



## **Modeling Hearing and Balance Disorders in Mice: The Hear@JAX Workshop**

**September 23-28, 2018**

### **EVENT SCHEDULE**

#### **Sunday, September 23, 2018**

- 5:00 pm **Registration at Highseas Conference Center**
- 5:00 pm **Organizer meeting**
- 6:00 pm **Dinner at Highseas**
- 7:00 pm **Welcome and introductions**  
Karen Avraham, Basile Tarchini, Matt Kelley, Amanda Lauer, Mike Bowl, Guy Richardson
- 7:30 pm **The deaf patient as seen by an ENT-geneticist**  
Ronna Hertzano
- 8:00 pm **The other side: The culture of deafness**  
Karen Avraham
- 8:30-9:15 pm **The history of inner ear mouse mutants and their present state**  
Karen Steel

#### **Monday, September 24**

- 7:30 am **Breakfast and late registration** (Highseas 1<sup>st</sup> floor)  
Highseas 3<sup>rd</sup> floor
- 8:30 am **Introduction to sound and sound perception**  
Amanda Laurer
- 9:00 am **A genetics primer**  
Karen Avraham
- 9:30 am **Mouse genetics, nomenclature, and MGI**  
Leona Gagnon
- 10:00 am **JAX KOMP2: Mouse phenotypes & resources for ear research**  
Steve Murray
- 10:30 am **Coffee break**
- 10:45 am **General introduction to inner ear development**  
Andy Groves
- 11:30 am **Hair cell development**  
Matt Kelley
- 12:15 pm **Lunch** (Highseas 1<sup>st</sup> floor)

- 1:00pm **Transportation to GRB Training Laboratory** (Meet in Highseas 1<sup>st</sup> floor lobby)
- 1:15 pm **Biomethods Training** (GRB 3245)  
Kristin Cough
- 2:30 pm **Laboratory Session: Dissection and culturing of the inner ear, tutorial on ABR/DPOAE**  
Matt Kelley, Guy Richardson, Basile Tarchini, Ronna Hertzano  
*Participants will learn how to dissect the cochlea and the vestibular organs from neonate wild-type mice. Some tissues will be fixed and processed for whole-mount immunofluorescence using various typical antibodies to label hair cells, supporting cells and neuron projections. Other tissues will be placed in culture. A tutorial will be presented on Auditory brainstem response (ABR) and Distortion product otoacoustic emission recordings (DPOAE).*
- 6:00 pm **Transportation to Highseas**
- 6:30 pm **Dinner** (Highseas 1<sup>st</sup> floor)
- 7:30-8:15 pm **Planar cell polarity in the inner ear**  
Basile Tarchini

## **Tuesday, September 25**

- 7:30 am **Breakfast** (Highseas 1<sup>st</sup> floor)
- 8:30 am **Deafness genes in humans**  
Uli Mueller
- 9:00 am **An epigenetics primer**  
Neil Segil
- 9:30 am **Transcriptome analysis & inner ear cell sorting**  
Ronna Hertzano
- 10:15 am **Break**
- 10:30 am **Single cell transcriptomics in the inner ear**  
Matt Kelley
- 11:15 am **Epigenetics of the auditory system**  
Neil Segil
- 12:00 pm **Lunch** (Highseas 1<sup>st</sup> floor)
- 12:45 pm **Transportation to GRB Training Laboratory** (Meet in Highseas 1<sup>st</sup> floor lobby)
- 1:00 pm **Microscopy in inner ear research**  
Uri Manor
- 1:45 pm **Laboratory Session: Dissection and culturing of the inner ear (continued), demonstration of paint fill and MET channel function**  
Matt Kelley, Guy Richardson, Basile Tarchini, Ronna Hertzano  
*Cochlear and vestibular wild-type samples collected by participants the previous day will be mounted on slides and observed using confocal microscopy in small groups. Mutant samples prepared in advance will enable participants to observe phenotypes leading to congenital hearing loss and circling behavior using confocal and scanning electron microscopy. Cultures set up the previous day will be used to perform dye uptake and show aminoglycoside toxicity in hair cells, and inner ear paint-fill will be demonstrated.*

6:00 pm **Transportation to Highseas**  
6:30 pm **Dinner** (Highseas 1<sup>st</sup> floor)  
7:15 pm **Free evening**

### **Wednesday, September 26**

7:30 am **Breakfast** (Highseas 1<sup>st</sup> floor)  
8:30 am Hair cells and **mechanotransduction**  
Guy Richardson  
9:15 am **Afferent & efferent innervation of the inner ear**  
Paul Fuchs  
10:00 am **Break**  
10:30 am **Central auditory system & effects of hearing loss on the brainstem**  
Amanda Lauer  
11:15 am **The vestibular system**  
Jennifer Stone  
12:00 pm **Hair cell damage/protection/hidden hearing loss**  
Uri Manor  
12:30 pm **Lunch** (Highseas 1<sup>st</sup> floor) and **free afternoon**  
1:00 pm **Optional Microscopy session** (participant's samples and mutant samples prepared in advance), Basile Tarchini, Uri Manor  
6:00 pm **Dinner** (Highseas 1<sup>st</sup> floor)  
7:00 pm **gEAR workshop** (bring your laptop)  
Ronna Hertzano  
8:00-8:45 pm **The state of the art for mice as models for human deafness** (Highseas 3<sup>rd</sup> floor)  
Uli Mueller

### **Thursday, September 27**

7:30 am **Breakfast** (Highseas 1<sup>st</sup> floor)  
8:30 am **CRISPR-Cas genome editing**  
Michael Wiles  
9:15 am **CRISPR-Cas applied to age-related hearing loss**  
Michael Bowl  
10:00 am Coffee  
10:15 am **Viral gene therapy**  
Gwen Geleoc  
11:00 am **Regeneration**  
Jennifer Stone  
11:45 am **Organoids**  
Karl Koehler  
12:30 pm **Lunch** (Highseas 1<sup>st</sup> floor)  
1:15 pm **Transportation to GRB Training Laboratory** (Meet in Highseas 1<sup>st</sup> floor lobby)  
1:30 pm **Laboratory session: Microscopy, demonstration of injection for gene therapy, tutorial on organoids**  
Karl Koehler, Gwen Geleoc (with help from Richardson, Tarchini, Hertzano, Kelley  
Observation of wild-type and mutant samples with confocal and scanning electron microscopy will be continued in small groups. Posterior canal injection will be

demonstrated as a powerful method for gene therapy in the inner ear. A tutorial will be delivered on the potential of stem-cell derived inner ear organoids.

6:00 pm **Transportation to Highseas**  
6:30 pm **Lobster Dinner** (Highseas 1<sup>st</sup> floor)

**Friday, September 28, 2018**

7:30 am **Breakfast** (Highseas 1<sup>st</sup> floor)  
7:30 am **Organizers' meeting**  
9:00 am **Room check-out and departures**

Course Organizers

Karen Avraham, Chair  
Mike Bowl  
Matt Kelley  
Amanda Lauer  
Guy Richardson  
Basile Tarchini

Alison Kieffer, JAX  
Melissa Johnson, JAX

Faculty

Ronna Hertzano, MD, PhD, University of Maryland School of Medicine  
Leona Gagnon, MS, The Jackson Laboratory  
Steve Murray, PhD, The Jackson Laboratory  
Karen Steel, PhD, King's College, London  
Michael Bowl, PhD, Medical Research Council, Harwell  
Matt Kelley, PhD, NIDCD/NIH  
Kristin Cough, BS, The Jackson Laboratory  
Guy Richardson, PhD, University of Sussex  
Basile Tarchini PhD., The Jackson Laboratory  
Uli Mueller, PhD, Johns Hopkins  
Neil Segil, PhD, University of Southern California  
Uri Manor, PhD, Salk Institute for Biological Studies  
Andy Groves, PhD, Baylor College of Medicine  
Jennifer Stone, PhD, University of Washington  
Paul Fuchs, PhD, Johns Hopkins University School of Medicine  
Amanda Lauer, Ph.D., Johns Hopkins University School of Medicine  
Michael Wiles, PhD, The Jackson Laboratory  
Gwen Geleoc, PhD, Boston Children's Hospital  
Karl Koehler, PhD, Indiana University School of Medicine