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Kevin.ess@vumc.org

Personal:

Kevin Christopher Ess
Birthday: January 9
Birthplace: Buffalo, NY
Citizenship: U.S.A.
Married: Dr. Jennifer Ess; 2 children, Grace and Gabriella

Education:

1989 Bachelor of Music, College-Conservatory of Music, University of Cincinnati, OH
1996 Ph.D., Department of Developmental Biology, University of Cincinnati, OH.
Dissertation “c-Myb: Organizer of Gene Expression in Thymocytes”.
Advisors: John Hutton, M.D. and Bruce Aronow, Ph.D.
1998 M.D., College of Medicine, University of Cincinnati

Postgraduate Training:

June 1998 to June 1999 Pediatric Internship, Denver Children’s Hospital, Denver, CO
June 1999 to June 2002 Pediatric Neurology Residency, St. Louis Children’s Hospital, Barnes-Jewish Hospital and Washington University School of Medicine, St. Louis, Missouri
July 2002 to June 2004 Clinical Neurophysiology Fellowship, St. Louis Children’s Hospital and Washington University
July 2003 to June 2005 Postdoctoral Fellow, Laboratory of David H. Gutmann, MD, PhD

Medical Licensure and Certification:

1999 – 2006 State of Missouri
2006 to present State of Tennessee
June 2004 to 12/31/2014 Neurology with Special Certification in Child Neurology, American Board of Psychiatry and Neurology
Recertified 2015-2025 Neurology with Special Certification in Child Neurology, American Board of Psychiatry and Neurology
May 2007 to 12/31/2017 Clinical Neurophysiology, American Board of Psychiatry and Neurology

Academic Appointments:

July 2004 to March 2006 Instructor, Depts. of Neurology and Pediatrics, Washington University
April 2006 to 2013 Assistant Professor, Department of Pediatrics, Vanderbilt University
April 2006 to 2013 Assistant Professor, Department of Neurology, Vanderbilt University, Nashville, TN
April 2006 to 2013 Investigator, Kennedy Center for Research on Human Development, Vanderbilt University
April 2009 to 2013 Assistant Professor, Department of Cell and Developmental Biology, Vanderbilt University

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| July 2009 to present | Member, Vanderbilt Center for Stem Cell Biology |
| August 2011 to 2013 2013 to present | Assistant Professor, Department of Biological Sciences, Vanderbilt University Director, Division of Pediatric Neurology Neurologist-in-Chief, Vanderbilt Children's Hospital |
| April 2014 to present 2015-present | Associate Professor, Departments of Pediatrics, Neurology, Cell & Developmental Biology, Vanderbilt University Gerald M. Fenichel Chair in Neurology |

Hospital Appointments:

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| July 2004 to March 2006 | St. Louis Children's Hospital, and Barnes-Jewish Hospital, St. Louis, MO |
| April 2006 to present | Monroe Carell Jr. Children's Hospital at Vanderbilt, Vanderbilt University Hospital, Vanderbilt University Medical Center, Nashville, TN |
| 2019 to present | St. Thomas Midtown Hospital, Nashville TN |

Other Employment:

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| Sept. 1988 to August 1991 | West Virginia Symphony Orchestra |
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Professional Organizations:

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| 2001 to present | American Epilepsy Society |
| 2003 to present | Tuberous Sclerosis Alliance Professional Advisory Board |
| 2006 to present | Society for Neuroscience |
| 2009 to present | Child Neurology Society |
| 2011 to present | Society for Developmental Biology |
| 2011 to present | International Society for Stem Cell Research |
| 2012 to present | Elected member and Fellow, American Neurological Association |
| 2013 to present | Fellow, American Academy of Pediatrics |
| 2014 to present | Member, Professors of Child Neurology |
| 2015 to present | Elected member, Society for Pediatric Research |

Professional Service

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| 2011 to 2013 | American Epilepsy Society Scientific Program Committee |
| 2010 to 2015 | Child Neurology Society, Awards Committee |
| 2011 to 2015 | Child Neurology Society, Research Committee |
| 2012 to 2015 | Association of University Professors of Neurology |
| 2013 to present | Member, Clinical Immunization Safety Assessment Network at Vanderbilt |
| 2014 to 2016 | Councillor, Elected Officer of the Executive Board, Child Neurology Society |
| 2015 to present | Tuberous Sclerosis Complex Preclinical Consortium |
| 2016 to present | Board of Directors, Thisbe and Noah Scott Foundation for Pediatric Neuromuscular Diseases |
| 2017 to present | Director, Vanderbilt Leukodystrophy Care Network, member Neurology Clinical Practice Guidelines committee |
| 2018 to present | Advisory Board Member, Developmental Synaptopathies Consortium |
| 2020 to present | Co-Director, Vanderbilt Center for Pediatric Rare Diseases |

Professional Activities:

Attending Physician

Inpatient Neurology (2-4 weeks per year) and Pediatric Epilepsy Monitoring Unit (2-4 weeks per year)

Pediatric EEG interpretation

Founder and Director: Vanderbilt Tuberous Sclerosis Clinic, Vanderbilt Alternating Hemiplegia of Childhood Clinic, Vanderbilt Leukodystrophy Care Network, Fabry Clinic, Adrenoleukodystrophy Clinic

Journal Review

Editorial Board:

Journal of Child Neurology
Neurobiology of Disease

Ad hoc reviewer for:

Neurology India
Pediatric Drugs
Journal of Child Neurology
Epilepsia
Acta Neurologica Scandinavica
Neurology
Annals of Human Genetics
Clinical and Experimental Dermatology
EMBO Molecular Medicine
Developmental Dynamics
Journal of Neuroscience
Kidney International
Archives of Neurology
Experimental Neurology
PLoS ONE
Neural Plasticity
Kidney International
New England Journal of Medicine
Behavior Genetics
Cellular and Molecular Life Sciences
Disease Models Mechanisms
AJP: Renal Physiology
Cell Reports

Grant Review

Ad hoc reviewer for:

Citizens United for Research in Epilepsy (CURE)
Autism Speaks
Tuberous Sclerosis Alliance (2006-present)
NICHD, NIH, Autism Centers of Excellence Study Section 2012
NINDS, NIH, NRSA/T32 Training Grants Study Section Fall 2012
Telethon (Italy) 2013
E-Rare, Bonn, Germany 2013
NIH August 2014, ZRG1 Study Section
Dept. of Defense Tuberous Sclerosis Complex Grant Programmatic Review, 2013-2015
2017 *Ad hoc* reviewer, Tuberous Sclerosis Complex Research Program (TSCR), Department of Defense Congressionally Directed Medical Research Programs (CDMRP)
NIH Special Emphasis Panel 2018/05 ZRG1 OBT-H (02)

NIH Study Section, NST-1, NINDS, (2018-present)

Clinical Studies

2010-2012 Data Monitoring Committee and Chair for Exist-1 and Exist-2, Novartis Pharmaceuticals
2013-2015 Data Monitoring Committee for Exist-3, Novartis Pharmaceuticals

- 2015-2017 Site P.I., SP848, "Open Label Study to Determine Safety, Tolerability, and Efficacy of Long-Term Oral Lacosamide as Adjunctive Therapy in Children with Epilepsy".
- 2019-2020 Buccal Diazepam for Epilepsy

Select Invited Lectures

- Tuberous Sclerosis Alliance National Meeting 2002, Washington, DC.
- Tuberous Sclerosis Alliance-LAM National Meeting 2005, Cincinnati, OH.
- Tuberous Sclerosis Alliance National Conference 2006, Chicago, IL.
- Tuberous Sclerosis Alliance National Meeting 2007, Annapolis, MD.
- "Neurobiology of Tuberous Sclerosis Complex" Keystone Symposia "Towards Identifying the Pathophysiology of Autistic Syndromes" 2008, Santa Fe, NM.
- "Advances in Tuberous Sclerosis Complex: Animal Models and New Therapies". 2008, University of Birmingham, AL.
- "Lessons from Tuberous Sclerosis: Seizures, Autism, and Easter Island". Adelyn Stroup Visiting Lectureship in Pediatric Epilepsy, May 2008, Johns Hopkins School of Medicine, Baltimore, MD.
- "IDDRC Seminar". Children's Research Institute at the Children's National Medical Center, October 2008, Washington DC.
- "Tuberous Sclerosis, Comorbidity and Connectivity". American Epilepsy Society Workshop "The comorbidities of epilepsy: The unification of multiple disease states through basic mechanisms." December 2008, Seattle, WA.
- "Epilepsy and Stigma in Africa", April 2009, Lusaka, Zambia
- "Zebrafish Model of TSC", Tuberous Sclerosis Alliance Conference 2009, Bloomington, IL.
- "Tuberous Sclerosis Complex: Astrocytomas, mTOR and Easter Island" AAN 2010 Meeting, Toronto, Canada.
- "Human Stem Cells to Study Neuronal Development and the Role of mTOR Signaling", 3rd Southwest Stem Cell Consortium Workshop 2010, Nashville, TN.
- "Combining Clinical and Research Careers: How I Am Doing It", NINDS/AUPN/ANA Sponsored Combining Clinical and Research Careers in Neuroscience Symposium 2011, Wash. DC.
- "Tumorigenesis in Zebrafish: TOR is Not Enough?", 7th European Zebrafish Meeting 2011, Edinburgh, Scotland.
- "Induced Pluripotent Stem Cells in Child Neurology: New Tools, New Hope?". Child Neurology Society 2011 Meeting Savannah, GA (Speaker and Chair of Symposium).
- "Sometimes it Takes a Zoo, My Journey from St. Louis to Easter Island and Back", Washington Univ. in St. Louis, Dept. of Developmental Biology, October 2012, St. Louis, MO.
- "Neurocutaneous Disorders" AAN March 2013 Meeting, San Diego, CA.
- "MADD about TSC" Seattle Children's Research Institute, Seattle WA, March 2013
- "Tuberous Sclerosis Complex, Sometimes it Takes a Zoo" San Raffaele Scientific Institute, July 2013, Milan, Italy
- "MADD about TSC: A Tale of Two Disorders" Rutgers Medical Center, New Jersey, November 2013
- "Lessons from a Menagerie: New Insights for Tuberous Sclerosis Complex", Neurology Grand Rounds, Montefiore Medical Center, January 2015
- "The Menagerie of Tuberous Sclerosis Complex", University of Virginia Stem Cell and Regenerative Biology Seminar September 2015, Charlottesville, VA.

“Epilepsy and Mimickers of Epilepsy”, 12th Annual Pediatric Review Conference, Al Khalidi Medical Center, March 2017, Amman, Jordan.

“Control of Myelination by mTOR Kinase Signaling in Tuberous Sclerosis Complex “. XIII European Meeting on Glial Cells in Health and Disease July 2017, Edinburgh, Scotland.

"Stem Cell Derived Human Neurons to Study *ATP1A3* Mutations in Alternating Hemiplegia of Childhood", 6th Symposium on *ATP1A3* in Disease, September 2017, Tachikawa, Tokyo, Japan.

Various lectures, 13th Annual Pediatric Review Conference, Al Khalidi Medical Center, March 2018, Amman, Jordan.

Various lectures, 14th Annual Pediatric Review Conference, Al Khalidi Medical Center, March 2019, Amman, Jordan.

“mTOR Pathway- a New Window for Precision Therapy” Pediatric Neurology and Developmental Medicine Update Course, Duke University Medical Center, March 2019.

Various lectures, 15th Annual Pediatric Review Conference, Al Khalidi Medical Center, February 2020, Amman, Jordan.

Conferences/Meetings/Symposium as Organizer:

Tuberous Sclerosis Alliance and LAM Regional Meeting, 2016, Nashville TN
7th Symposium on *ATP1A3* in Disease, October 2018, Chicago, IL

Honors:

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| 1996 | Graduate Student Research Award, College of Medicine, Univ. of Cincinnati |
| 1998 | Alpha Omega Alpha |
| 2003 | Epifellows Award |
| 2006 | Vanderbilt Physician Scientist Development Award |
| 2007 | Milken Family Career Development Award |
| 2008 | Adelyn Stroup Visiting Lectureship in Pediatric Epilepsy, Johns Hopkins U. |
| 2008 | Hazinski-Turner Award, Vanderbilt Dept. of Pediatrics |
| 2009 | Doris Duke Clinical Scientist Development Award |
| 2012 | Elected Member and Fellow, American Neurological Association |
| 2013, 14 | Five Star Award for Excellence in Healthcare, sponsored by Professional Research Consultants |
| 2014-22 | “Nashville Top Doctor” <i>Nashville Lifestyles Magazine</i> |

Teaching Activities:

Medical School: Brain and Behavior “Molecular Development of the CNS” Lecture, Vanderbilt University Medical Students Oct. 2013
Third year Neurology clerkship, Child Neurology lecture (wrote lecture)
Third year Neurology clerkship LP training session
Third year Pediatrics: Rotation, Meharry Medical College (wrote lecture)
Developed and Course Director for Advanced Clinical Electives in Pediatric Epilepsy (PEACE) and Pediatric Neurology (2015-present)
BMM 2015, “Pathophysiology of epilepsy and “Approach to Epilepsy”
First year students, Foundations of Medicine, (2015-present)

Graduate School: NURO345: Forebrain Development Lectures and journal club (2007-2014)

Cell Biology 310 “Stem Cells” and journal club (2009-present)
NURO310: “Neurobiology of Disease” (2008-present)
CellBio: “Primary Cilia in Development and Disease” (2010)
NURO366 “Epilepsy and Brain Development” (2010, 2012, 2013, 2015, 2017)
“Epilepsy”, HHMI sponsored Certificate Program in Molecular Medicine (2012)
CBIO320 “mTOR and Development” in Cancer & Embryonic Development class (2013)

Dissertation and qualifying exam committee member for >17 Ph.D. students in Cell & Developmental Biology, Pharmacology, Biological Sciences and Neuroscience.

Postgraduate lectures Neurology residents and Clinical Neurophysiology fellows (2-3 lectures/year)
Child Neurology lecture to 3rd year medical students at Meharry Medical School 1-2 times per year (2008-present)

Continuing Medical Education, Lectures:

“First Time and Febrile Seizures”. Emergency Department Grand Rounds, Williamson Medical Center, May 2009.
“Ketogenic Diet” with Mary Montgomery, RD. East TN Children’s Hospital, April 2012.
General Pediatric Update 2012, Univ. South Carolina CME (4 lectures), Hilton Head, SC.
“Genetically Determined Epilepsies” Vanderbilt Contemporary Clinical Epilepsy Symposium 2016, 2017

Clinical Supervision:

Vanderbilt Child Neurology, Adult Neurology, and Pediatric residents, Neurophysiology fellows, medical students in various outpatient and inpatient settings

Research Supervision:

Robert Carson, MD, PhD. Postdoctoral fellow (2009-2010, 2011-present), now Assistant Professor, K08 mentee 2013-2018

Cary Fu, MD. Postdoctoral fellow (2009--2014), now Assistant Professor

Eric Armour, MD, PhD. MSTP student (2009-2013). Defended Ph.D. dissertation Sept. 6th 2013, now Neurophysiology fellow, Vanderbilt

Seok-Hyung Kim, Ph.D., Research Assistant Professor (2010-2014), now Assistant Professor, Univ. South Carolina

Lynsa Nguyen, MD. Vanderbilt undergraduate (2008-2010), Maternal Fetal Medicine Fellow, Vanderbilt

Adrienne Bruce, MD. Vanderbilt undergraduate (2009-2011), Instructor in Surgery, University of Pennsylvania

Brandon Goodman, MD. Vanderbilt undergraduate (2010-2012), current Psychiatry fellow, UNC Chapel Hill

William Clinkscales, MD. Vanderbilt undergraduate (2011-2013), ENT resident, University TN, Memphis

Inan Olmez, MD. Vanderbilt Child Neurology Resident (2011-2013), now Assistant Professor, Ohio State University

Laura Armstrong, MD, PhD. MSTP student (2012-2017), defended Ph.D. dissertation Aug. 11th 2017, now PGY-3 Child Neurology Resident, Vanderbilt

John P. Snow, MD, PhD. MSTP student (2015-2020), defended Ph.D. dissertation April 27th 2020, now PGY-1 Neurology Resident, Mass. General Hospital

Lindsay Klofas, MD, PhD. MSTP student (co-mentor with R. Carson) (2016-2020) defended Ph.D. dissertation April 21st 2020, now PGY-1 Ophthalmology Resident, Mass. General Hospital

Krystine Sandlin, MD. post-doctoral fellow (2018-2020), now Assistant Professor, University Texas Austin

Daniel Pomerantz, MD. Vanderbilt Medical student, Emphasis (2012-2013, 2014-2015), current resident Genetics Univ. Michigan

Emily Krainer, MD. Vanderbilt Medical student, Emphasis (2015), current resident Child Neurology Univ. Rochester, NY

Mary Chalkley, PhD student (co-mentor with R. Ihrie) (2019-present)

Sonia Joeng, Vanderbilt undergraduate (2017-2020), in medical school

Sofia Hall, Vanderbilt undergraduate (2017-2020), in medical school

Maria Osipovich, Vanderbilt School for Science and Math (2018-2019) High School program, now undergraduate Boston University Honor's College

Kevin Lazenby, Vanderbilt undergraduate (2018-2020), in medical school

Sydney Nelson Vanderbilt undergraduate (2019-2021), in medical school

Samantha Mallahan Vanderbilt undergraduate (2020-present)

Bill Nobis, MD, PhD, Assistant Professor, Dept Neurology, Vanderbilt

Administration/Service:

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| 2006 to present | Recruitment of Neurophysiology Fellows |
| 2006 to present | Recruitment of Child Neurology Residents |
| 2008 to 2011 | Pediatric EMU Quality Committee |
| 2012 to 2016 | Faculty Advisory Committee of the Vanderbilt Medical Scientist Training Program |
| 2012 to 2015 | Program Director, Child Neurology Residency Program |
| 2013 to present | Director, Division of Pediatric Neurology, Neurologist-in-Chief, Vanderbilt Children's Hospital |
| 2015 to 2017 | Member, Pharmacy and Therapeutics Committee |
| 2015 to present | Member, Pediatric Telemedicine Steering Committee |
| 2016 to | Vanderbilt Kennedy Center Director Selection Committee |
| 2016 to | Vanderbilt Neurology Chairperson Selection Committee |
| 2016-present | VARs CDC program, support Drs. Edwards and Creech to review possible vaccine related adverse reactions, review cases with neurological concerns. |
| 2016-present | SDY Sudden Death Young, CDC funded state program to review all deaths under the age of 18 years, classify patients based on histories and autopsy findings. |

Patient Advocacy

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| 2004 to present | Tuberous Sclerosis Alliance, Member Professional Advisory Board |
| 2015 to present | Thisbe and Noah Foundation, Board of Directors |
| 2018 to 2022 | Member, Genetics Advisory Committee, State of TN |

Research Program:

Active Grants (as PI)

- 2012 – 2022 “Molecular Physiology and Pharmacology of *ATP1A3* Mutations in AHC”
Principal Investigator: **Kevin C. Ess**, MD, PhD
Funding agency: Peay and Gipson Foundation
- 2017 – 2022 “Drug development for tuberous sclerosis complex and other pediatric epileptogenic diseases using neurovascular and cardiac microphysiological models”
Principal Investigators: Diana Neely, **Kevin C. Ess**, John Wikswo,
1 UG3 TR002097-01, 4 UH3 TR002097-03, Funding agency: NCATS, NIH
- 2020 – 2025 “Identifying mTOR Dependent Periods During Brain Development”
Principal Investigators: **Kevin C. Ess**, Rebecca Ihrie,
1R01NS118580-01, Funding agency: NINDS, NIH

Other funding

- 04/01/2018 - 03/31/2023 2R01ES010563-17 (Bowman)
NIH/NIEHS
Mechanisms of Manganese Neurotoxicity
Project Role: Co-Investigator
- 09/29/2017 - 09/28/2022 CISA Clinical Vaccine Safety (Edwards)
Center for Disease Control
Clinical Immunization Safety Assessment (CISA)
Project Role: Co-Investigator
- 08/01/2020 - 04/30/2025 1R01 NS118483-01 (Trevathan)
NIH/NINDS
Childhood Status Epilepticus and Epilepsy Determinants of Outcome
Role: Co-Investigator
- 09/01/2020 – 09/30/2021 5U01HG007674-08 (Hamid)
NIH
Vanderbilt Center for Undiagnosed Diseases
Role: Co-Investigator

Past

- 2003 – 2004 “Preclinical Therapeutic Evaluation in A Mouse Epilepsy Model Of Tuberous Sclerosis Complex”
Principal Investigator: **Kevin C. Ess**
Funding agency: National Epifellows Foundation
- 2005 – 2006 “Tuberous Sclerosis Natural History Database”
Principal Investigator: **Kevin C. Ess**
Funding agency: Centers for Disease Control
- 2006 – 2007 “The Role of *Tsc* Genes During Brain Development”
Principal Investigator: **Kevin C. Ess**
Funding agency: McDonnell Foundation, Washington University
(Withdrawn after acceptance)

- 2007 “GABAergic Interneurons in Tuberous Sclerosis Complex”
Milken Family/American Epilepsy Society Development Award
Mentor Robert Macdonald, M.D., Ph.D.
Principal Investigator: **Kevin C. Ess**
Funding agency: American Epilepsy Society
- 2007 – 2012 “The Role of the TSC Genes During Brain Development”
K08 Mentored Clinical Scientist Research Career Development Award
Mentor Christopher Wright, Ph.D. (previous mentor Pat Levitt, Ph.D.)
Principal Investigator: **Kevin C. Ess**, MD, PhD
5K08NS050484, Funding agency: NINDS, NIH
- 2007 – 2010 “Role of the Tuberin GAP Domain in Neural Progenitor Cells”
Junior Investigator Research Award
Principal Investigator: **Kevin C. Ess**, MD, PhD
Funding agency: Tuberous Sclerosis Alliance
- 2008 – 2009 iPS Cell Equipment Grant
Principal Investigators: **Kevin C. Ess**, M. Gallagher, A. Bowman
Vanderbilt Institute for Clinical Translational Research
- 2009 “Spinning Disk Confocal Microscope System”
Principal Investigator: Donna Webb, Ph.D.
Co-P.I. (with 7 other Vanderbilt faculty) **Kevin C. Ess**, MD, PhD
National Center For Research Resources, NIH
1S10RR025524-01
- 2009 – 2012 “Neurogenesis in Tuberous Sclerosis Complex”
Clinical Scientist Development Award
Principal Investigator: **Kevin C. Ess**, MD, PhD
2009032, Funding agency: Doris Duke Charitable Foundation
- 2010 – 2013 “Zebrafish Model of TSC” (In NCE through February 2014)
Principal Investigator: **Kevin C. Ess**, MD, PhD
W81XWH-10-1-0854, Funding agency: Dept. of Defense
Total Award: \$673,000
- 2012 – 2018 “Regulation of Neurogenesis in TSC by mTORC1 and mTORC2”
Principal Investigator: **Kevin C. Ess**, MD, PhD
1R01 NS078289, Funding agency: NINDS, NIH
(no cost extension through May 31st 2018)
- 2018 “7th Symposium on ATP1A3 function in health and disease” Chicago, IL October 13-14 2018
Principal Investigators: Brashear, Allison (Contact); **Ess, Kevin C**; George, Alfred
1 R13 NS108697-01

Publications:

Book Chapters/Books

1. **Ess KC**, and Gutmann D. Tuberous Sclerosis Complex and Other Neurocutaneous Disorders, and Neurofibromatosis. In: *Saunders Manual of Neurologic Practice*. (R. Evans, editor). WB Saunders: Orlando. (2003).

2. **Ess KC**. Modeling Tuberous Sclerosis Complex: Brain Development and Hyperexcitability. In: *Animal Models of Epilepsy*. (S. Baraban, editor). Humana Press: Totawa, New Jersey (2008).
3. Neely MD, Tidball AM, **Ess KC**, Bowman AB. Induced pluripotent stem cells (iPSCs) - an emerging model system for the study of human neurotoxicology. (2010). In: **Neuromethods: Cell Culture techniques**. Humana Press: New York, NY. Editors: Michael Aschner, Cristina Suñol, Anna Price.
4. Bowman AB, **Ess KC**, Kumar KK, Summar KL. Chapter 130 – Down Syndrome (2012). In: **Comprehensive Developmental Neuroscience**. Elsevier.
5. Bassel Abou-Khalil, Hasan Sonmezturk, **Kevin Ess**, Karl E Misulis *Atlas of EEG, Seizure Semiology, and Management, 3rd Edition*. Oxford Press (2021)

Review Articles/Editorials/Book Review

1. **Ess KC**. The Neurobiology of Tuberous Sclerosis Complex. *Seminars in Pediatric Neurology*.13:37-42, (2006).
2. **Ess KC**. Tuberous Sclerosis Complex: Everything Old is New Again. *Journal of Neurodevelopmental Disorders* (1 (2): 141-149, (2009).
3. **Ess KC**. Treatment of infantile spasms in tuberous sclerosis complex: dismal outcomes but future hope? *Nature Clinical Practice Neurology* (2009) ;5(2):72-3
4. **Ess KC**. Tuberous Sclerosis Complex: A Brave New World? *Current Opinion in Neurology*. (2010) Apr;23(2):189-93.
5. **Ess KC** and E. Steve Roach. New therapies for tuber-less sclerosis: White matter matters? *Neurology* (2012) Feb 21;78(8):520-1. Epub 2012 Jan 18.
6. **Ess KC**, Review of Pediatric Neurology: A Color Handbook by James F. Bale, Jr., Joshua L. Bonkowsky, Francis M. Filloux, Gary L. Hedlund, Paul D. Larsen, and Denise M. Nielsen, 350 pp., Mason Publishing, (2012), for *Neurology* 2012.
7. **Ess KC**. Patient Heal Thyself: Modeling and Treating Neurological Disorders Using Patient Derived Stem Cells *Experimental Biology and Medicine* 2013 Mar;238(3):308-14. doi: 10.1177/1535370213480713.
8. **Ess KC** and H. Chugani. Dynamic tubers in Tuberous Sclerosis Complex: A Window for Intervention? *Neurology* 2015 Oct 2. pii: 10.1212/WNL. [Epub ahead of print]
9. Sahin M, Henske EP, Manning BD, **Ess KC**, Bissler JJ, Klann E, Kwiatkowski DJ, Roberds SL, Silva AJ, Hillaire-Clarke CS, Young LR, Zervas M, Mamounas LA. Advances and Future Directions for Tuberous Sclerosis Complex Research: Recommendations From the 2015 Strategic Planning Conference. Tuberous Sclerosis Complex Working Group to Update the Research Plan. *Pediatr Neurol*. 2016 Jul;60:1-12. doi: 10.1016/j.pediatrneurol.2016.03.015. Review. PMID: 27267556
10. **Ess KC** and Franz DN. Everolimus for Cognition/Autism in Children with TSC: Definitive Outcomes Deferred. *Neurology* 2019

Government Reports

1. Hannah G. Rosenblum, Stephen C. Hadler, Danielle Moulia, Tom T. Shimabukuro, John R. Su, Naomi K. Tepper, **Kevin C. Ess**; Emily Jane Woo; Adamma Mba-Jonas, Meghna Alimchandani, Narayan Nair, Nicola P. Klein, Kayla E. Hanson, Lauri E. Markowitz, Melinda Wharton, Veronica V. McNally, José R. Romero, H. Keipp

Talbot, Grace M. Lee, Matthew F. Daley, Sarah A. Mbaeyi, Sara E. Oliver. Use of COVID-19 Vaccines After Reports of Adverse Events Among Adult Recipients of Janssen (Johnson & Johnson) and mRNA COVID-19 Vaccines (Pfizer-BioNTech and Moderna): Update from the Advisory Committee on Immunization Practices — United States, July 2021. *MMWR Morb Mortal Wkly Rep.* ePub: 10 August 2021. DOI: http://dx.doi.org/10.15585/mmwr.mm7032e4external_icon.

Peer Reviewed Original Research

1. **Ess KC**. Hutton JJ. Aronow BJ. Double-stranded phosphorothioate oligonucleotide modulation of gene expression. *Annals of the New York Academy of Sciences.* 716:321-3, 1994.
2. Hutton JJ. **Ess KC**. Witte DP. Aronow BJ. c-Myb and the coordinate regulation of thymic genes. *Transactions of the American Clinical & Climatological Association.* 107:115-24, 1995.
3. **Ess KC**. Whitaker TL. Cost GJ. Witte DP. Hutton JJ. Aronow BJ. A central role for a single c-Myb binding site in a thymic locus control region. *Molecular & Cellular Biology.* 15(10):5707-15, 1995.
4. **Ess K**. Chen H. Kier A. Brackenbury R. Suppression of tumorigenicity, but not invasion, in glioblastoma/HeLa cell hybrids. *Journal of Cellular Physiology.* 162(3):341-7, 1995.
5. **Ess KC**. Witte DP. Bascomb CP. Aronow BJ. Diverse developing mouse lineages exhibit high-level c-Myb expression in immature cells and loss of expression upon differentiation. *Oncogene.* 18(4):1103-11, 1999.
6. Wong M, **Ess K**, Landt M. Cerebrospinal fluid neuron-specific enolase following seizures in children: role of etiology. *Jrl.Child Neurol.* 17(4):261-4, 2002.
7. Wong M, **Ess K**, Uhlmann E, Jansen L, Li W, Phelan C, Crino P, Mennerick P, Yamada K, Gutmann D. Astrocyte-Specific *Tsc1* Inactivation Leads to Impaired Glutamate Transport in a Mouse Epilepsy Model of Tuberous Sclerosis Complex. *Annals of Neurology* 54 251-56, 2003
8. **Ess K**, Uhlmann E, Li W, Hongzhen L, DeClue J, Crino P, Gutmann D. Expression Profiling in Tuberous Sclerosis Complex (TSC) Knockout Mouse Astrocytes to Characterize Human TSC Brain Pathology. *Glia* 46 28-40 2004.
9. **Ess K**, Camp K, Tu B, Gutmann D. Developmental Origin of Subependymal Giant Cell Astrocytoma in Tuberous Sclerosis Complex. *Neurology* 64, 1446-1449, 2005.
10. Zeng L, Ouyang Y, Gazit V, Cirrito J, Jansen L, **Ess K**, Yamada K, Wozniak D, Holtzman D, Gutmann D., Wong M. Abnormal glutamate homeostasis and impaired synaptic plasticity and learning in a mouse model of tuberous sclerosis complex. *Neurobiology of Disease.* 2007.
11. Kim, S. H., Speirs, C. K., Solnica-Krezel, L. and **Ess, K. C.** (2011) Zebrafish model of tuberous sclerosis complex reveals cell-autonomous and non-cell-autonomous functions of mutant tuberin. *Dis Model Mech* 4(2): 255-67.
12. Fu C, Cawthon C, Clinkscales W, Bruce A, Winzenburger P and **Ess, K.** GABAergic Interneuron Development and Function is Modulated by the *Tsc1* Gene. *Cerebral Cortex* (2011).
13. Carson R, Van Nielen D, Winzenburger P and **Ess, K.** Neuronal and glia abnormalities in *Tsc1*-deficient forebrain and partial rescue by rapamycin. *Neurobiology of Disease* (2012) 10.1016/j.nbd.2011.08.024
14. Neely MD, Litt M, Tidball A, Li G, Aboud A, Hopkins C, Chamberlin R, Hong C, **Ess KC**, and Bowman A. DMH1, a highly selective small molecule BMP inhibitor promotes neurogenesis of hiPCs: *ACS Chemical Neuroscience* (2012) Jun 20;3(6):482-91. Epub 2012 Mar 5. PMID: 22860217

15. Armour, EA, Carson O, and **Ess KC**. "Cystogenesis and Elongated Primary Cilia in Tsc1-Deficient Distal Convolved Tubules" *American Journal of Physiology - Renal Physiology* 303(4):F584-92 (2012). PMID:22674026
16. Carson, RP, Fu, C, Winzenburger P and **Ess, KC**. "Deletion of Rictor in Neural Progenitor Cells Reveals Contributions of mTORC2 Signaling to Tuberous Sclerosis Complex" *Human Molecular Genetics* 22:140-52 (2013). PMID: 23049074
17. Aboud, A, Tidball, A, Kumar, K, Neely, D, **Ess KC**, Erikson, KM and Bowman, AB, "Genetic risk for Parkinson's disease correlates with alterations in neuronal manganese sensitivity between two human subjects" *Neurotoxicology* (2012). Dec;33(6):1443-9. doi: 10.1016/j.neuro.2012.10.009. Epub 2012 Oct 22. PMID:23099318
18. Fu, C, and **Ess, KC**. "Conditional and Domain Specific Inactivation of the *Tsc2* Gene in Neural Progenitor Cells" *genesis* (2013). PMID:23359422
19. Boglev, Y, Badrock, P, Trotter, A, Du, Q, Richardson, E, Parslow, A, Markmiller, S, Hall, N, de Jong-Curtain, T, Ng, A, Verkade, H, Ober, E, Field, H, Shin, D, Shin, C, Hannan, K, Hannan, R, Pearson, R, Kim, S-H, **Ess, KC**, Lieschke, G, Stainier, D and Joan K. Heath, "Autophagy Induction is a Tor and Tp53-independent Cell Survival Response in a Zebrafish Model of Disrupted Ribosome Biogenesis" *PLOS Genetics* (2013) 9(2):e1003279. doi: 10.1371/journal.pgen.1003279. Epub 2013 Feb 7. PMID:23408911
20. Kim SH, Carson, R, Kowalski N, Bridges, R and **Ess, KC**. "Genetic interaction of *tsc2* and *p53* enhance tumorigenesis in zebrafish" *DMM* (2013). Jul;6(4):925-33. doi: 10.1242/dmm.011494. Epub 2013 Mar 27. PMID:23580196
21. Kim, SH, Scott, SA, Bennett, MJ, Carson, RP, Brown, HA, and **Ess, KC**. "Multi-organ Abnormalities and mTORC1 Activation in Zebrafish Model of Multiple Acyl-CoA Dehydrogenase Deficiency" *PLOS Genetics* (2013) Jun;9(6):e1003563. doi: 10.1371/journal.pgen.1003563. Epub 2013 Jun 13. PMID:23785301
22. International Tuberous Sclerosis Complex Consensus Group (**Ess KC**) Tuberous Sclerosis Complex Surveillance and Management: Recommendations of the 2012 International Tuberous Sclerosis Complex Consensus Conference *Pediatric Neurology* [Volume 49, Issue 4](#) , Pages 255-265, October 2013.
23. International Tuberous Sclerosis Complex Consensus Group (**Ess KC**) Tuberous Sclerosis Complex Diagnostic Criteria Update: Recommendations of the 2012 International Tuberous Sclerosis Complex Consensus Conference *Pediatric Neurology* [Volume 49, Issue 4](#) , Pages 243-254, October 2013.
24. Srinivasakumar, N, Zaboikin, M, Tidball, A, Aboud, A., Neely, N, **Ess, K**, Bowman, A, Schuening, F. "Gammaretroviral vector encoding a fluorescent marker to facilitate detection of reprogrammed human fibroblasts during iPSC generation". *PeerJ* 2013 Dec 10;1:e224. doi: 10.7717/peerj.224. eCollection 2013.
25. West JD, Austin ED, Gaskill C, Marriott S, Baskir R, Bilousova G, Jean JC, Hemnes AR, Menon S, Bloodworth NC, Fessel JP, Kropski JA, Irwin DC, Ware LB, Wheeler LA, Hong CC, Meyrick BO, Loyd JE, Bowman AB, **Ess KC**, Klemm DJ, Young PP, Merryman WD, Kotton D, Majka SM. Identification of a Common Wnt Associated Genetic Signature Across Multiple Cell Types in Pulmonary Arterial Hypertension. *Am J Physiol Cell Physiol*. 2014 May 28. pii: ajpcell.00057.2014. [Epub ahead of print]
26. Asad A Aboud; Andrew M Tidball; Kevin K Kumar; M D Neely; Bingying Han; **Kevin C Ess**; Charles C Hong; Keith M Erikson; Peter Hedera. PARK2 patient neuroprogenitors show increased mitochondrial sensitivity to copper. *Neurobiol Dis*. 2014 Oct 12;73C:204-212. doi: 10.1016/j.nbd.2014.10.002. PMID: 25315681

27. de Vries, P, Vicky H Whittemore, Loren Leclezio, Anna Byars, David Dunn, **Kevin C. Ess**, Dena Hook, Brian King, Mustafa Sahin, Anna Jansen. Tuberous Sclerosis Associated Neuropsychiatric Disorders (TAND) and the TAND Checklist *Pediatric Neurology*. 2015 Jan;52(1):25-35. doi: 10.1016/j.pediatrneurol.2014.10.004. Epub 2014 Oct 16. PMID: 25532776
28. Thomas LR, Wang Q, Grieb BC, Phan J, Foshage AM, Sun Q, Olejniczak ET, Clark T, Dey S, Lorey S, Alicie B, Howard GC, Cawthon B, **Ess KC**, Eischen CM, Zhao Z, Fesik SW, Tansey WP. Interaction with WDR5 Promotes Target Gene Recognition and Tumorigenesis by MYC. *Mol Cell*. 2015 Mar 25. pii: S1097-2765(15)00142-2.
29. Tidball AM, Bryan MR, Uhouse MA, Kumar KK, Aboud AA, Feist JE, **Ess KC**, Neely MD, Aschner M, Bowman AB. A novel manganese-dependent ATM-p53 signaling pathway is selectively impaired in patient-based neuroprogenitor and murine striatal models of Huntington's disease. *Hum Mol Genet*. 2015 Apr 1;24(7):1929-44.
30. Seok-Hyung Kim, Shu-Yu Wu, Jeong-In Baek, Soo Young Choi, Yanhui Su, Charles R. Flynn, Joshua T. Gamse, **Kevin C. Ess**, Gary Hardiman, Joshua H. Lipschutz, Najj N. Abumrad, Don C. Rockey. A Post-Developmental Genetic Screen for Zebrafish Models of Inherited Liver Disease. *PLOS One*. 2015 May 7;10(5):e0125980. doi: 10.1371/journal.pone.0125980. eCollection 2015.
31. Nathaniel D Kelm, Kathryn L West, Robert P Carson, Daniel F Gochberg, **Kevin C Ess**, Mark Does. Evaluation of Diffusion Kurtosis Imaging in Hypomyelinated Mouse Models. *NeuroImage*. 2015 PMID: 26400013.
32. Robert Carson, Nathaniel Kelm, Kathryn West, Mark Does, Mark, Cary Fu, Grace Weaver, Eleanor McBrier, Mark Grier, and **Kevin C Ess**, Hypomyelination following deletion of *Tsc2* in oligodendrocyte precursors". 2015 *Annals Clinical and Translational Neurology*. PMID: 26982737.
33. Chun YW, Balikov DA, Feaster TK, Williams CH, Sheng CC, Lee JB, Boire TC, Neely MD, Bellan LM, **Ess KC**, Bowman AB, Sung HJ, Hong CC. Combinatorial polymer matrices enhance in vitro maturation of human induced pluripotent stem cell-derived cardiomyocytes. *Biomaterials*. 2015 Oct;67:52-64. PMID: 26982737.
34. Tidball AM, Neely MD, Chamberlin R, Aboud AA, Kumar KK, Han B, Bryan MR, Aschner M, **Ess KC**, Bowman AB. Genomic Instability Associated with p53 Knockdown in the Generation of Huntington's Disease Human Induced Pluripotent Stem Cells. *PLoS One*. 2016 Mar 16;11(3):e0150372. PMID: 26982737.
35. West KL, Kelm ND, Carson RP, Gochberg DF, **Ess KC**, Does MD Myelin Volume Fraction Imaging with MRI. *Neuroimage*. 2016 Dec 23. pii: S1053-8119(16)30793-5. doi: 10.1016/j.neuroimage.2016.12.067. PMID: 28025129
36. Hendrik Rosewich, Matthew Thomas Sweney, Suzanne DeBrosse, **Kevin Ess**, Laurie Ozelius, Eva Andermann, Frederick Andermann, Gene Andrasco, Alice Belgrade, Allison Brashear, Sharon Ciccodicola, Lynn Egan, Al George, Aga Lewelt, Joshua Magelby, Mario Merida, Tara Newcomb, Vicky Platt, Dominic Poncelin, Sandra Reyna-Merida, Masayuki Sasaki, Marcio Sotero De Menezes, Kathleen Sweadner, Louis Viollet, Mary Zupanc, Kenneth Silver, and Kathryn J. Swoboda. Research Conference Summary from the 2014 International Task Force On ATP1A3-Related Disorders. *Neurology Genetics* 2017 Mar 2;3(2) PMID: 28293679
37. Armstrong LC, Westlake G, Snow JP, Cawthon B, Armour E, Bowman AB, **Ess KC**. Heterozygous loss of *TSC2* alters p53 signaling and human stem cell reprogramming. *Hum Mol Genet*. 2017 Sep 5. doi: 10.1093/hmg/ddx345. PMID:2897354
38. Grier M, West K, Kelm N, Fu C, Does M, Parker B, McBrier, Lagrange A, **Ess KC**, Carson R. Loss of mTORC2 signaling in oligodendrocyte precursor cells delays myelination. *PLoS One*. 2017 Nov 21;12(11):e0188417. PMID: 29161318

39. Xie Y, Schutte RJ, Ng NN, **Ess KC**, Schwartz PH, O'Dowd DK. Reproducible and efficient generation of functionally active neurons from human iPSCs for preclinical disease modeling. *Stem Cell Res.* 2017 Dec 9;26:84-94. doi: 10.1016/j.scr.2017.12.003. [Epub ahead of print] PubMed PMID: 29272856
40. Simmons C, Thompson C, Cawthon BE, Westlake G, Swoboda KJ, Kiskinis E, **Ess KC**, and George A. Direct Evidence of Impaired Neuronal Na/K-ATPase Pump Function in Alternating Hemiplegia of Childhood. *Neurobiology of Disease.* 2018 Jul;115:29-38. PubMed PMID: 29567111
41. Darcy A. Krueger, Jamie K. Capal, Paolo Curatolo, Orrin Devinsky, **Ess KC**, Michal Tzadok, Mary K. Koenig, Vinodh Narayanan, Federico Ramos, Sergiusz Jozwiak, Petrus de Vries, Anna C. Jansen, Michael Wong, David Mowat, Stephanie Bruns, David Neal Franz. Safety of mTOR Inhibitors in infants and very young children with Tuberous Sclerosis Complex (TSC): *Eur J Paediatr Neurol.* 2018 Jul 4. pii: S1090-3798(17)31969-4. PMID: 30005812
42. Rushing, G, Brockman, A, Bollig, M, Leelatian, N, Mobley, B, Irish, J, **Ess, K**, Fu, K, Ihrie, RA. Location-dependent maintenance of an intrinsic susceptibility to mTORC1-driven tumorigenesis. *Life Science Alliance* (2019) PMID: 30910807, PMCID: PMC6435042
43. Emma H. Neal, Nicholas A. Marinelli, Yajuan Shi, P. Mason McClatchey, Kylie M. Balotin, Dalton R. Gullett, Kameron A. Hagerla, Aaron B. Bowman, Kevin C. **Ess**, John P. Wikswo, and Ethan S. Lippmann. A simplified, fully defined differentiation scheme for producing blood-brain barrier endothelial cells from human iPSCs. *Stem Cell Reports.* 2019 Jun 11;12(6):1380-1388. doi: 10.1016/j.stemcr.2019.05.008. PMID: 31189096
44. Short B, Kozek L, Harmsen H, Zhang B, Wong M, **Ess KC**, Fu C, Nafter R, Pearson M, Carson RP. Cerebral aquaporin-4 expression is independent of seizures in tuberous sclerosis complex. *Neurobiol Dis* 2019 May 9. pii: S0969-9961(19)30118-4. doi: 10.1016/j.nbd.2019.05.003.
45. Gewin L, Hopp K, Summers M, Harral J, Gaskill C, Nlandu Khodo S, Neelisetty S, Sullivan T, Reese J, Klemm D, Kon V, **Ess K**, Shi W, Majka S. Inactivation of *Tsc2* in *Abcg2* Lineage Derived Cells Drives the Appearance of Polycystic Lesions and Fibrosis in the Adult Kidney. *American Journal of Physiology-Renal Physiology* (In Press)
46. Xie Y, Ng NN, Safrina OS, Ramos CM, **Ess KC**, Schwartz PH, Smith MA, O'Dowd DK. Comparisons of dual isogenic human iPSC pairs identify functional alterations directly caused by an epilepsy associated SCN1A mutation. *Neurobiol Dis.* 2019 Nov 28;134:104627. doi: 10.1016/j.nbd.2019.104627. PMID: 31786370
47. Klofas L, Short, B, Snow, J, Sinnaeve J, Rushing G, Weinstein W, Westlake, W, Ihrie R, **Ess KC**, Carson, R. DEPDC5 haploinsufficiency drives increased mTORC1 signaling and abnormal morphology in human cortical neurons. *Neurobiology of Disease* 143: 10495- (2020). PMID: 32574724, PMCID: PMC7462127
48. Snow, J.P., Westlake, G., Klofas, L.K., Jeon, S., Armstrong, L.C., Swoboda, K.J., George, A.L, Jr., and **Ess, K.C.** Neuronal modeling of alternating hemiplegia of childhood reveals transcriptional compensation and replicates a trigger-induced phenotype. *Neurobiol Dis*, 2020. PMID: 32348881
49. Fu C, Armstrong L, Short B, **Ess KC**. Decreased neocortical interneuron survival and altered growth and behavior in conditional interneuron *Tsc2*-deficient mice. *Neurobiology of Disease* (Under revision)
50. Keller S, Mallack E, Rubin J, Accardo J, Brault J, Corre C, Elizondo C, Garafola J, Jackson-Garcia A, Rhee J, Seeger E, Shullanberger K, Tourjee A, Trovato M, Waldman A, Wallace J, Wallace M, Werner K, White A, **Ess KC**, Becker C, Eichler F. Neurology Care Guidelines for Pediatric Leukodystrophy. *Journal of Child Neurology* 2020. PMID: 32875938
51. Dusty R. Miller, David K. Schaffer, M. Diana Neely, Ethan S. McClain, Adam R. Travis, Frank E. Block III, Jennifer McKenzie, Erik M. Werner, Laura Armstrong, Dmitry A. Markov, Aaron B. Bowman, **Kevin C. Ess**,

David E. Cliffler, and John P. Wiksw. A bistable, multiport valve enables microformulators creating microclinical analyzers that reveal aberrant glutamate metabolism in astrocytes derived from a tuberous sclerosis patient. *Sensors & Actuators: B. Chemical* Aug 2021. PMID: 34092923

52. Lana Jeradeh Boursoulian, Victoria Adeseye, Beth A. Malow, **Kevin Ess**. Recurrence Rate of the First Non-Febrile Seizure in Children with Autism Spectrum Disorder. *Epilepsy and Behavior* (In Press 2021)

53. A. Hall, G. Westlake, B. P. Short, M. Pearson, **K. C. Ess**. *TSC1* Mosaicism leading to Subependymal Giant Cell Astrocytoma but not Tuberous Sclerosis Complex. *Pediatric Neurology* (In press 2021)