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CONTACT INFORMATION

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PROFESSIONAL STATEMENT

Human epilepsy arising from variants in genes for voltage-gated sodium and potassium channels is the major focus of our lab. We discovered that KCNQ potassium channels were concentrated in the axon, and identified the first molecular mechanisms for KCNQ channel targeting at the axonal initial segment and nodes of Ranvier. We have contributed to the first physiological demonstrations of KCNQ2 channel function in identified PNS and CNS axons. We have used molecular phylogenetics and comparative anatomy to identify previously unknown functional domains of KCNQ channels and interacting proteins. Using such approaches, we showed that the targeting of certain KCNQ channels to axons is an outcome of convergent evolution in early vertebrates. In an unusual example of molecular convergence, a newly duplicated KCNQ gene acquired the property of selective axonal targeting by mimicking a motif present on the sodium channel. This sharing establishes an essential balance conserved in all vertebrates between restraint (mediated by KCNQ channels) and excitability (mediated by sodium channels). This balance ensures proper action potential initiation and saltatory conduction. Loss of this balance leads to epilepsy.

KCNQ2 encephalopathy: mechanisms, models, and new therapies

Human KCNQ2 mutations cause a spectrum of disease phenotypes, including a transient form of neonatal epilepsy, peripheral nerve hyperexcitability manifesting as muscle twitches, and severe epilepsy with profound intellectual disability (epileptic encephalopathy). We are analyzing the basis for KCNQ2 genotype-phenotype relationships through multidisciplinary studies combining protein immunolocalization, electrophysiology in cultured cells, neurons and in vivo, and computational modeling. We are testing strategies for reversing the effects of KCNQ2 mutations as treatments in cases with persistent epilepsy and developmental delay.

ANK3-related mechanisms in epilepsy and psychiatric disease

We first showed that KCNQ channel clustering on axons depends on interaction with ankyrin-G, a large adaptor protein, encoded by a very complex gene, ANK3. Recently, variation at ANK3 has been implicated in epilepsy, autism, schizophrenia, and bipolar disorder. We have begun new work analyzing the molecular diversity of ankyrin-G isoforms, based on the hypothesis that altered expression of particular isoforms contributes to ANK3's association with disease.

CERTIFICATION

- American Board of Psychiatry and Neurology, Neurology

EDUCATION

- M.D./Ph.D., Yale University, Conn.
- Internship, Internal Medicine, University of California, San Francisco, School of Medicine, San Francisco, Calif.
- Residency, Neurology, University of California, San Francisco, School of Medicine, San Francisco, Calif.
- Fellowship, Epilepsy Research, University of California, San Francisco, School of Medicine, San Francisco, Calif.
- Fellowship, Molecular Neurobiology, Howard Hughes Medical Institute, University of California, San Francisco, School of Medicine, San Francisco, Calif.

WEBSITE

- Molecular Neuropharmacology Lab (https://www.bcm.edu/research/labs/edward-cooper)

CONSULT

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CLINICAL INTERESTS

Epilepsy and seizures; family history of epilepsy

RESEARCH INTERESTS

- Voltage-gated sodium and potassium channel diseases of the axon: evolution, mechanisms, animal models, and therapeutics
- Epilepsy and seizures
- Family history of epilepsy

GRANTS (NIH FUNDING)

- Cooper EC (Principal Investigator). KCNQ2/3 channels in neonatal-onset epilepsy and encephalopathy. National Institute of Neurological Disorders and Stroke R01 grant (PA-11-260); $1,002,516 awarded in direct costs, April 1, 2005–Jan. 31, 2018.
- Cooper EC (Principal Investigator). KCNQ2 Epileptic Encephalopathy: Overcoming Hurdles to Effective Disease-Modifying Therapy. National Institute of Neurological Disorders and Stroke R13 grant (PA-13-347); $15,000 awarded in direct costs, Sept. 21, 2015–Sept. 20, 2016.

JOURNAL PUBLICATIONS

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2. Bayat A, Iavarone S, Miceli F, Jakobsen AV, Johannesen KM, Nikanorova M, et al. Phenotypic and functional assessment of two novel KCNQ2 gain-of-function variants Y141N and G239S and effects of amitriptyline treatment. Neurotherapeutics. 2024;21(1):e00296. PMID: 38241158.
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4. Boets S, Johannesen KM, Destree A, Manti F, Ramantani G, Lesca G, et al. Adult phenotype of KCNQ2 encephalopathy. J Med Genet. 2022;59(6):528-35. PMID: 33811133.
5. Cooper EC, Abreo T, Tran B. KCNQ channel PIP2 modulation: Two loose links, three rings, and a twist. Neuron. 2022;110(2):178-80. PMID: 35051360.
6. Jing J, Dunbar C, Sonesra A, Chavez A, Park S, Yang R, et al. Removal of KCNQ2 from parvalbumin-expressing interneurons improves anti-seizure efficacy of retigabine. Exp Neurol. 2022;355:114141. PMID: 35691372.
7. Mazzaferro S, Msekela DJ, Cooper EC, Maheshwari A, Sine SM. Genetic variant in nicotinic receptor alpha4-subunit causes sleep-related hyperkinetic epilepsy via increased channel opening. Int J Mol Sci. 2022;23(20):. PMID: 36292983.
8. Miceli F, Millevert C, Soldovieri MV, Mosca I, Ambrosino P, Carotenuto L, et al. KCNQ2 R144 variants cause neurodevelopmental disability with language impairment and autistic features without neonatal seizures through a gain-of-function mechanism. EBioMedicine. 2022;81:104130. PMID: 35780567.
9. Vanoye CG, Desai RR, Ji Z, Adusumilli S, Jairam N, Ghabra N, et al. High-throughput evaluation of epilepsy-associated KCNQ2 variants reveals functional and pharmacological heterogeneity. JCI Insight. 2022;7(5):. PMID: 35104249.
10. Miceli F, Carotenuto L, Barrese V, Soldovieri MV, Heinzen EL, Mandel AM, et al. A Novel Kv7.3 Variant in the Voltage-Sensing S4 Segment in a Family With Benign Neonatal Epilepsy: Functional Characterization and in vitro Rescue by beta-Hydroxybutyrate. Front Physiol. 2020;11:1040. PMID: 33013448.

11. Tran B, Ji ZG, Xu M, Tsuchida TN, Cooper EC. Two KCNQ2 encephalopathy variants in the calmodulin-binding helix A exhibit dominant-negative effects and altered PIP2 interaction. Front Physiol. 2020;11:1144. PMID: 33041849.
12. Goto A, Ishii A, Shibata M, Ihara Y, Cooper EC, Hirose S. Characteristics of KCNQ2 variants causing either benign neonatal epilepsy or developmental and epileptic encephalopathy. Epilepsia. 2019;60(9):1870-80. PMID: 31418850.
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14. Soldovieri MV, Ambrosino P, Mosca I, Miceli F, Franco C, Canzoniero LMT, et al. Epileptic encephalopathy in a patient with a novel variant in the Kv7.2 S2 transmembrane segment: Clinical, genetic, and functional features. Int J Mol Sci. 2019;20(14):. PMID: 31295832.
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17. Mulkey SB, Ben-Zeev B, Nicolai J, Carroll JL, Gronborg S, Jiang YH, et al. Neonatal nonepileptic myoclonus is a prominent clinical feature of KCNQ2 gain-of-function variants R201C and R201H. Epilepsia. 2017;58(3):436-45. PMID: 28139826.
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24. Ho TS, Zollinger DR, Chang KJ, Xu M, Cooper EC, Stankewich MC, et al. A hierarchy of ankyrin-spectrin complexes clusters sodium channels at nodes of Ranvier. Nat Neurosci. 2014;17(12):1664-72. PMID: 25362473.
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27. Millichap JJ, Cooper EC. KCNQ2 potassium channel epileptic encephalopathy syndrome: Divorce of an electro-mechanical couple? Epilepsy Curr. 2012;12(4):150-2. PMID: 22936888.
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Neurology. 2007;69(13):1310-1. PMID: 17893291.

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43. Devaux JJ, Kleopa KA, Cooper EC, Scherer SS. KCNQ2 is a nodal K+ channel. J Neurosci. 2004;24(5):1236-44. PMID: 14762142.
44. Cooper EC, Jan LY. M-channels: Neurological diseases, neuromodulation, and drug development. Arch Neurol. 2003;60(4):496-500. PMID: 12707061.
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BOOK CHAPTERS and OTHER PUBLICATIONS

1. Miceli F, Soldovieri MV, Weckhuysen S, Cooper E, Taglialatela M. KCNQ2-related disorders. In: Adam MP, Mirzaa GM, Pagon RA, Wallace SE, Bean LJH, Gripp KW, Amemiya A, editors. GeneReviews((R)). Seattle (WA): 2022. Available from: http://www.ncbi.nlm.nih.gov/pubmed/20437616.
2. Miceli F, Soldovieri MV, Joshi N, Weckhuysen S, Cooper E, Taglialatela M. KCNQ2-related disorders. In: Pagon RA, Adam MP, Ardinger HH, Wallace SE, Amemiya A, Bean LJH, Bird TD, Fong CT, Mefford HC, Smith RJH, Stephens K, editors. GeneReviews(R). Seattle (WA): 2016. Available from: http://www.ncbi.nlm.nih.gov/pubmed/20437616.
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POSTER and PLATFORM PRESENTATIONS

1. Cohen S, Smith L, Parthasarathy S, Bonkowski E, Hong W, Wiltrout K, et al. Voltage clamp and genetic variant pathogenicity: Epilepsy sodium channel variant curation expert panel consensus. Abstract No 3.356, 2023, American Epilepsy Society Annual Meeting, www.aesnet.org.
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3. Cooper E, Joshi N, Millichap J, Tsuchida T, Nesbitt G, Taglialatela M, et al. The RIKEE database: Genotype-phenotype patterns in KCNQ channel related disease, based on 784 individuals and pedigrees. Neurology. 2021;96(15 Suppl):2427.
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5. Park S, Dunbar C, Tarkunde Y, Akbar A, Marks R, Lee M, et al. Pharmacodynamic dependence of retigabine on Kcnq2 expression in PV+ interneurons. Abstract No 3.053, 2018, American Epilepsy Society Annual Meeting, www.aesnet.org.
6. Zhang J, Chen C, Kim E, Procko E, Patel J, Choi R, et al. Missense epilepsy mutations in neuronal KCNQ/Kv7 channels occur at hotspots within highly conserved functional domains of Kv7.2 and Kv7.3. Program No. 558.12. 2018 Neuroscience Meeting Planner. Washington, DC: Society for Neuroscience, 2018. Online.
7. Sands TT, Miceli F, Lesca G, Beck A, Cimino M, Strong N, et al. Autism with benzodiazepine-responsive electrical status epilepticus in sleep (ESES) caused by KCNQ3 gain-of-function variants. Abstract No 2.380, 2017, American Epilepsy Society Annual Meeting, www.aesnet.org.
8. Ji Z, Tran B, Li L, Xu M, Cooper E. Functional characterization of three de novo KCNQ2 encephalopathy variants in the pore helix and responses to SF0034 and ICA-069673. Abstract No 3.256, 2016, American Epilepsy Society Annual Meeting, www.aesnet.org.
9. Joshi N, Taglialatela M, Weckhuysen S, Nesbitt G, Cooper E. An informatics infrastructure for KCNQ2 encephalopathy research including a patient registry, database, curation platform, and website. Abstract No 3.329, 2016, American Epilepsy Society Annual Meeting, www.aesnet.org.

10. Taglialatela M, Millichap J, Miceli F, De Maria M, Keator C, Joshi N, et al. Infantile spasms and encephalopathy without preceding neonatal seizures caused by KCNQ2 R198Q, a gain-of-function variant. Abstract No 2.366, 2016, American Epilepsy Society Annual Meeting, www.aesnet.org.
11. Wang X, Li L, Xu M, Mays J, Joshi N, Cooper E. Genotype-phenotype relationships In KCNQ2 related epilepsy and encephalopathy caused by variants in the pore region. Abstract No 3.134, 2016, American Epilepsy Society Annual Meeting, www.aesnet.org.
12. Joshi N, Cooper EC, Taglialatela M, Weckhuysen S. An international, curated KCNQ2 registry, database and website. Abstract No 1.308, 2015, American Epilepsy Society Annual Meeting, www.aesnet.org.
13. Li L, Cooper EC. KCNQ2 encephalopathy: Novel single amino acid deletion variants strongly suppress currents and are responsive to SF0034. Abstract No 1.346, 2015, American Epilepsy Society Annual Meeting, www.aesnet.org.
14. Millichap J, Miceli F, Tran B, Keator C, Joshi N, Virginia Soldovieri M, et al. KCNQ2 p.Arg198Gln, a gain-of-function variant presenting recurrently as West syndrome without preceding neonatal seizures. Abstract No 3.337, 2015, American Epilepsy Society Annual Meeting, www.aesnet.org.
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18. Park K, Millichap J, Cooper E. KCNQ2-deficiency: Clinical spectrum of epilepsy, encephalopathy, and response to ezogabine. Abstract No 3.210, 2014, American Epilepsy Society Annual Meeting, www.aesnet.org.
19. Tran B, Xu M, Cooper E. A KCNQ2/3 mutation causing severe epilepsy disrupts channel targeting to the axon initial segment. Abstract No 3.017, 2014, American Epilepsy Society Annual Meeting, www.aesnet.org.
20. Li L, Tran B, Xu M, Millichap J, Porter B, Cooper E. Pore loop KCNQ2 mutations causing epileptic encephalopathy strongly suppress wild type KCNQ2 currents when co-expressed in mammalian cells. Abstract No 3.022, 2013, American Epilepsy Society Annual Meeting, www.aesnet.org.
21. Tran B, Xu M, Cooper E. KCNQ2 targeting to the axon initial segment is disrupted in a mutant form causing severe epilepsy. Abstract No 1.022, 2013, American Epilepsy Society Annual Meeting, www.aesnet.org.
22. Xu M, Cooper E. Ankyrin-G as a potential molecular mechanism linking epilepsy and comorbid mood disorder. Abstract No 1.029, 2013, American Epilepsy Society Annual Meeting, www.aesnet.org.
23. Chang K-J, Zollinger DR, Susuki K, Ho TS, Cooper EC, Bennett V, et al. Paranodal ankyrins: Enigmatic glial anchors. Program No. 699.05. 2013 Neuroscience Meeting Planner. San Diego, CA: Society for Neuroscience, 2013. Online.
24. Ho T, Zollinger DR, Xu M, Cooper EC, Stankewich MC, Bennett V, et al. The roles of ankyrin-G in node of Ranvier formation in vivo. Program No. 699.17. 2013 Neuroscience Meeting Planner. San Diego, CA: Society for Neuroscience, 2013. Online.
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26. Millichap JJ, Levisohn PM, Tsuchida TN, Cooper EC. Treatment of KCNQ2 encephalopathy with ezogabine. Abstract No 1.355, 2012, American Epilepsy Society Annual Meeting, www.aesnet.org.
27. Cooper E. Ion channel modifications in epilepsy. Presented at the American Academy of Neurology (AAN), 64th Annual Meeting in New Orleans, La. (April 21-28, 2012).