC Neurosci.* *11*, 99.
- Moran, O., and Conti, F. (1995). Properties of the Kv1.1 rat brain potassium channels expressed in mammalian cells: temperature effects. *Biochem. Biophys. Res. Commun.* *215*, 915–920.
- Moreno, H., Kentros, C., Bueno, E., Weiser, M., Hernandez, A., Vega-Saenz de Miera, E., Ponce, A., Thornhill, W., and Rudy, B. (1995). Thalamocortical projections have a K⁺ channel that is phosphorylated and modulated by cAMP-dependent protein kinase. *J. Neurosci. Off. J. Soc. Neurosci.* *15*, 5486–5501.
- Mortensen, L.S., Schmidt, H., Farsi, Z., Barrantes-Freer, A., Rubio, M.E., Ufartes, R., Eilers, J., Sakaba, T., Stühmer, W., and Pardo, L.A. (2015). KV 10.1 opposes activity-dependent increase in Ca²⁺ influx into the presynaptic terminal of the parallel fibre-Purkinje cell synapse. *J. Physiol.* *593*, 181–196.
- Mourre, C., Chernova, M.N., Martin-Eauclaire, M.F., Bessone, R., Jacquet, G., Gola, M., Alper, S.L., and Crest, M. (1999). Distribution in rat brain of binding sites of kaliotoxin, a blocker of Kv1.1 and Kv1.3 alpha-subunits. *J. Pharmacol. Exp. Ther.* *291*, 943–952.
- Nakajo, K., and Kubo, Y. (2008). Second coiled-coil domain of KCNQ channel controls current expression and subfamily specific heteromultimerization by salt bridge networks. *J. Physiol.* *586*, 2827–2840.
- Nicolaou, S.A., Szigligeti, P., Neumeier, L., Lee, S.M., Duncan, H.J., Kant, S.K., Mongey, A.B., Filipovich, A.H., and Conforti, L. (2007). Altered dynamics of Kv1.3 channel compartmentalization in the immunological synapse in systemic lupus erythematosus. *J. Immunol. Baltim. Md 1950* *179*, 346–356.
- Noma, K., Kimura, K., Minatohara, K., Nakashima, H., Nagao, Y., Mizoguchi, A., and Fujiyoshi, Y. (2009). Triple N-glycosylation in the long S5-P loop regulates the activation and trafficking of the Kv12.2 potassium channel. *J. Biol. Chem.* *284*, 33139–33150.
- Oliver, K.L., Franceschetti, S., Milligan, C.J., Muona, M., Mandelstam, S.A., Canafoglia, L., Boguszewska-Chachulska, A.M., Korczyn, A.D., Bisulli, F., Di Bonaventura, C., et al. (2017). Myoclonus epilepsy and ataxia due to KCNC1 mutation: Analysis of 20 cases and K⁺ channel properties. *Ann. Neurol.* *81*, 677–689.
- Oliveras, A., Roura-Ferrer, M., Solé, L., de la Cruz, A., Prieto, A., Etxebarria, A., Manils, J., Morales-Cano, D., Condom, E., Soler, C., et al. (2014). Functional assembly of Kv7.1/Kv7.5 channels with emerging properties on vascular muscle physiology. *Arterioscler. Thromb. Vasc. Biol.* *34*, 1522–1530.

- Orhan, G., Bock, M., Schepers, D., Ilina, E.I., Reichel, S.N., Löffler, H., Jezutkovic, N., Weckhuysen, S., Mandelstam, S., Suls, A., et al. (2014). Dominant-negative effects of KCNQ2 mutations are associated with epileptic encephalopathy. *Ann. Neurol.* **75**, 382–394.
- Pahapill, P.A., and Schlichter, L.C. (1990). Modulation of potassium channels in human T lymphocytes: effects of temperature. *J. Physiol.* **422**, 103–126.
- Peretz, A., Pell, L., Gofman, Y., Haitin, Y., Shamgar, L., Patrich, E., Kornilov, P., Gourgy-Hacohen, O., Ben-Tal, N., and Attali, B. (2010). Targeting the voltage sensor of Kv7.2 voltage-gated K⁺ channels with a new gating-modifier. *Proc. Natl. Acad. Sci. U. S. A.* **107**, 15637–15642.
- Pérez-García, M.T., López-López, J.R., and González, C. (1999). Kvbeta1.2 subunit coexpression in HEK293 cells confers O₂ sensitivity to kv4.2 but not to Shaker channels. *J. Gen. Physiol.* **113**, 897–907.
- Po, S., Roberds, S., Snyders, D.J., Tamkun, M.M., and Bennett, P.B. (1993). Heteromultimeric assembly of human potassium channels. Molecular basis of a transient outward current? *Circ. Res.* **72**, 1326–1336.
- Rangaraju, S., Khoo, K.K., Feng, Z.-P., Crossley, G., Nugent, D., Khaytin, I., Chi, V., Pham, C., Calabresi, P., Pennington, M.W., et al. (2010). Potassium channel modulation by a toxin domain in matrix metalloprotease 23. *J. Biol. Chem.* **285**, 9124–9136.
- Rashid, A.J., Morales, E., Turner, R.W., and Dunn, R.J. (2001). The contribution of dendritic Kv3 K⁺ channels to burst threshold in a sensory neuron. *J. Neurosci. Off. J. Soc. Neurosci.* **21**, 125–135.
- Remillard, C.V., Tigno, D.D., Platoshyn, O., Burg, E.D., Brevnova, E.E., Conger, D., Nicholson, A., Rana, B.K., Channick, R.N., Rubin, L.J., et al. (2007). Function of Kv1.5 channels and genetic variations of KCNA5 in patients with idiopathic pulmonary arterial hypertension. *Am. J. Physiol. Cell Physiol.* **292**, C1837–1853.
- Rezazadeh, S., Kurata, H.T., Claydon, T.W., Kehl, S.J., and Fedida, D. (2007). An activation gating switch in Kv1.2 is localized to a threonine residue in the S2-S3 linker. *Biophys. J.* **93**, 4173–4186.
- Rich, T.C., and Snyders, D.J. (1998). Evidence for multiple open and inactivated states of the hKv1.5 delayed rectifier. *Biophys. J.* **75**, 183–195.
- Roeper, J., Sewing, S., Zhang, Y., Sommer, T., Wanner, S.G., and Pongs, O. (1998). NIP domain prevents N-type inactivation in voltage-gated potassium channels. *Nature* **391**, 390–393.
- Rudy, B., Chow, A., Lau, D., Amarillo, Y., Ozaita, A., Saganich, M., Moreno, H., Nadal, M.S., Hernandez-Pineda, R., Hernandez-Cruz, A., et al. (1999). Contributions of Kv3 channels to neuronal excitability. *Ann. N. Y. Acad. Sci.* **868**, 304–343.
- Russell, S.N., Publicover, N.G., Hart, P.J., Carl, A., Hume, J.R., Sanders, K.M., and Horowitz, B. (1994). Block by 4-aminopyridine of a Kv1.2 delayed rectifier K⁺ current expressed in Xenopus oocytes. *J. Physiol.* **481 (Pt 3)**, 571–584.
- Sankaranarayanan, K., Varshney, A., and Mathew, M.K. (2005). N type rapid inactivation in human Kv1.4 channels: functional role of a putative C-terminal helix. *Mol. Membr. Biol.* **22**, 389–400.
- Schönherr, R., Gessner, G., Löber, K., and Heinemann, S.H. (2002). Functional distinction of human EAG1 and EAG2 potassium channels. *FEBS Lett.* **514**, 204–208.
- Schrøder, R.L., Jespersen, T., Christoffersen, P., Strøbaek, D., Jensen, B.S., and Olesen, S.P. (2001). KCNQ4 channel activation by BMS-204352 and retigabine. *Neuropharmacology* **40**, 888–898.

- Smith, J.S., Iannotti, C.A., Dargis, P., Christian, E.P., and Aiyar, J. (2001). Differential expression of kcnq2 splice variants: implications to m current function during neuronal development. *J. Neurosci. Off. J. Soc. Neurosci.* **21**, 1096–1103.
- Sprung, L.K., Stewig, N.J., and O'Grady, S.M. (1996). Effects of charybdotoxin on K⁺ channel (KV1.2) deactivation and inactivation kinetics. *Eur. J. Pharmacol.* **314**, 357–364.
- Stühmer, W., Ruppertsberg, J.P., Schröter, K.H., Sakmann, B., Stocker, M., Giese, K.P., Perschke, A., Baumann, A., and Pongs, O. (1989). Molecular basis of functional diversity of voltage-gated potassium channels in mammalian brain. *EMBO J.* **8**, 3235–3244.
- Sung, M.J., Hahn, S.J., and Choi, B.H. (2009). Effect of psoralen on the cloned Kv3.1 currents. *Arch. Pharm. Res.* **32**, 407–412.
- Syrbe, S., Hedrich, U.B.S., Riesch, E., Djémié, T., Müller, S., Møller, R.S., Maher, B., Hernandez-Hernandez, L., Synofzik, M., Caglayan, H.S., et al. (2015). De novo loss- or gain-of-function mutations in KCNA2 cause epileptic encephalopathy. *Nat. Genet.* **47**, 393–399.
- Tian, S., Liu, W., Wu, Y., Rafi, H., Segal, A.S., and Desir, G.V. (2002). Regulation of the voltage-gated K⁺ channel KCNA10 by KCNA4B, a novel beta-subunit. *Am. J. Physiol. Renal Physiol.* **283**, F142–149.
- Tinel, N., Diochot, S., Borsotto, M., Lazdunski, M., and Barhanin, J. (2000). KCNE2 confers background current characteristics to the cardiac KCNQ1 potassium channel. *EMBO J.* **19**, 6326–6330.
- Tipparaju, S.M., Li, X.-P., Kilfoil, P.J., Xue, B., Uversky, V.N., Bhatnagar, A., and Barski, O.A. (2012). Interactions between the C-terminus of Kv1.5 and Kvβ regulate pyridine nucleotide-dependent changes in channel gating. *Pflugers Arch.* **463**, 799–818.
- Trudeau, M.C., Titus, S.A., Branchaw, J.L., Ganetzky, B., and Robertson, G.A. (1999). Functional analysis of a mouse brain Elk-type K⁺ channel. *J. Neurosci. Off. J. Soc. Neurosci.* **19**, 2906–2918.
- Unsold, B., Kerst, G., Brousos, H., Hübner, M., Schreiber, R., Nitschke, R., Greger, R., and Bleich, M. (2000). KCNE1 reverses the response of the human K⁺ channel KCNQ1 to cytosolic pH changes and alters its pharmacology and sensitivity to temperature. *Pflugers Arch.* **441**, 368–378.
- Wang, H.S., Pan, Z., Shi, W., Brown, B.S., Wymore, R.S., Cohen, I.S., Dixon, J.E., and McKinnon, D. (1998). KCNQ2 and KCNQ3 potassium channel subunits: molecular correlates of the M-channel. *Science* **282**, 1890–1893.
- Wang, S., Ding, W.-G., Bai, J.-Y., Toyoda, F., Wei, M.-J., and Matsuura, H. (2016). Regulation of human cardiac Kv1.5 channels by extracellular acidification. *Pflugers Arch.* **468**, 1885–1894.
- Weiser, M., Vega-Saenz de Miera, E., Kentros, C., Moreno, H., Franzen, L., Hillman, D., Baker, H., and Rudy, B. (1994). Differential expression of Shaw-related K⁺ channels in the rat central nervous system. *J. Neurosci. Off. J. Soc. Neurosci.* **14**, 949–972.
- Wettwer, E., and Terlau, H. (2014). Pharmacology of voltage-gated potassium channel Kv1.5--impact on cardiac excitability. *Curr. Opin. Pharmacol.* **15**, 115–121.
- Wible, B., Murawsky, M.K., Crumb, W.J., and Rampe, D. (1997). Stable expression and characterization of the human brain potassium channel Kv2.1: blockade by antipsychotic agents. *Brain Res.* **761**, 42–50.
- Wible, B.A., Yang, Q., Kuryshev, Y.A., Accili, E.A., and Brown, A.M. (1998). Cloning and expression of a novel K⁺ channel regulatory protein, KChAP. *J. Biol. Chem.* **273**, 11745–11751.

- Xie, C., Su, H., Guo, T., Yan, Y., Peng, X., Cao, R., Wang, Y., Chen, P., Wang, X., and Liang, S. (2014). Synaptotagmin I delays the fast inactivation of Kv1.4 channel through interaction with its N-terminus. *Mol. Brain* 7, 4.
- Xie, G., Harrison, J., Clapcote, S.J., Huang, Y., Zhang, J.-Y., Wang, L.-Y., and Roder, J.C. (2010). A new Kv1.2 channelopathy underlying cerebellar ataxia. *J. Biol. Chem.* 285, 32160–32173.
- Yan, L., Liu, X., Liu, W.-X., Tan, X.-Q., Xiong, F., Gu, N., Hao, W., Gao, X., and Cao, J.-M. (2015). Fe2O3 nanoparticles suppress Kv1.3 channels via affecting the redox activity of Kv β 2 subunit in Jurkat T cells. *Nanotechnology* 26, 505103.
- Yang, F., and Zheng, J. (2014). High temperature sensitivity is intrinsic to voltage-gated potassium channels. *ELife* 3.
- Yang, T., Snyders, D.J., and Roden, D.M. (1997). Inhibition of cardiac potassium currents by the vesnarinone analog OPC-18790: comparison with quinidine and dofetilide. *J. Pharmacol. Exp. Ther.* 280, 1170–1175.
- Yang, Y., Xia, M., Jin, Q., Bendahhou, S., Shi, J., Chen, Y., Liang, B., Lin, J., Liu, Y., Liu, B., et al. (2004). Identification of a KCNE2 gain-of-function mutation in patients with familial atrial fibrillation. *Am. J. Hum. Genet.* 75, 899–905.
- Yang, Y., Vasylyev, D.V., Dib-Hajj, F., Veeramah, K.R., Hammer, M.F., Dib-Hajj, S.D., and Waxman, S.G. (2013). Multistate structural modeling and voltage-clamp analysis of epilepsy/autism mutation Kv10.2-R327H demonstrate the role of this residue in stabilizing the channel closed state. *J. Neurosci. Off. J. Soc. Neurosci.* 33, 16586–16593.
- Yeung, S.Y.M., Thompson, D., Wang, Z., Fedida, D., and Robertson, B. (2005). Modulation of Kv3 subfamily potassium currents by the sea anemone toxin BDS: significance for CNS and biophysical studies. *J. Neurosci. Off. J. Soc. Neurosci.* 25, 8735–8745.
- Yunoki, T., Takimoto, K., Kita, K., Funahashi, Y., Takahashi, R., Matsuyoshi, H., Naito, S., and Yoshimura, N. (2014). Differential contribution of Kv4-containing channels to A-type, voltage-gated potassium currents in somatic and visceral dorsal root ganglion neurons. *J. Neurophysiol.* 112, 2492–2504.
- Zaika, O., Hernandez, C.C., Bal, M., Tolstykh, G.P., and Shapiro, M.S. (2008). Determinants within the turret and pore-loop domains of KCNQ3 K $^{+}$ channels governing functional activity. *Biophys. J.* 95, 5121–5137.
- Zhang, X., Bursulaya, B., Lee, C.C., Chen, B., Pivaroff, K., and Jegla, T. (2009). Divalent cations slow activation of EAG family K $^{+}$ channels through direct binding to S4. *Biophys. J.* 97, 110–120.
- Zhang, Y., Zhang, X.-F., Fleming, M.R., Amiri, A., El-Hassar, L., Surguchev, A.A., Hyland, C., Jenkins, D.P., Desai, R., Brown, M.R., et al. (2016). Kv3.3 Channels Bind Hax-1 and Arp2/3 to Assemble a Stable Local Actin Network that Regulates Channel Gating. *Cell* 165, 434–448.
- Zhong, X.Z., Abd-Elrahman, K.S., Liao, C.-H., El-Yazbi, A.F., Walsh, E.J., Walsh, M.P., and Cole, W.C. (2010). Stromatoxin-sensitive, heteromultimeric Kv2.1/Kv9.3 channels contribute to myogenic control of cerebral arterial diameter. *J. Physiol.* 588, 4519–4537.
- Zhou, W., Qian, Y., Kunjilwar, K., Pfaffinger, P.J., and Choe, S. (2004). Structural insights into the functional interaction of KChIP1 with Shal-type K(+) channels. *Neuron* 41, 573–586.
- Zhu, S., Peigneur, S., Gao, B., Luo, L., Jin, D., Zhao, Y., and Tytgat, J. (2011). Molecular diversity and functional evolution of scorpion potassium channel toxins. *Mol. Cell. Proteomics MCP* 10, M110.002832.

Zou, A., Lin, Z., Humble, M., Creech, C.D., Wagoner, P.K., Kraft, D., Jegla, T.J., and Wickenden, A.D. (2003). Distribution and functional properties of human KCNH8 (Elk1) potassium channels. *Am. J. Physiol. Cell Physiol.* **285**, C1356-1366.