The Faculty of Medicine of Harvard University
Curriculum Vitae

Date Prepared: Friday, February 17, 2023
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Place of Birth: Evreux (France)

Education:

1992	D.E.A (Master)	Neurobiology (Alain Sans)	Université de Montpellier II, France
1996	Doctorat Européen (PhD)	Sensory Neurobiology (Alain Sans)	Université de Montpellier II, France

Postdoctoral Training:

10/96-09/99	Postdoctoral Fellow	Physiology (Jonathan F. Ashmore)	University College London, U.K.
10/99-10/01	Research Associate	Neurobiology (David Corey)	Harvard Medical School, MGH Howard Hughes Medical Institute, Boston, MA

Faculty Academic Appointments:

10/92-09/95	Instructor	Neurobiology	Université de Montpellier II, France
10/01-10/07	Assistant Professor of Research	Neuroscience and Otolaryngology	University of Virginia, VA
10/07-06/11	Associate Professor of Research	Neuroscience and Otolaryngology	University of Virginia, VA
07/11-03/21	Assistant Professor	Otolaryngology	Boston Children's Hospital Harvard Medical School, MA
04/21- Present	Associate Professor	Otolaryngology	Boston Children's Hospital Harvard Medical School, MA

Appointments at Hospitals/Affiliated Institutions:

10/99-10/01	Research Associate	Neurobiology	Massachusetts General Hospital
07/11- present	Research Associate with Harvard Appointment		Boston Children's Hospital

Major Administrative Leadership Positions:

08/19-04/21	Director of Student Affairs	Harvard Graduate Program in Speech, Hearing Bioscience and Technology	Harvard Medical School, MA
04/21-02/22	Associate Director	Harvard Graduate Program in Speech, Hearing Bioscience and Technology	Harvard Medical School, MA
02/22-present	Director	Harvard Graduate Program in Speech, Hearing Bioscience and Technology	Harvard Medical School, MA

Committee Service:

Local

2005-2007	Curriculum Committee	University of Virginia/ Neuroscience Graduate Program Member
2009	Focus Group	University of Virginia/ Neuroscience Graduate Program Member
2009-2014	Thesis Committee- Michaela Levin	University of Virginia/ Neuroscience Graduate Program Examiner
2009-2015	Thesis Committee- Charles Askew	University of Virginia/ Neuroscience Graduate Program Examiner
2011-2020	Research Faculty Council: Educational Committee	Boston Children's Hospital Member
2017	Qualifying exam, Sarah Gluck	Harvard Medical School/ Harvard Speech Hearing Bioscience and Technology (SHBT) Graduate program Examiner
2017-present	Graduate Advisor (5 entering SHBT students)	Harvard Medical School/ Harvard Speech Hearing Bioscience and Technology (SHBT) Graduate program Advisor

2019-2021	Graduate Program Dissertation Advisory Committee- John Lee	Harvard Medical School/ Harvard Speech Hearing Bioscience and Technology (SHBT) Examiner
2019-present	SHBT Executive Committee	Harvard Medical School/ Harvard Speech Hearing Bioscience and Technology (SHBT) Graduate program Member Role: Review of the graduate program budget; Review of the qualifications of those applying to join the program as faculty members; Approval of graduate credits for the courses.
2019-present	SHBT tracking Committee	Harvard Medical School/ Harvard Speech Hearing Bioscience and Technology (SHBT) Graduate program Member Role: Evaluating applications for admission, and monitoring the progress and academic performance of graduate students in the program. Member
2021-2022	Joint Committee on the Status of Women (JCSW) Sub committee: Work/Life balance Dean's award	To facilitate and promote leadership, career development, professional advancement, community building, and work life integration for women faculty, trainees (fellows and residents), students (graduate, medical and dental) and staff at Harvard Medical School (HMS) and Harvard School of Dental Medicine (HSDM).
2022-	Standing Committee on Higher Degrees in Medical Sciences, Harvard University, Faculty of Arts and Sciences	Executive decision-making body, discussing and approving policy changes or issues of great importance within the Division of Medical Sciences.

International

2018	Thesis Committee- Margot Tertrais	Neuroscience Graduate Program- University of Bordeaux, France External examiner - Thesis Defense
2020	Thesis Committee- Virginia Mayeux	Neuroscience Graduate Program - University of Sussex, UK. External examiner - Thesis Defense

Professional Societies:

1996-present	Association for Research in Otolaryngology (ARO)	Affiliated Member (1996-2001) Member (2001-Present)
1997-1999	Physiological Society, UK	Affiliated Member

2004-2012	Society for Neuroscience	Member
2005- 2010	Harvard Graduate Program in Neuroscience 2005-2007 2007-2010	 Junior Faculty Member Faculty Member
2009-2012	American Physiological Society	Member
2016-present	Harvard Graduate Program in Speech, Hearing and Biotechnologies	Faculty Member
2016-present	Harvard Graduate Program in Neuroscience	Faculty Member
2018-2023	American Otological Society Research Advisory Board	Consultant (Appointed for 5 years)
2018-2020	Association for Otolaryngology (ARO)	Council member
2022-	Association for Otolaryngology (ARO)	Nominating committee

Grant Review Activities:

2010-present	Wellcome Trust	Ad hoc member
2011	Italian Ministry of Health- Health Research Grants	Ad hoc member
2013	NIH/NIDCD	Ad hoc member- Fellowship Awards
2013- 2021	Action for Hearing Loss (UK)	Ad hoc member
2014	Translational Rsesearch Center; Boston Children's Hospital	Ad hoc member
2016	NIH/NIDCD	Ad hoc member- Fellowship Awards
2017	Agir Pour l'Audition (APA)	Ad hoc member
2017	Translational Research Center, Boston Children's Hospital	Ad hoc member
2017	European Research Council	Ad hoc member
2018	Action for Hearing Loss	Ad hoc member
2018-2023	American Otological Society	AOS Research advisory board member
2019	Agence Nationale de la Recherche (France)	Ad hoc member
2019	Fondation Pour l'Audition (FPA)	Ad hoc member

2020-present	Actions Concertées Inter-Pasteuriennes (France)	Ad hoc member
2020	Harvard Medical School	Reviewer of the Blavatnik Award
2021	NIDCD/NIH	Ad hoc member
2021- 2023	RIND Hearing Research (UK)	Discovery Research Grant Review Panel

Editorial Activities:

- **Ad hoc Reviewer**

Gene Therapy; Journal of Neurophysiology; Journal of Physiology; Journal of the Association for Research in Otolaryngology (JARO); Journal of Neuroscience; Journal of Comparative Neurology; Hearing Research; Elife; Journal of Clinical Investigation; Journal of Neurophysiology; Neuron; EMBO Molecular Medicine; Human Molecular Genetics; Nature;

- **Other Editorial Roles**

2019-2021	Editorial Board Member	Scientific Report (Nature)
2020-2022	Topic Co- editor with Dr. Sadhegi	Frontier in Neuroscience- Research Topics; *Commonalities and Differences in Vestibular and Auditory Pathways*

Honors and Prizes:

1992-1995	Research Scholarship	French Ministry of Research	Awarded by order of merit to the first three D.E.A. students; Finished 1st/12
2011	Pilot Award	Manton Center for Rare Disease	Boston Children's Hospital and Harvard Medical School
2012	Prix Scientifique de l'expatriation	Racine Sud	Prize for graduates of the region "Occitanie, south of France, International representation in the Science category
2012	Pilot Project Award	Research Faculty Council	Boston Children's Hospital
2019	Kirby Innovation Award	Kirby Foundation	Boston Children's Hospital
2020	NIDCD Director recognition Award	NIDCD, NIH	Awarded to EARssentials Session Instructors for supporting or leading the first virtual EARssentials course, allowing for inner ear expertise to be passed to the next generation of inner ear researchers on an international scale during a global pandemic.
2021	ARO Pioneer Award	Association of Research in Otolaryngology	For the co-discovery of Tmc1 as the sensory transduction channel in hair cells of the inner ear.

Report of Funded and Unfunded Projects

Past

08/01- 07/08 Ion channel function in auditory and vestibular hair cells
NIDCD/RO1 DC 05439
Co-Investigator; Jeffrey Holt (PI)
This grant was focused on a family of potassium channels, known as KCNQs, and their function in hearing and deafness. I recorded and analyzed voltage-dependent potassium current in the presence and absence of numerous KCNQ constructs applied to sensory hair cells of the mouse utricle.

05/03-05/08 Stem cells and repair in the auditory and vestibular systems
NIDCD/R01 DC 006182
Co-Investigator; Jeffrey Holt (PI)
This grant took a multifaceted approach aimed at regeneration of sensory hair cells in the inner ear. My role on the project was to test the functionality of candidate hair cells.

01/04-12/06 Hair cell development in the mouse embryo
NIDCD/R03 DC006183
Principal Investigator
The objective of this grant was to provide an electrophysiological and molecular characterization of hair cell development before birth in the mouse utricle.

07/09-07/11 Functional development of hair cells
NIH-NIDCD/Administrative Supplement RO1 DC008853-S
Principal Investigator
This supplemental grant was designed to accelerate the generation of a new transgenic mouse model to assess a modified hair cell specific promoter.

11/11-10/12 Rescue of inner ear function in mouse model of Usher Syndrome type I
Manton Center Award
Principal Investigator
This grant was instrumental in allowing the import of a humanized mouse model of Usher Syndrome type I. To investigate possible treatment strategies for Usher Syndrome, I proposed to introduce wild-type harmonin at various development stages into inner ear tissue harvested from a mouse model of human USH1C using advanced generation adenoviral constructs.

07/11-07/13 Inducible expression of exogenous potassium channels in mouse hair cells
NIH-NIDCD/ R21
Collaborator; Jeffrey Holt (PI)
For this project, we proposed to develop a new mouse model using the modified myo7a promoter which I had developed with my R01 grant. This model provided a unique opportunity to decipher several functional roles of sensory hair cells in vivo (in young, mature and aging animals).

12/12-11/13 Conditional Expression of TMC genes in the Mouse Inner Ear
Pilot Project Research Award- Children's Hospital Research Council
Principal Investigator
This grant was awarded to support a startup project and the generation of preliminary data to support an upcoming grant application. With the support from the Research Council at Boston Children's Hospital we generated a new mouse model to study the function of transmembrane channel-like gene 2 (Tmc2) in hair cells of the inner ear.

04/14-03/15 International Usher Syndrome Symposium
NIH-NIDCD/R13
Principal Investigator (Co-PI with Margaret Kenna)
This grant was awarded to support a conference held at Harvard Medical School in Boston, July 10-12th, 2014 that included two days of scientific reports (talks and posters) and one family day. This award also supported attendance of 10 students and postdoctoral fellow whose research pertained to Usher Syndrome.

12/14-12/16 Gene therapy for animal model of retinal degeneration
Foundation Fighting Blindness (FFB)
Consortium agreement with MEEI; PI: Luk Vandenberghe
To assess treatment strategies for hearing impairment in a mouse model of Usher syndrome type IC

1/15-12/17 Development of Genome Editing Tools to Target Deafness Genes
Bertarelli Foundation
Co- Investigator; Jeffrey Holt and Patrick Aebischer (PIs)
For this project we developed new technologies to treat inherited forms of deafness. These new gene therapy technologies were developed to allow repair of specific DNA sequences in the mutant genes of inner ear sensory cells. Our goal was to restore hearing function in two mouse models of human deafness.

6/1/2017 Development of a knockout mouse model of USH2A
IDDRC, BCH
Principal Investigator;
With the assistance of the Mouse Gene Manipulation Core facility at Boston Children's Hospital, we proposed to generate a knockout mouse model of USH2A with targeted deletion using the CRISPR/Cas9 technology.

12/17-11/18 International Usher Syndrome Conference
NIDCD/ R13 DC016829
Principal Investigator (Margaret Kenna, Co-PI);
This grant supported the second International Symposium on Usher Syndrome and 10th Annual USH Connection Conference that will be held in Mainz, Germany from July 19-21, 2018. This conference brought together scientists, clinicians and USH patients and families

07/14-09/16 Rescue of inner ear function in mouse model of Usher Syndrome type I
Kids B Kids 07/14-09/19
Principal Investigator;
For this project we investigated the use of novel therapeutics to treat genetic inner ear disorders, with focus on antisense oligonucleotides to rescue function in hair cells of USH1C mice.

01/19-07/20 The inner ear as a model for the development of gene therapy for Peroxisome Biogenesis Disorders
The Global Foundation for Peroxisomal Disorders (GFPD);
Principal Investigator;
This proposal aimed to assess inner ear gene therapy for human peroxisomal biogenesis disorders (PBD) and Zellweger syndrome spectrum disorders (ZSD). As PBD-ZSD patients suffer from moderately severe to profound hearing loss, we seek to use the enclosed inner ear as a model for the development and assessment of viral gene therapies for this disease. An R21 proposal was submitted in October, 2020 to continue this work.

01/19-07/20 Generation of a large animal model for USH2A
Usher Syndrome Society (Seed funding in response of RFP); And Gift from a patient
Principal Investigator;

Gene manipulation in large animals is now highly efficient thanks to the use of the CRISPR/Cas9 system to generate animals with altered DNA sequences. In this proposal, we aimed to take advantage of the technology to transfer CRISPR/Cas9 constructs to induce in the pig genome a common *USH2A* mutation found in humans.

07/19-07/20 A Pig Model for Usher Syndrome for Advancement of Gene Therapy Toward Clinical Application
Kirby Innovation Award
Principal Investigator;
Funds for this pilot project were used to characterize and perform the initial phenotyping of the new USH2A swine model. Preliminary data from this project were used to secure industry sponsorship to support translation of the inner ear gene therapy into clinical application.

Current

01/16-01/25 Research in Gene Therapy for Genetic Deafness
Jeff and Kimberly Barber Fund for Gene Therapy
Co-Principal Investigator
Total direct cost: $950,000 (until 01/2025)
For this project we are developing novel therapies for genetic and acquired forms of hearing loss.

07/20-12/23 Development of Gene Therapy for Usher 2A
BioMarin Pharmaceuticals- Sponsored Research Agreement
Principal Investigator
Total direct cost: $377,771
The purpose of this collaborative project is to develop an AAV gene therapeutic for the treatment of patients with USH2A. The strategy will be designed for treatment of the auditory manifestations of the disease but may also be useful for treating the retinal disease.

04/07-11/25 Functional Development of Hair cells and Neurons in the Inner Ear
NIDCD /RO1 DC008853
Principal Investigator
Total Direct cost: $1,914,648
This R01 grant has provided continuous funding for my laboratory since April 2007. Currently the work focusses on the study of the maturation of the peripheral auditory circuit which largely takes place during the first 2 post-natal weeks in mice. We hypothesize that modulation by external inputs and sensory hair cell activity including sensory transduction may affect this maturation process. With this proposal, we seek to establish how changes in sensory transduction alter maturation of the synapse and innervation to the hair cells and determine if gene therapy can restore normal function.

10/20-09/24 Gene Therapy for Deafness in the Middle Eastern Population
Us-Israel Binational Science Foundation
Principal Investigator
Total direct cost: $160,000
This project is designed to develop and validate state-of-the-art gene therapies to rescue hearing in mouse models of Middle Eastern deafness variants.

01/22-12/23 Piloting Preclinical Development of Patient-Customized ASO Therapies for Usher Syndrome
Usher Syndrome Society (FRA- Invitation upon LOI)
Principal Investigator; Co-PI: Timothy Yu
Total direct cost: $125,000 (Y1- Budget for Y2 will be determined upon progress report)
The goal of this project is to develop and test novel gene therapies for treating genetic hearing and vision loss in human patients suffering from Usher syndrome, type 2A.

01/23-12/27 Vestibular dysfunction and the development of therapies for Usher syndrome
NIDCD/NIH- R01 DC020243-01A1
Principal Investigator (MPI Project)
This research project aims to use multidisplinary approaches to define the progression of vestibular dysfunction in an Usher syndrome type 1C mouse model and in patients, and further test two novel treatments (antisense and gene replacement) that restore balance in the mouse model. Success in this project will provide the foundation for translation of the treatments into clinical trials.

Unfunded Current Projects

2019- Mechanisms and therapies for Hearing loss associated with Zellweger Spectrum Disorders
This project aims to assess inner ear gene therapy for human peroxisomal biogenesis disorders (PBD) and Zellweger syndrome spectrum disorders (ZSD). As PBD-ZSD patients suffer from moderately severe to profound hearing loss, we seek to use the enclosed inner ear as a model for the development and assessment of viral gene therapies for this disease.

2020- Characterization of novel mouse models of Usher Syndrome Type III
In collaboration with Dr. Dinculescu from the University of Florida, Gainesville, we are studying several novel mouse models of USH3, a deaf-blindness syndrome. Currently we are processing inner ear tissues from two different HA-tagged mouse models and determining the auditory phenotype of a new mouse model for a non-sense human mutation in USH3.

Report of Local Teaching and Training

NIH Training Grants:

Project Number:	T32DC000038
Contact PI / Project Leader:	**Geleoc, Gwenaelle**
Title:	Training for Speech and Hearing Science
Awardee Organization:	Harvard Medical School

Project Number:	5T32NS007473-17
Contact PI / Project Leader:	Schwarz, Thomas L.
Title:	Developmental Neurology
Awardee Organization:	Boston Children's Hospital

Teaching Students in Courses:

2015	Speech and Hearing Bioscience and Technology MIT and Harvard Graduate program Lecture and paper discussion	Massachusetts Eye and Ear Infirmary, Boston 2 hrs

Formal Teaching of Residents, Clinical Fellows and Research Fellows (post-docs):

Post-doc: 09/10-07/11 **Varadamurthy Srinivasanarada,** PhD/ Associate Director Regulatory Affairs/ AstraZeneca, Gaithersburg, MD
Varada was a postdoctoral fellow working on a project funded by my R01 grant and supplement. I supervised his research and monitored his progress routinely.

Post-doc: 04/06-01/10 03/11-12/14 Instructor: 01/15-04/19	**Yukako Asai**, PhD/ Senior Scientist, Akouos, Boston, MA Yukako designed different constructs that we used to produce new mouse models to study hearing and deafness and to assess the role of Transmembrane Channel-Like (TMC) genes in hair cell function. I worked closely with her and mentored her towards a more independent position. She was appointed to the level of Instructor in Otolaryngology at Harvard Medical School in 2014.
Post-doc: 01/19-07/20	**Xiaohan Wang,** PhD Xiahoan has worked with me on the development of electrophysiological recordings from IHCs synapses.
Post-doc: 10/2019-present	**Stephanie Mauriac, PhD** Stephanie works on two projects: 1) The study of the role of peroxisome on hair cell function and survival and how mutations associated with Peroxisomal Biogenesis Disorders affect hearing. 2) The development of gene therapy for Usher syndrome IIA (USH2A). Importantly, Stephanie was just awarded the **Manton Center Fellowship** to develop her own research project, now, focusing on USH1B.
Post-doc: 09/2022-present	**Thibault Peinau** Thibault received his PhD from the University of Bordeaux, France, in January 2022. He is a trained electrophysiologist and he will be working on my main R01 grant, studying the sensory cells physiology in different mouse models of altered or absent transduction.

Research Supervisory and Training responsibilities:

2011-2014	Supervision of post-doctoral research fellow/ Boston Children's Hospital	One hour lab meeting per week and 1:1 supervision 1h per week
2011-2015	Supervision of graduate students at the University of Virginia	One hour lab meeting per week and 1:1 supervision 3h per week
2014-2016	Supervision of medical faculty/ Harvard Medical School Office for Diversity Inclusion and Community Partnership	1:1 supervision 1h per week
2011-	Supervision of graduate student/ MIT and Harvard Medical School	One hour lab meeting per week and 1:1 supervision 3h per week
2011-	Supervision and training of technicians/ Boston Children's Hospital (leading to publication)	1:1 supervision 2h per week

Formally Mentored Harvard Medical, Dental and Graduate Students:

05/16	**Sarah Gluck,** SHBT Graduate student/ Boston Children's Hospital Qualifying exam committee- Speech and Hearing Harvard/MIT graduate program

10/16-11/20 **Hannah Goldberg**, SHBT Graduate student/ Boston Children's Hospital
Hannah was a graduate student in the Speech and Hearing Harvard/MIT graduate program (Entering class of 2015). She joined the lab to do a 3-month rotation studying gene therapy of Usher Syndrome and decided to do her thesis with us. As her mentor and Dissertation Advisor, I met with her regularly and offered her guidance and support along the way. Hannah successfully defended her thesis in November 2020.

10/17-08/21 **John Lee**, SHBT Graduate student/ Boston Children's Hospital, Boston
John was a third-year graduate student in the Speech and Hearing Harvard/MIT graduate program (Entering class of 2016) when he joined our lab to do a 6 month rotation. He since decided to continue his thesis in our lab. He is studying balance recovery after gene therapy treatment of several genes with mutations that result in auditory and balance deficits. We are currently preparing two papers on his thesis work.

Other mentored trainees and Faculty:

06/02-09/02 **Jessica (Risner) Janiczek**, PhD/ Current: Research Funding Manager, Research UK, London, UK
Jessica was a graduate student in the Neuroscience Program at the University of Virginia. I supervised her daily during 3-month lab rotation. The research that she performed during her rotation was the basis of her first publication in Journal of Neuroscience (Geleoc et al. 2004).

08/05-11/13 **Michaela Levin**, PhD/ Licencing Manager/ Partner's Healthcare, Boston, MA
Michaela was a graduate student in the Neuroscience Program at the University of Virginia. I supervised Michaela daily during 3-month lab rotation.

07/12-07/15 **Charlie Askew**, PhD/ Current: Postdoctoral Fellow, UNC, Chapel Hill, NC
Charlie was a graduate student in the Neuroscience Program at the University of Virginia. He had initiated the study of hair cell function in a novel mouse model of Ush1C.

07/14-07/16 **Selena E. (Heman-Ackah) Briggs**, MD, PhD, MBA, FACS/ Current: Associate Professor, Georgetown University Medical Center, Georgetown, Washington DC.
Selena was sponsored by HMS to participate in a mentored research project that she conducted under my supervision. Selena's research was focused on the rescue of auditory function in a mouse model of USH1C. This work was included in our published report (Pan et al., *Nature Biotech.* 2017).

10/14-11/16 **Alice Galvin**, Research Assistant/ Current position unknown
I trained and supervised Alice in the different tasks that she accomplished on a regular basis over the two years she worked with me. Alice became proficient with genotyping, mouse colony management and Auditory Brainstem recordings (ABR). Alice is co-first author on Pan et al. *Nature Biotech.* (2017) and a co-author on Lentz et al (2020).

06/16-07/20 **Carl Nist-Lund**, Graduate student, Program in Neuroscience, Harvard, Boston, MA
Carl was a research associate in the lab. I trained him at the bench and taught him multiple techniques that he eventually ran independently. I also met with him regularly to discuss his projects, along with his short term and long-term plans. Carl became a co-author on several of my manuscripts and was recently admitted to the Graduate Program in Neuroscience at Harvard Medical School.

Local Invited Presentations: *No presentations below were sponsored by 3rd parties/outside entities*

2011 "Rescue of inner ear function in mouse models of Usher Syndrome"/ Annual Otolaryngology Retreat, Department of Otolaryngology, BCH, Boston, MA

2011 "Development of a new transgenic mouse model to target hair cell genes"/ Kirby Lab Results, Kirby Neurobiology Center, BCH, Boston, MA

2011 "Functional and molecular development of mechanosensory complex in sensory cells in the mouse inner ear"/ Eaton Peabody Laboratory Seminar Series, Massachusetts Eye and Ear Infirmary, Boston, MA

2012 "Transmembrane channels are essential for mechanotransduction in hair cells of the auditory system"/ 8th Annual Symposium in Cellular, Molecular and Clinical Research in Surgery, BCH, Boston, MA

2012 "A Switch in TMC Expression Underlies Auditory Development"/ Kirby Neurology Faculty Retreat/ Woods Hole, MA

2013 "Rescue of Inner Ear Function in Mouse Models of Usher Syndrome"/ Joint lab presentations with the Neurobiology Department, Harvard Medical School, Boston, MA

2013 "Possible gene therapy strategies for treatment of Usher Syndrome, a family of genetic disorders that cause deafness and blindness"/ Sensory Focus Group, Kirby Neurobiology Center, BCH, Boston, MA

2014 "Rescue of Inner Ear Function in Mouse Models of Usher Syndrome"/ Kirby Neurobiology Center Faculty Talk, BCH, Boston, MA

2016 "Gene therapy restores auditory and vestibular function in a mouse model of Usher syndrome, type IC"/ Invited Speaker, EPL Seminar Series, MEEI, Boston, MA

2016 "Gene therapy restores auditory and vestibular function in a mouse model of Usher syndrome, type IC"/ Kirby Neurobiology center, Lab results, BCH, Boston, MA

2017 "Gene therapy restores hearing and balance in a mouse model of Usher syndrome type IC"/ BU-Tufts Alumni Day honoring Kenneth Grundfast, BU, Boston, MA

2017 "TMC2 partially compensates hair cells and auditory function in mice lacking TMC1 channels"/ Kirby Neurobiology Faculty Talk, Boston, MA

2018 "Gene Therapy for Usher Syndrome Type IC"/ Fetal Medicine, BCH seminar series, Boston, MA

2018 "The inner ear as a model for Peroxisomal disorders"/ Kirby Neurobiology Retreat. BCH, Boston, MA

2022 "Development of therapies for Usher syndrome"/ Harvard Brain Initiative, Boston, MA

Report of Regional, National and International Invited Teaching and Presentations

No presentations below were sponsored by 3rd parties/outside entities

Regional (Teaching and presentations given in proximity of appointment at that time)

1993-1995 Tutorial in Neurobiology
Undergraduate/ Groups of 35-40 students, 35 x 2hrs class/ year, Université de Montpellier, France

2009-2010 Lecture in Neurobiology NESC 703
Graduate students/ Group of 8-12 students, 1 x 2hrs/year
University of Virginia, VA

1995 "Mechano-electrical transduction in neonatal mammalian vestibular hair cells"/ Patch club of Montpellier, Annual meeting, Lauret, France

1996 "Mechanoelectrical transduction in neonatal mammalian vestibular and cochlear hair cells"/ Departmental Seminar Series, Department of Physiology, University of Sussex, Brighton, U.K.

1997 Tutorial in Anatomy; Undergraduate/Groups of 30 students/ 2hrs, University College London, UK

1997 Lab: Audiometry; Undergraduate/Groups of 30 students/ 2hrs, University College London, UK

1998 "Sugar transport and outer hair cell motility"/ Departmental Seminar Series, Department of Physiology, University College London, U.K.

1998 Tutorial in Biology; Undergraduate/Groups of 30 students/ 3hrs, University College London, UK

2002 "Prenatal acquisition of mechano-electrical transduction in mouse vestibular hair cells."/ Departmental NERD Seminar Series, Neuroscience Department, University of Virginia, Charlottesville,VA

2002 "Molecular motors in the sensory cells of the mammalian inner ear."/ Interdisciplinary Workshop on Sensory Systems (Talk), University of Virginia, Charlottesville

2003 "Prenatal acquisition of voltage-gated conductances in vestibular hair cells of the developing mouse embryo." / Departmental NERD Seminar Series, Neuroscience Department, University of Virginia, Charlottesville, VA

2003 "Development of sensory hair cells"/ Annual Audiology Conference, James Madison University, VA

2003-2010 Tutorial in Neurobiology/ Graduate students;12x 1hr/ year
University of Virginia, Charlottesville, VA

2003-2008 Lecture in Neurobiology NESC 703
Graduate students/ Group of 8-12 students; 2 x 1hr/ year University of Virginia, VA

2010 "A new transgenic model to study hair cell function in the mouse"/ Departmental NERD Seminar Series, Neuroscience Department, University of Virginia, Charlottesville

2011 "Rescue of inner ear function in mouse models of Usher Syndrome"/ Otolaryngology Retreat, Boston Children's Hospital, Boston

2011 "Development of a new transgenic mouse model to target hair cell genes"/ Kirby Lab Results, Kirby Neurobiology Center, Boston Children's Hospital, Boston

2011 "Functional and molecular development of mechanosensory complex in sensory cells in the mouse inner ear"/ Eaton Peabody Laboratory Seminar Serie, Massachusetts Eye and Ear Hospital, Boston

2011 Labs: Biology of the inner ear. Summer course, 3 weeks, MBL- Woods Hole, MA

2012 "Transmembrane channels are essential for mechanotransduction in hair cells of the auditory system"/ 8th Annual Symposium in Cellular, Molecular and Clinical Research in Surgery, Boston Children's Hospital, Boston, MA

2012 "A Switch in TMC Expression Underlies Auditory Development"/ Neurology Faculty Retreat, Woods Hole, MA

2013 "Rescue of Inner Ear Function in Mouse Models of Usher Syndrome"/ Joint lab presentations with the Neurobiology Department, Harvard Medical School, Boston, MA

2013 "Possible gene therapy strategies for treatment of Usher Syndrome, a family of genetic disorders that cause deafness and blindness." Sensory Focus Group., Kirby Center, BCH. Boston, MA

2013 Responsible Research: Authorship/ Department of Otolaryngology, Boston Children's Hospital/ 1x 1hr, Boston, MA

2013 Labs: Biology of the inner ear; Summer course, 3 weeks, MBL- Woods Hole, MA

2014 "Rescue of Inner Ear Function in Mouse Models of Usher Syndrome"/ Kirby Center Faculty Talk, Boston, MA

2014 "Usher Syndrome Research: Update for the families"/ International Symposium for Usher Syndrome, Boston, MA

2016 "Gene therapy restores auditory and vestibular function in a mouse model of Usher syndrome, type IC"/ Invited Speaker, EPL Seminar Series, MEEI, Boston, MA

2016 "Gene therapy restores auditory and vestibular function in a mouse model of Usher syndrome, type IC"/ Kirby Neurobiology Center, Lab results, Boston, MA

2016 "Hair cells and Mechanotransduction"/ Invited Speaker, Earssentials: Concepts and Techniques of Contemporary Hearing Research. NIDCD (NIH), Bethesda, MN

2017 Labs: Biology of the inner ear
Summer course, 2 weeks, MBL- Woods Hole, MA

2017 "Gene therapy restores auditory and vestibular function in a mouse model of Usher syndrome, type IC"/MEEI Grand rounds, Lecture, Boston, MA

2017 "Gene therapy restores auditory and vestibular function in a mouse model of Usher syndrome, type IC"/ Otolaryngology Department, Lecture, Boston, MA

2018 "Sensory transduction in the mammalian inner ear"
Brown University, Guest lecture, Undergraduate program, Providence, RI

2018 "Gene Therapy for Usher Syndrome"/ Decibel Therapeutics, Boston, MA

2019 "Sensory transduction in the mammalian inner ear"
Brown University, Guest lecture, Undergraduate program, Providence,RI

2019 "Hair cells and Mechanotransduction"- 2 lectures
Invited Speaker/ Earssentials: Concepts and Techniques of Contemporary Hearing Research. NIDCD (NIH), Bethesda, MN

2020 "Hair cells and Mechanotransduction"- 2 lectures
Invited Speaker, Remote presentation/ Earssentials: Concepts and Techniques of Contemporary Hearing Research. NIDCD (NIH), Bethesda, MN

2021 "Sensory transduction in the mammalian inner ear"
Brown University, Guest lecture, Undergraduate program, Providence, RI

2022 "Prospects for treatments of progressive hearing loss and balance dysfunction" Audiology Research Conference, St Louis, MO

2022 "Sensory transduction in the mammalian inner ear"
Brown University, Guest lecture, Undergraduate program, Providence, RI

2022 "Development of therapeutic approaches for Usher Syndrome". Midwest Auditory Research Conference, Ann Arbor, MI

Others: Oral presentations from selected abstract (Regional)

1995 **Géléoc G.S.G.*,** Lennan G.W.T., and Kros C.J. Mechanoelectrical transduction in mammalian vestibular hair cells. (1995) 32nd Inner Ear Biology Workshop- Montpellier, France

1997 **Géléoc G.S.G.*,** Lennan G.W.T., and Kros C.J. Inward rectifier potassium currents in cultured cochlear hair cells of neonatal mice. (1997) 8eme Colloque Canaux Ioniques, P36.

National (Presentations given in country of appointment at the time)

1997 "Sugar transport in isolated outer hair cells of the guinea-pig cochlea" / Physiological Society Meeting (Selected from abstract). Cambridge, U.K

2001 "Molecular motors in sensory hair cells: outer hair cell motility and transducer" /Job Talk, Department of Neuroscience, UVA, Charlottesville

2003 "Prenatal Acquisition of Voltage-Gated Conductances in Vestibular Hair Cells of the Developing Mouse Embryo"/ Association for Research in Otolaryngology, 26th Midwinter Research Meeting (Selected from abstract), St Petersburg, FL

2004 "Functional Maturation of Type I and Type II Hair Cells in the Embryonic Mouse Utricle"/ Association for Research in Otolaryngology, 27th Midwinter Research Meeting (Selected from abstract), Daytona, FL

2005 "Functional development of mechanosensory hair cells of the mammalian inner ear"/ Job Talk, Departments of Neurobiology and Otolaryngology, Stanford, Palo-Alto, CA

2010 "Putting the pieces together: How hair cells of the inner ear become functional"/ Job Talk; FM Kirby Neurobiology Center, Children Hospital, Harvard Medical School, Boston, MA

2010 "Putting the pieces together: How hair cells of the inner ear become functional"/ NIDCD Seminar Series, NIH, Bethesda, MN

2012 "Development of transduction in hair cells of the inner ear"/ Gordon Conference on Auditory Systems, Bates College, ME

2012 "Mechanotransduction in hair cells of the mouse inner ear: Development and molecular players"/ Invited Speaker, Hearing, Balance, and Chemical Senses Program (HBCS) seminar series at Kresge Hearing Research Institute (Seminar series cross-disciplinary, NIH-funded program in sensory systems to train predoctoral students and postdoctoral fellow) – University of Michigan

2016 "Molecular Pathophysiology in a mouse model of Usher Syndrome type IC"/ 39th Association for Research in Otolaryngology Meeting (Abstract), San Diego, CA

2016 "Hair cells and Mechanotransduction"/ Invited Speaker, Earssentials: Concepts and Techniques of Contemporary Hearing Research. NIDCD (NIH), Bethesda, MD

2017 "Gene Therapy Restores Auditory and Vestibular Function in a Mouse Model of Usher Syndrome Type 1C"/ Invited Speaker, University of Washington, Seattle

2017 "Direct Delivery of Antisense Oligonucleotides to the Middle and Inner Ear Improves Hearing and Balance in Usher Mice"/ 40th Association for Research in Otolaryngology Meeting (Abstract), Baltimore, MD

2018 "TMC2 partially restores hair cell and auditory function in mice lacking TMC1"/ 41th Association for Research in Otolaryngology Meeting (Abstract), San Diego, MD

2018 "Genetic Therapies for Hearing Loss in PBD-ZSD"/ Global Foundation for Peroxisomal Disorders, June 2018, Washington

2018 "Vestibular Periphery"/ Jackson Laboratories- Workshop Hear@Jax, ME

2018 "Viral Gene Therapy "/ Jackson Laboratories- Workshop Hear@Jax, ME

2019 "Gene therapy restores hearing and balance in a mouse model of Usher Syndrome type IC"/ Wayne State University, MI

2019 "Gene therapy for Usher syndrome"/ University of Mississippi Medical Center, NAS seminar series, Jackson, MS

2019 "Exploring gene replacement or antisense therapy to restore hearing and balance in a mouse model of Usher syndrome type IC"/ New York Academy of Sciences, Hair cell regeneration and Hearing restoration, New York

Others: Oral presentations from selected abstract (National)

2003 **Géléoc G.S.G.*,** Risner J.R. and Holt J.R. Prenatal Acquisition of Voltage-Gated Conductances in Vestibular Hair Cells of the Developing Mouse Embryo. (2003) *26th Association for Research in Otolaryngology*, #1071- *Vestibular Peripheral symposium*

2004 **Géléoc G.S.G.*** Risner J.R. and Holt J.R. Functional Maturation of Type I and Type II Hair Cells in the Embryonic Mouse Utricle. (2004), *27th Association for Research in Otolaryngology Meeting*, #797- *Podium: Hair cells: Stereocilia, Transduction and Motility.*

2008 Lelli A.*, Asai Y., Holt J.R. and **Géléoc G.S.G.** Developmental Acquisition of Sensory Transduction in Mouse Cochlear Hair Cells. (2008) *31th Association for Research in Otolaryngology Meeting- #486 - Podium- Hair cell Development and Transduction – Presented by A. Lelli.*

2009 Steigelman K. , Lelli A. , Wu X., **Géléoc G.S.G.,** Lin S., Gao J., Piontek K., Feng Q. Potential Roles of Pkd1 in Mechanotransduction of Mouse Hair Cells (2009) *32nd Association for Research in Otolaryngology Meeting- #511- Podium: Hair Cells: Stereocilia & Transduction – Presented by K. Steigelman*

2012 Kawashima Y.*, **Géléoc G.S.G**., Kurima K, Lelli A., Labay V., Asai Y., Makishima T., Askew C., Wu D., Holt J.*, Griffith A. Postnatal Switch in Expression of Transmembrane Channel-Like Genes 1 and 2 Underlies Mechanotransduction in Auditory Hair Cells. *35th Association for Research in Otolaryngology Meeting- #580 - Podium- Hair cell Development and Transduction – Presented by Y. Kawshima*

2012 Kawashima Y.*, **Géléoc G.S.G**., Kurima K. Labay V., Asai Y., Horwitz G., Makishima T., Wu D., Della Santina C., Griffith A., Holt J. * Mechanotransduction in Mouse Vestibular Hair Cells Requires Transmembrane Channel-Like Genes 1 or 2. *35th Association for Research in Otolaryngology Meeting- #581 - Podium- Hair cell Development and Transduction – Presented by J. Holt*

2017 Asai Y., **Géléoc. G.S.G.**and Holt J.R. An Inducible Trek1 Mouse Model and the Role of Spontaneous Activity in the Developing Auditory Organ. *Symp #27- 40th Association for Research in Otolaryngology Meeting- Presented by Y. Asai*

2018 Asai Y., Pan B., Nist-Lund C., Galvin A., Lukashin A., Ludkaskina V., Russell I., Holt J. & **Géléoc G.S.G.** (2018) TMC2 partially restores hair cell and auditory function in mice lacking TMC1. *41th Association for Research in Otolaryngology Meeting, San Diego- Podium #39 Hair cells- Presented by Y. Asai*

2019 Nist-Lund C., Pan P., Patterson A., Asai Y., Zhou W., Zhu H., Chen T., Romero S., Resnick J., Polley D.B. **Géléoc G.S.G.**, Holt J.R. (2019) Next Generation Gene Therapy Improves Hearing, Balance, and Secondary Outcomes in Mouse Models of Genetic Inner Ear Disorder. *42th Association for Research in Otolaryngology Meeting, Baltimore- presented by C. Nist-Lund*

International

1996 "Mechano-electrical transduction in mammalian vestibular and cochlear hair cells" EMBO lecture, University of Bielefeld, Germany

1998 "Modulation of the mechanism of fast outer hair cell motility by sugars in guinea-pigs"/ Physiological Society Meeting (Abstract), Prague, Czech republic

2008 "Developmental acquisition of sensory transduction in sensory cells of the mouse cochlea"/ Invited Seminar, Ear Institute, University College London, UK.

2016 "Molecular Pathophysiology & Gene Therapy in a Mouse Model of Usher Syndrome, Type IC"/ Molecular Biology of Hearing and Deafness (Abstract)

Welcome Genome Center, Hinxton, UK.

2016 "Gene therapy restores auditory and vestibular function in a mouse model of Usher syndrome, type IC"/ Inner Ear Biology Meeting, Montpellier, France.

2017 "Gene therapy restores auditory and vestibular function in a mouse model of Usher syndrome, type IC"/ Bertarelli Symposium, Geneva, Switzerland

2017 "Gene therapy treatment of congenital deafness in a mouse model of Usher syndrome type IC"/ European Society of Gene and Cell Therapy, Berlin, Germany

2018 "Gene therapy of Usher syndrome type IC"/ New horizon in vision and hearing research", Tubingen, Germany

2018 "Gene therapy of Usher syndrome type IC"/ Connection Conference, Moscow, Russia

2018 "Update on therapies of Usher syndrome"/ Usher syndrome symposium, Mainz, Germany

2019 "Gene therapy of Usher syndrome type IC"/ Inner Ear Biology meeting, Padova, Italy

2021 "Therapeutic Perspectives in the treatment of hearing loss and balance disorders in Usher syndrome patients". Usher Syndrome International Scientific Research Conference, Usher 1F Collaborative, Virtual presentation.

2021 "Antisense oligonucleotides or gene replacement therapy? Lessons learned from USH1C preclinical studies." International Usher Info Scientific Symposium, Fondation pour l'Audition.

Oral presentations from selected abstract (International)

1993 Chabbert C., **Géléoc G.S.G.***, Lehouelleur J. and Sans A. Calcium influx through mechanotransduction channels in isolated type I vestibular hair cells. (1993), *30th Inner Ear Biology Workshop- Budapest, Hungary*

1998 **Géléoc G.S.G.*** and Ashmore J.F. Sugar transport in isolated outer hair cells of the guinea-pig cochlea. (1998) *Proceedings of the Physiological Society*, 506P, 131P.- Prague, Czech Republic

2013 **Géléoc G. S. G.***, Pan B., Asai Y., Lentz J. and Askew C. Mechanotransduction Defects in Sensory Hair Cells of USH1C Knock-In Mice. (2013) *Molecular Biology of Hearing and Deafness Meeting. La Jolla, California*

2014 Askew C., Heman-Ackah S., Asai Y., Pan B., Lentz J. and **Géléoc G.S.G**.* Gene augmentation therapy to treat Usher Syndrome Type IC. (2014) *International Symposium on Usher Syndrome.* Boston, MA

2016 **Géléoc. G.S.G.***, Pan B., Haman-Ackah S., Galvin A., Indzhykulian A., Askew C., Lentz J., Vandenberghe L., Holt J.R. and Asai Y. Gene therapy restores auditory and vestibular function in a mouse model of Usher syndrome, type IC. (2016) *Molecular Biology of Hearing and Deafness (UK)*

Report of Teaching and Education Innovations

2012	Role: Session leader (Formal presentation followed by a discussion) on "***Responsible authorship and publications***". Program implemented by: Children's Hospital Program in Responsible Research (CHPRR).	Audience: ~ 50 Trainee, fellow, scholar, or participant receiving support from any NIH training grant (D, F or T), career development award (individual or institutional) (K), research education grant (R25), and/or dissertation research grant (R36), as well as students and postdoctoral fellows on any National Science Foundation (NSF) awards.
2012-2017	Role: Session leader (Formal presentation followed by a discussion) on "***Collaborative Research***". Program implemented by: Children's Hospital Program in Responsible Research (CHPRR).	
2013-2014	Role: Program coordinator and PI on NIH conference grant *International Symposium for Usher Syndrome*	The International Usher Syndrome Symposium was held in Boston, Massachusetts from July 9-12, 2014. This conference was the fourth of its kind and was meant to convene scientists whose research and interests are closely related to Usher Syndrome, with families and patients who suffer from the disease.
2017-2018	Role: Program coordinator and PI on NIH conference grant. *International Symposium for Usher Syndrome*	The second International Usher Syndrome Symposium took place in Mainz, Germany from July 19-21, 2018.
2017	Role: Workshop leader: Presentation & discussion "***Work/Life Balance in Academia***" ARO-SPARO program	Audience: ~ 30 Trainees
2018	Role: Session leader (Formal presentation followed by a discussion) on "***Responsible authorship and publications***". Program implemented by: Children's Hospital Program in Responsible Research (CHPRR).	Audience: ~ 50 Trainees, fellow, scholar, or participant receiving support from any NIH training grant (D, F or T), career development award (individual or institutional) (K), research education grant (R25), and/or dissertation research grant (R36), as well as students and postdoctoral fellows on any National Science Foundation (NSF) awards.
2018	Role: Discussion leader Gordon Research Conference: Function, Dysfunction and Restoration of the Auditory System.	This conference was meant for PIs and trainees and encompassed the fundamental mechanisms of auditory function, the etiology and pathology of inherited and acquired auditory dysfunction and innovative new approaches to restore auditory function.

2018	Role: Workshop faculty member The HEar@JAX Workshop *Modeling Hearing and Balance Disorders in Mice*	This is a pioneering training event that highlights the strengths of using the mouse as a model system to understand processes involved in human hearing, balance and deafness.
2018 & 2019	Role: Session leader (Formal presentation followed by a discussion) on "***Collaborative research***". Program implemented by: Children's Hospital Program in Responsible Research (CHPRR).	Audience: ~ 50 Trainee, fellow, scholar, or participant receiving support from any NIH training grant (D, F or T), career development award (individual or institutional) (K), research education grant (R25), and/or dissertation research grant (R36), as well as students and postdoctoral fellows on any National Science Foundation (NSF) awards.
2020	Coming to Our Senses: Vestibular Research- From Molecules to Systems- Commonalities and Differences with the Auditory System **Chairs: Gwenaelle S. Geleoc & Hong Zhu**	ARO- Symposium organizer

Report of Technological and Other Scientific Innovations

2017-06-15 WO2017100791A1 Application
Former: 2015-12-11 WO2017100791A1/ 2016-12-12 PCT/US2016/066225
Inventor: Konstantina Stankovic, Luk H. Vandenberghe, Jeffrey Holt, Gwenaelle Geleoc
Original Assignee: Massachusetts Eye & Ear Infirmary
The Children's Medical Center Corporation

2018-02-06 PCT Patent Application No.: PCT/US18/17104
Inventors: Jeffrey Holt, Gwenaelle Geleoc and Yukako Asai
Assignee: Children's Medical Center Corporation
"MATERIALS AND METHODS FOR DELIVERING NUCLEIC ACIDS TO COCHLEAR AND VESTIBULAR CELLS"

Report of Education of Patients and Service to the Community

No activities below were sponsored by outside entities

2004	Guest Lecturer, Montessori School of Anderson	1, 1hr special lecture
2008 – 2009	UVA Mini Med School Program	2 hours lab tour and demo
2014	Radio Interview- Radio Canada	20 min
2017-	USH Talks- Web based presentation Gene Therapy for Usher Syndrome Type IC	
2017-	Skype Talk to a Connection Conference for Usher patients and families in Russia	

Report of Scholarship

Peer-Reviewed Scholarship in print or other media

Research investigations

1. C. Chabbert, **G.S.G. Géléoc**, J. Lehouelleur and A. Sans (1994) Intracellular calcium variations evoked by mechanical stimulation of mammalian isolated vestibular hair cells. *European Journal of Physiology*, *Pflüger Arch.* 427: 162-168
2. G.W.T. Lennan, **G.S.G. Géléoc** and C.J. Kros (1996) Displacement sensitivity of mammalian vestibular hair cells. *Annals of New York Academy of Sciences*, New directions in vestibular research, 781: 650-652
3. **G.S.G. Géléoc**, G.W.T. Lennan, G.P. Richardson, and C.J. Kros (1997) A quantitative comparison of mechano-electrical transduction in vestibular and auditory hair cells of neonatal mice. *Proceedings of the Royal Society London*, 264: 611–621
4. **G.S.G. Géléoc**, S.O. Casalotti, A. Forge and J.F. Ashmore (1999) A sugar transporter as a candidate for the outer hair cell motor. *Nature neuroscience*, 8: 713-719
5. W. Marcotti, **G.S.G. Géléoc**, G.W.T. Lennan and C.J. Kros (1999) Transient expression of an inwardly rectifying potassium conductance in developing inner and outer hair cells along the mouse cochlea. *European Journal of Physiology, Pflüger Arch*, 439: 113-122
6. J.F.Ashmore, **G.S.G. Géléoc**, L. Harbott (2000) Molecular mechanisms of sound amplification in the mammalian cochlea. *Proceeding of the National Academy of Sciences USA,* 97: 11759-11764
7. **G.S.G. Géléoc** and J.R. Holt (2003) Developmental acquisition of sensory transduction in hair cells of the mouse inner ear. *Nature Neuroscience,* 6 :1019-1020
8. D.P Corey*, J. Garcia-Anoveros*, J.R. Holt*, K.Y. Kwan*, S.Y. Lin*, M.A. Vollrath*, A. Amalfitano, E. Cheung., B.H. Derfler, A. Duggan, , **G.S.G. Géléoc**, P. Gray, M.P. Hoffman, N. Hopkins, H.L. Rehm, D. Tamasauskas, and D.S. Zhang. (2004) TRPA1 is a candidate for the mechanosensitive transduction channel of vertebrate hair cells. *Nature*, 432(7018):723-730
9. **G.S.G. Géléoc**, J.R. Risner and J.R. Holt (2004) Developmental acquisition of voltage dependent conductances and sensory signaling in hair cells of the embryonic mouse inner ear. *J Neuroscience*, 24:11148-11159. Cover picture of the 2004 Dec 08 Journal issue.
10. M. Senften, M. Schwander, P. Kazmierczak, C. Lillo, J-B. Shin, T. Hasson, **G.S.G. Géléoc**, P.G. Gillespie, D. Williams, J.R. Holt and U. Müller (2006). Physical and functional interaction between protocadherin 15 and myosin VIIa in mechanosensory hair cells. *J Neuroscience*, 26:2060-71
11. K. Oshima, E. Corrales, C. Grimm, P. Senn, R. Martinez Monedero, **G.S.G. Géléoc**, A. Edge, J.R. Holt and S. Heller (2007) Differential distribution of stem cells in the auditory and vestibular organs of the inner ear. *J Assoc Res Otolaryngol.* 1:18-31 PMID: 17171473 PMCID: PMC2538418
12. J.R. Holt, E. Stauffer, D. Abraham and **G.S.G. Géléoc** (2007) Dominant-negative inhibition of M-like potassium conductances in hair cells of the mouse inner ear. *J Neuroscience*, 27:8940-8951. Cover picture of the 2007 Dec 19 Journal issue. PMID: 17699675 PMCID: PMC2647843
13. A. Lelli, Y. Asai, A. Forge, J.R. Holt and **G.S.G. Géléoc** (2009) Tonotopic gradient in the developmental acquisition of sensory transduction in outer hair cells of the mouse cochlea. *J Neurophysiology*, 101: 2961-2973. PMID: 19339464 PMCID: PMC2694104
14. C. Grimsley-Myers, C. Sipe**, G.S.G. Géléoc** and Xiaowei Lu (2009) The small GTPase Rac1 regulates auditory hair cell morphogenesis. *J Neuroscience*, 29:15859-15869.
15. Y. Asai, J. R. Holt and **G.S.G. Géléoc** (2010) A quantitative analysis of the spatiotemporal pattern of transient receptor potential gene expression in the developing mouse cochlea. *J Assoc Res Otolaryngol* 11(1):27-37- Epub 2009 Oct16 PMID: 19834762 PMCID: PMC2820207
16. G. Horwitz, **G.S.G. Géléoc**, A. Lelli and J.R. Holt (2010) HCN channels are not required for

mechanotransduction in sensory hair cells of the mouse inner ear. *PLoS ONE* 5(1): e8627. doi:10.1371/journal.pone.000862731) PMID: 20062532 PMCID: PMC2797612

17. K.A. Steigelman, A.Lelli, X.D.Wu, J.Gao, S.Lin, K.Piontek, C.Wodarczyk, A.Boletta, H.Kim, F.Qian, G.Germino, **G.S.G.Géléoc**, J.R.Holt and J. Zuo (2011). Polycystine-1 is required for stereocilia structure and maintenance but not for mechanotransduction in inner ear hair cells. *J Neuroscience*, 31:12241-12250. PMID: 21865467 PMCID: PMC3164988

18. Y. Kawashima*, **G.S.G. Géléoc*,** K. Kurima*,V. Labay, A. Lelli, Y. Asai, T. Makishima,D.K. Wu, C. C. Della Santina, J.R. Holt, A. J. Griffith (2011). Mechanotransduction in mouse inner ear hair cells requires transmembrane channel–like genes. *J. Clinical Investigation*, 121(12):4796-809. doi: 10.1172/JCI60405. (*co-first authors); PMID: 22105175 PMCID: PMC3223072

19. B. Pan, **G.S.G. Géléoc**, Y. Asai, G.C. Horwitz, K. Kurima, K. Ishikawa, Y. Kawashima, A.J. Griffith, J.R. Holt (2013). TMC1 and TMC2 Are Components of the Mechanotransduction Channel in Hair Cells of the Mammalian Inner Ear. *Neuron.* 79(3):504-515. doi: 10.1016/j.neuron.2013.06.019. Epub 2013 Jul 18. PMID: 23871232 PMCID: PMC3827726

20. **G.S.G Géléoc** and J.R. Holt (2014) Sounds strategies for hearing restoration. *Science 344* (6184):1241062. doi: 10.1126/science.1241062. Review. PMID: 24812404 PMCID: PMC4148779

21. B. Pan, C. Askew, A. Galvin, S. Heman-Ackah, Y. Asai, A.A. Indzhykulian, F.M. Jodelka, M.L. Hastings, J.J. Lentz, L.H. Vandenberghe, J.R. Holt, and **G.S.G Géléoc** (2017) Gene Therapy Restores Auditory and Vestibular Function in a Mouse Model of Usher Syndrome Type 1c. *Nature Biotechnology* doi:10.1038/nbt.3801 Epub 2017 Feb 06. PMID: 28165476 PMCID: PMC5340578

22. M.L. Shannon, R.M. Fame, K.F. Chau, N. Dani, M.L. Calicchio, **G.S.G Géléoc**, H.G.W. Lidov, S .Alexandrescu, M.K. Lehtinen. (2018) Mice Expressing c-MYC in Neural Precursors Develop Choroid Plexus and Ciliary Body Tumors. *Am J Pathol*. pii: S0002-9440(17)31033-7. doi: 10.1016/j.ajpath.2018.02.009.

23. Y. Asai, B. Pan, C. Nist-Lund, A. Galvin, A.N. Lukashkin, V.A. Lukashkina, T. Chen, W. Zhou, H. Zhu, I.J. Russell, J.R. Holt & **G.S. G. Géléoc.** (2018) Transgenic Tmc2 expression preserves inner ear hair cells and vestibular function in mice lacking Tmc1. *Scientific Reports,* 8(1):12124. doi: 10.1038/s41598-018-28958-x. PMID: 30108254 PMCID: PMC6092434

24. H. Nakanishi, K. Kurima, B. Pan, P. Wangemann, T.S. Fitzgerald, **G.S.G. Géléoc**, J.R. Holt, A.J. Griffith. (2018) Tmc2 expression partially restores auditory function in a mouse model of DFNB7/B11 deafness caused by loss of Tmc1 function. *Scientific Reports* 8(1):12125. doi: 10.1038/s41598-018-29709-8. PMID: 30108230 PMCID: PMC6092339

25. M.R. Avenarius, J-Y. Jung, C. Askew, S.M. Jones, K.L. Hunker, H. Azaiez, A.U. Rehman, M. Schraders, H. Najmabadi, H. Kremer, R.J.H. Smith, **G.S.G. Géléoc**, D.F. Dolan, Y. Raphael, and D.C. Kohrman (2018). Grxcr2 is required for stereocilia morphogenesis in the cochlea. *Plos One* 13(8):e0201713. doi: 10.1371 PMID: 30157177 PMCID: PMC611452

26. C.A. Nist-Lund, B. Pan, A. Patterson, Y. Asai, T. Chen, W. Zhou, H. Zhu, S. Romero, J. Resnik, D.B. Polley, **G.S.G Géléoc**, J.R. Holt. (2019) Improved TMC1 gene therapy restores hearing and balance in mice with genetic inner ear disorders. *Nat Commun*. 10(1):236. doi: 10.1038/s41467-018-08264-w. PMID: 30670701 PMCID: PMC6342993

27. B. György, C. Nist-Lund, B. Pan, Y. Asai, K. D. Karavitaki, B. Kleinstiver, S.P. Garcia, M.P. Zaborowski, P. Solanes, S. Spataro, B. Schneider, J. K. Joung, **G.S.G. Géléoc**, J.R. Holt*, D.P. Corey* (2019) Allele-specific gene editing prevents deafness in a model of dominant progressive hearing loss. *Nat Med.* 25(7):1123-1130. doi: 10.1038/s41591-019-0500-9. Epub 2019 Jul 3. PMID 31270503

28. **G.S.G Géléoc**, A. EL-Amraoui. (2020) Disease mechanisms and gene therapy for Usher syndrome. *Hearing Res.* 394:107932. doi: 10.1016/j.heares.2020.107932. Epub 2020 Mar 4. PMID: 32199721

29. J. J. Lentz, B. Pan, A. Ponnath, C.M. Tran, C. Nist-Lund, A. Galvin, H. Goldberg, K.N. Robillard, F.M. Jodelka, H.E. Farris, J. Huang, T. Chen, H. Zhu, W. Zhou, F. Rigo, M.L. Hastings, **G.S.G. Géléoc.** (2020) Direct delivery of antisense oligonucleotides to the middle and inner ear improves hearing and balance in Usher mice. *Mol. Ther.* 2020 28(12):2662-2676. doi: 10.1016/j.ymthe.2020.08.002. Epub 2020 Aug 5. PMID: 32818431

30. J. Lee, C. Yergeau, K. Kawai, N. Braverman, **G.S.G. Géléoc** (2021) A Retrospective Study of Hearing Loss in Patients Diagnosed with Peroxisome Biogenesis Disorders in the Zellweger Spectrum. *Ear and Hearing* doi: 10.1097/AUD.0000000000001126. PMID: 34534157

31. J. Lee, J.R. Holt and **G.S.G. Géléoc** (2021) Sensory Transduction is Required for Normal Development and Maintenance of Cochlear Inner Hair Cell Synapses. *Elife* 10:e69433. doi: 10.7554/eLife.69433. PMID: 34734805- PMCID: PMC8598158

32. S.A. Mauriac, T. Peineau, A. Zuberi, C. Lutz and **G.S.G. Géléoc** (2022) Loss of Pex1 in Inner Ear Hair Cells Contributes to Cochlear Synaptopathy and Hearing Loss. Cells. 2022 Dec 9;11(24):3982. doi: 10.3390/cells11243982. PMCID: PMC9777190; PMID: 36552747

Non-peer reviewed scholarship in print or other media

Reviews, chapters

1. J.F Ashmore and **G.S.G. Géléoc** (1999) Hearing in the fast lane. *Current biology*, 9: R572-R574

2. **G.S.G. Géléoc** and J.R. Holt (2003) Auditory amplification: outer hair cells pres the issue. *Trends in Neuroscience,* 26:115-117

3. J.R. Holt and **G.S.G. Géléoc** (2009) Hair cells: Sensory transduction. Encyclopedia of Neuroscience pp1015-1020.

4. **G.S.G. Géléoc** (2012) TMC1 and TMC2: Key partners in hearing and vestibular function. [TMC1 et TMC2 : Partenaires essentiels dans l'audition et l'équilibre.] Med Sci (Paris), 28 (8-9): 701-703.

5. B.W. Kesser, J.Burns, B. Thiede and **G.S.G. Géléoc** (2013) Experimental Approaches to Auditory Neuroscience. "Otology" Thieme Medical Publishers, Inc. Kirtane, Milind V. et al.: 2013 Otology and Neurotology- DOI: 10.1055/b-0034-8056

6. J.R. Holt and **G.S.G. Géléoc** (2017) Hair cells: Sensory transduction. Encyclopedia of Neuroscience Online Publication Date: May 2017 DOI: 10.1093/acrefore/9780190264086.013.8

7. J.R. Holt, **G.S.G Géléoc** (2019) Split otoferlins reunited. *EMBO Mol Med*. 2019 Jan;11(1). pii: e9995. doi: 10.15252/emmm.201809995. PMID: 30573543; PMCID: PMC6328905.

8. S.A. Mauriac and **G.S.G Géléoc** (2021) A hop, skip and a jump to evade USH2A deaf-blindness mutations. *Molecular Therapy, Commentary,* Mol Ther. 2021 Aug 4;29(8):2391-2393. doi: 0.1016/j.ymthe.2021.07.008. Epub 2021 Jul 23. PMID: 34297918

9. Indzhykulian, S. Johnson and **G.S.G Géléoc** (2022) "Electrophysiological recordings of voltage dependent and mechanosensitive currents in sensory hair cell of the auditory and vestibular organs of

the mouse." *Book Chapter,* Neuromethods, Volume 176, pp 221-264; DOI: 10.1007/978-1-0716-2022-9_10.; Developmental, Physiological, and Functional Neurobiology of the Inner Ear; Editor: Andrew K. Groves

10. S.G. Sadeghi, **G.S.G. Géléoc** (2022) Editorial: Commonalities and Differences in Vestibular and Auditory Pathways., Front Neurosci. 2022 Mar 23;16:876798. doi: 10.3389/fnins.2022.876798. eCollection 2022; PMID: 35401079 PMCID: PMC8984178

Thesis

Diplôme de Doctorat label Européen avec mention très honorable et les félicitations du jury
PhD degree Summa Cum Laude
G.S.G. Géléoc (1996) Mécano-transduction des cellules ciliées de l'oreille interne chez le mammifère : approche électrophysiologique et microfluorimétrique. [Mechanotransduction of mammalian inner ear sensory hair cells: Electrophysiological and microfluorimetric study.]
Université de Montpellier II, Sciences et Techniques du Languedoc, Montpellier, France

Abstracts, Poster Presentations and Exhibits Presented at Professional Meetings (From the last three years- Unpublished work)

1. Zhu H., Chen T, Huang J., Xu Y., Ou Y., Tu Y., Sandlin D., Arteaga A., **Géléoc G.S.G**., Holt J.R., Zhou W. (2019) Single Unit Recording Studies of Vestibular Afferents in Mice Lacking Tmc1 and Tmc2. *42th Association for Research in Otolaryngology Meeting, Baltimore.*

2. Goldberg H., Asai Y., Pan B., Isgrig K., Yang J., Chien W.W., **Géléoc G.S.G.** (2019) AAV Mediated Gene Therapy Restores Partial Auditory Sensitivity in Mouse Models of Autosomal Recessive Non-Syndromic Deafness DFNB31 and Usher Syndrome type IID. *42th Association for Research in Otolaryngology Meeting, Baltimore*

3. Isgrig K., Zhu J., Cheng C., Goldberg H., Belyantseva I.A., Friedman T.B., Cunningham L., **Géléoc G.S.G.,** Chien W.W. (2019) Effect of Increased Outer Hair Cell Infection and Whirlin Long Isoform in Whirlin Mutant Mice. *42th Association for Research in Otolaryngology Meeting, Baltimore*

4. Lentz J.J., Nist-Lund C., Goldberg H., Pan B. Jodelka F.M., Huang J., Chen T., Rigo F., Zhu H., Zhou W., Hastings M.L., and **Géléoc G.S.G.*** (2019) Comparative study: Gene replacement versus antisense oligonucleotide therapy to restore hearing and balance in a mouse model of Ush1c. *International Symposium on Auditory and Audiological Research, Nyborg, Danemark.*

5. Nist-Lund C., Pan B., Hastings M.L., Rigo F., Lentz J.J., and **Géléoc G.S.G.** (2019) Exploring gene replacement or antisense therapy to restore hearing and balance in a mouse model of Usher syndrome type IC. *Hair cell regeneration and Hearing Restoration, New York Academy of Sciences. NY.*

6. Du W., **Géléoc G.S.G**, Zhang A., Chen T.; Huang J.; Zhu H.; Zhou W., Zheng Q (2020) PS 419. Long-Term Expression Stability of Anc80L65- Containing Virus Infected Hair Cells in the USH1C Mouse Model. *43rd Association for Research in Otolaryngology Meeting, San Jose*

7. Goldberg H., Asai Y., Pan B., Isgrig K., Chien W., Yang J., **Géléoc G.S.G.** (2020) PS 681- AAV mediated gene therapy restores auditory sensitivity in mouse models of autosomal recessive non syndromic deafness DFNB31 and Usher syndrome type IID. *43rd Association for Research in Otolaryngology Meeting, San Jose*

8. Lee J., **Géléoc G.S.G.,** Holt J. (2021) Exploring the Roles of TMC1 and TMC2 in Hair Cells of the Vestibular End Organs. *44rd Association for Research in Otolaryngology, Virtual Midwinter Meeting*

9. Omar Akil, Lei Xu, Susan Bolch, Frank M. Dyka, William N. Ruddick, Chiab Simpson, Carl Nist-Lund, W.Clay Smith, Gwenaelle **S.G. Géléoc**, Astra Dinculescu (2022) A novel epitope-tagged Clarin-1 knock-in mouse displays congenital deafness. *45th Association for Research in Otolaryngology, Virtual Midwinter Meeting*

10. Stephanie Rouse, Astra Dinculescu and Gwenaelle **S.G. Géléoc** (2022) A novel mouse model of USH3 displays auditory and balance deficits. *45th Association for Research in Otolaryngology, Virtual Midwinter Meeting*

11. Tianwen Chen, Jun Huang, Peyton Overstreet, Youguo Xu, Chunming Zhang, Yue Yu, David Sandlin, Alberto A. Arteaga, Gwenaelle **S.G. Géléoc**, Jeffrey R. Holt, Hong Zhu, Wu Zhou (2022). Pupillary responses of mice lacking Tmc1 and Tmc2. *45th Association for Research in Otolaryngology, Virtual Midwinter Meeting*

12. Tianwen Chen, John Lee, Jun Huang, Youguo Xu, Tracy Rappai, Michael Zhang, Jennifer Lentz, Gwenaelle **S.G Géléoc**, Hong Zhu, Wu Zhou. (2022) Progression of vestibular dysfunction in Ush1c mice. 45th *Association for Research in Otolaryngology, Virtual Midwinter Meeting*

13. Evan Ratzan, John Lee, Gwenaelle **S.G. Géléoc**, Jeffrey Holt (2023) AAV Delivery of Tmc1 Rescues Vestibular Function in Tmc1 KO Mice. *46th Association for Research in Otolaryngology Midwinter Meeting, Orlando, Florida.*

14. Tianwen Chen, Caroline Sit, Jun Huang, Zelma Iriarte-Oporto, Youguo Xu, Gwenaelle **S.G. Géléoc**, Jeffrey Holt, Hong Zhu, Wu Zhou (2023). Roles of Tmc1 and Tmc2 Channels in Vestibular Function. *46th Association for Research in Otolaryngology Midwinter Meeting, Orlando, Florida.*

Narrative Report

A significant focus of my research has been on the characterization of sensory transduction in the inner ear using mouse models. During my PhD, I developed a first-of-its-kind preparation to characterize sensory transduction in the vestibular system and performed a comparative study with auditory hair cells (Geleoc et al., Proc. Biol. Soc. 1997). As an independent investigator at the University of Virginia, I expanded this work and characterized the developmental acquisition of sensory transduction and voltage-sensitive conductances in mouse vestibular hair cells, and the tonotopic gradient in the acquisition of sensory transduction in the mouse auditory organ (Géléoc and Holt, Nature Neurosci. 2003; Géléoc et al., J. Neurosci. 2004; Lelli et al., J. Neurophysiol. 2009). Based on the timelines identified in those studies, I was able to determine the role of molecular players such as KCNQ4 (Holt et al., J. Neurosci. 2007) and the developmental expression patterns of several genes necessary for normal hair cell transduction, including deafness genes, cadherin-23, protocadherin-15, myosin-7a, myosin-1c and myosin-15.

To identify the elusive sensory transduction channel, a major focus in the hearing research field for over 3 decades, I performed spatiotemporal quantitative analysis of transient receptor potential (Trp) (Asai et al. JARO, 2010) and transmembrane channel-like (Tmc) gene expression (Kawashima, Géléoc et al., JCI 2011). Interestingly, analysis of Tmc1&2 mRNA expression revealed tonotopic (base to apex) expression that coincided with the developmental milestones (Lelli et al., J. Neurophysiol. 2009). These results motivated further studies in my lab that revealed molecular, cellular, and biophysical evidence that demonstrated the role of TMC1 and TMC2 proteins as components of hair cell transduction channels (Kawashima et al., JCI 2011; Pan et al., Neuron 2013;). Further studies in my lab also revealed the important role of TMC2 in the balance organs (Pan et al., Sci Rep. 2018; Nakanishi et al., Sci Rep, 2018).

My expertise and knowledge have placed me in position to investigate other hair cell genes associated with hearing loss and to develop novel therapies to protect or restore auditory function. To explore the broad applicability of inner ear gene therapy, my lab develops tools to tackle genetic deafness that results from mutations in Usher genes, which are responsible for combined hearing impairment and blindness. Our first study focused on Usher syndrome type IC using a mouse model for a known French-Acadian USH1C mutation. Early postnatal injection of synthetic adeno-associated viral vectors carrying the correct Ush1c gene lead to unprecedented recovery of hearing and balance to near wild type levels (Pan et al., Nature Biotech. 2017). I also obtained similar recovery using short antisense oligonucleotides targeting the same USH1C mutation (Lentz et al., Mol Therapy 2020). The data suggest that biological therapies to treat deafness may be suitable for translation to humans with genetic inner ear disorders. Indeed, more recent work from my lab shows recovery of function after Tmc1 gene therapy with gene replacement or gene editing (Gyorgy et al., Nature Med. 2019; Nist-Lund et al., Nature Comms. 2019).

My fundamental and translational research work has brough international interest. As such, I have developed important collaborations with scientists worldwide and relationships with several family foundations, in part through the organization of two consecutive International Symposia on Usher syndrome, one in Boston, in 2014, and one in Germany, in 2019. I was co-principal investigator (2014) and principal investigator (2019) on two R13 NIH conference grants that supported these events.

My significant supporting activity is in teaching. I have been involved with teaching undergraduate students at the University of Montpellier (France), graduate students in the Neuroscience Graduate Program at the University of Virginia, summer students at the Marine Biology Laboratories, Biology of the Inner Ear and at the NIH, Ear Essentials courses and graduate students and postdocs at Harvard Medical School. In 2019, I took on the role of Director of Student Affairs for the Harvard, Speech and Hearing, Bioscience and Technology (SHBT) graduate program. One of my tasks has been to assemble and maintain a handbook for the program and prepare new guidelines clarifying policies and expectations for Ph.D. students. I have worked closely with the students (currently 43 of them) to monitor academic progress and communicate individually with each student on a regular basis to ensure their progress and well-being, something that has

been even more important during the pandemic. I have enjoyed my interaction with the students and feel fortunate to be able to teach and serve as a mentor to all the students. To my delight, in spring 2021, I was selected to serve as the co-director of the SHBT graduate program. The SHBT program is unique in that it encompasses a wide range of topics and includes faculty working at various locations in and around Boston. I intend to work with students and faculty to develop a better sense of community for the program, include new efforts towards equity, diversity and inclusion as well as mental health support and importantly, provide a cohesive and competitive curriculum designed to train future scientists and leaders in the field of speech and hearing sciences.