

### (217

# DIVISION OF NEPHROLOGY

# Mission

The mission of the Division of Nephrology is:

- To care for children with renal and urinary tract disorders, directly and/or in consultation with other physicians
- To provide high-quality care, using the highest moral, ethical, and professional conduct
- To promote knowledge of renal topics to primary physicians, residents, medical students, advanced practice providers, and nurses through direct teaching and mentoring
- To extend knowledge of pathophysiology of renal disease through basic, clinical, and translational research
- To train pediatric nephrology fellows to become outstanding clinical nephrologists and to have the fellows engage in high-quality kidney/ urinary tract-based research projects

218

#### **FACULTY**

#### Carlton M. Bates, MD

Professor of Pediatrics Chief, Division of Nephrology Codirector, Pediatric Nephrology Fellowship Program Vice Chair of Basic Research

#### Michael Moritz, MD

Professor of Pediatrics Clinical Director, Division of Nephrology Medical Director, Pediatric Dialysis

#### Rannar Airik, PhD

Assistant Professor of Pediatrics

#### **Demetrius Ellis, MD**

Professor of Pediatrics

#### Dana Fuhrman, DO

Assistant Professor of Critical Care Medicine and Pediatrics

#### Jacqueline Ho, MD

Assistant Professor of Pediatrics Codirector, Pediatric Scientist Development Program Codirector, Pediatric Nephrology Fellowship Program

#### Yosuke Miyashita, MD, MPH

Assistant Professor of Pediatrics Assistant Director, Pediatric Nephrology Fellowship Program

#### Christina Nguyen, MD

Assistant Professor of Pediatrics Director, Pediatric Kidney Transplant Program

#### Sunder Sims-Lucas, PhD

Assistant Professor of Pediatrics

#### Agnieszka Swiatecka-Urban, MD

Associate Professor of Pediatrics

#### **OVERVIEW OF DIVISION**

he faculty consists of eight pediatric nephrologists and two PhD investigators. Among the nephrologists, Demetrius Ellis, Michael Moritz, Yosuke Miyashita, and Christina Nguyen are primarily clinically oriented, whereas Carlton Bates, Agnieszka Swiatecka-Urban, Jacqueline Ho, and Dana Fuhrman (dual appointment to Nephrology and Critical Care) are more focused on research, spanning clinical to basic discovery. Sunder Sims-Lucas, PhD, and Rannar Airik, PhD, are engaged in basic scientific research.

Major accomplishments in the division included new and ongoing research grants, including National Institutes of Heath (NIH) R01 awards (Bates, Ho, Swiatecka-Urban [co-investigator], and Nguyen [co-investigator]), an NIH R56 award (Swiatecka-Urban), NIH P30 pilot and feasibility sub-awards (Airik and Swiatecka-Urban), an NIH cooperative agreement sub-award (Swiatecka-Urban), an NIH U01 (Nguyen [co-investigator]), an NIH R00 Pathway to Independence Award (Airik), an NIH K01 award (Sims-Lucas), an NIH R03 award (Sims-Lucas), an NIH U24 award (Ho and Sims-Lucas [co-investigators]), an NIH GUDMAP award (Sims-Lucas [co-investigator]), an NIH T32 Pediatric Nephrology Fellowship training grant (Bates), an American Heart Association award (Swiatecka-Urban), University of Pittsburgh Vascular Medicine Institute pilot awards (Sims-Lucas), a University of Pittsburgh Research Advisory Committee awards (Airik and Sims-Lucas).

The division engaged in many clinical research programs, quality-improvement projects, and educational initiatives. In association with the Midwest Pediatric Nephrology Consortium, Miyashita led ongoing studies focused on diagnosis and follow-up of white-coat hypertension, which will soon lead to a practice guideline. The kidney transplant program, led by Nguyen, recently joined the Improving Renal Outcomes Collaborative (IROC) Network, which is devoted to eliminating the survival disparity between children with kidney diseases and the general population. In 2017, Sims-Lucas became the director of student research training for the Department of Pediatrics and led another highly successful summer undergraduate student research program with 45 participants (chosen from more than 400 applicants), including three high school students from the Pittsburgh City School District. The division hosted its fourth annual Nephrotic Syndrome Symposium on September 28, 2017 (led by Swiatecka-Urban), which featured local, national, and international experts in nephrotic syndrome research.

The division continues to recruit high-quality trainees into the fellowship program, including Melissa Anslow, Caitlin Peterson, and Cassandra Formeck. Paul Fadakar and Emily Joyce graduated from the program in June 2017 and joined the faculty in July 2017. The division successfully recruited two new fellows to the program to start in fiscal year 2018: Amy Lucier and Christine Crana.

#### **CLINICAL ACTIVITIES**

he pediatric nephrology division at Children's Hospital of Pittsburgh of UPMC is the sole provider of specialty nephrology care, including dialysis, for children in the Pittsburgh area and for most of the children within a 100-mile radius of the hospital. The pediatric nephrology program is among the top five largest pediatric kidney transplant programs in the country and has included patients from 18 states outside Pennsylvania. The division continues to see patients at the main hospital campus in Lawrenceville as well as two satellites (Wexford and Monroeville) and four outreach sites (Erie, Johnstown, Chippewa, and Sharon), with plans to add an outreach site in Wheeling, W.Va., in 2018.

The division had nearly 3,000 outpatient visits and saw more than 500 inpatients over the past fiscal year. The division offered clinical nutrition consultations for all its patients in the outreach locations via telemedicine, saving patients extra trips to Lawrenceville. The ambulatory blood pressure monitoring program, led by Miyashita, continued to expand. Nguyen and Moritz also performed renal ultrasound exams, saving patients extra trips to the radiology department. Under the direction of Moritz, the division began using low-density lipoprotein apheresis, a novel therapy to treat patients with steroid-resistant forms of nephrotic syndrome, with some striking successes.

#### RESEARCH AND OTHER SCHOLARLY ACTIVITIES

#### Carlton M. Bates, MD

#### **RESEARCH**

Carlton Bates conducts NIH-funded studies on the role of fibroblast growth factor receptors (FGFRs) and their signaling adapter proteins in the developing kidney and lower urinary tract using the mouse as a model. The studies are clinically relevant given that congenital abnormalities of the kidney and urinary tract are responsible for most cases of chronic kidney disease in children. Using various genetargeting approaches, the laboratory reveals how FGFRs and their adapters are critical for temporal patterning of multiple renal and bladder tissue lineages. They generate mouse models of pediatric structural kidney and lower urinary diseases, including obstructive nephropathy, renal aplasia/dysplasia/hypoplasia, vesicoureteral reflux and reflux nephropathy, progressive cystic kidney disease, atonic bladders, and functional bladder obstruction. The laboratory continues to elucidate the molecular pathways through which FGFRs pattern developing bladder urothelium and repair postnatal urothelium after injury. The laboratory recently discovered novel critical links between FGFR signaling and glucose metabolism in developing nephron progenitors. These approaches may lead to new biomarkers and therapeutic targets for congenital kidney and lower urinary tract disease.

#### **STUDY SECTIONS**

- Basil O'Conner Research Advisory Committee, March of Dimes
- Urologic and Hematologic Diseases D Subcommittee (reviewing training [T32, T35] and career development [K01, K08, K23, K24, K00, R25, R03] applications),

National Institute of Diabetes and Digestive and Kidney Diseases (NIDDK), NIH

#### ADVISORY COMMITTEE MEMBERSHIPS

- Council of the International Pediatric Nephrology Association
- Council of the Society for
  - Pediatric Research (Nephrology Section)
- Organizing Committee, 16th Congress of the International Pediatric Nephrology Association
- Training and Certification Committee, American Society of Pediatric Nephrology
- Internal Advisory Board, Center for Critical Care Nephrology, University of Pittsburgh School of Medicine

#### **EDITORSHIPS**

- Associate editor (review articles), Pediatric Nephrology
- Editorial Board, Journal of the American Society of Nephrology
- Editorial Board, Bladder

#### MAJOR LECTURESHIPS AND SEMINARS

 "Renal and Urogenital Development and Their Developmental Abnormalities," Developmental Renal



Carlton M. Bates, MD
Division Chief, Nephrology

- "From Tubes to Cysts: Novel Mechanisms Driving Pathology in Cystic Kidney Disease," 17th Congress of the International Pediatric Nephrology Association, Iguassu Falls, Brazil, September 2016
- "From Tubes to Cysts: Novel Mechanisms Driving Pathology in Cystic Kidney Disease," pediatric nephrology grand rounds, University of California, Los Angeles, October 2016
- "FGFR Signaling in Nephron Progenitors: Links to Metabolism and Cystogenesis," renal grand rounds, Cincinnati Children's Medical Center, Cincinnati, Ohio, March 2017

#### PROFESSIONAL AFFILIATIONS/SOCIETY MEMBERSHIPS

- American Pediatric Society
- American Physiological Society
- American Society for Clinical Investigation
- American Society of Nephrology
- American Society of Pediatric Nephrology
- International Society of Nephrology
- International Pediatric Nephrology Association
- National Kidney Foundation
- Salt and Water Club
- Society for Pediatric Research

### Ÿ

#### Michael Moritz, MD

#### **RESEARCH**

Michael Moritz focuses on the epidemiology and treatment of dysnatremias in children. He introduced the concept of using 0.9% sodium chloride in maintenance parenteral fluids for the prevention of hospital-acquired hyponatremia, and he has been an outspoken advocate for this therapy. This has resulted in a worldwide change in medical practice in both children and adults.

He also introduced the concept of using intermittent 3% sodium chloride boluses for the treatment of hyponatremic encephalopathy, and this is now being accepted as the treatment of choice by consensus guidelines. He is an internationally recognized expert in the controversial topic of salt poisoning in children and has served as a medical expert in high-profile trials in the United Kingdom and United States. He also has conducted seminal work in the epidemiology and prevention of breastfeeding-associated hypernatremia. He was the first to report on bleeding complications from high-concentration heparin to pack hemodialysis catheters in children.

He is clinical director of the Division of Nephrology and the medical director of Children's Hospital of Pittsburgh's Pediatric Dialysis Unit. His specific research interests include disorders in sodium and water metabolism (central pontine myelinolysis, hyponatremic encephalopathy, hypernatremic dehydration, salt poisoning, fluid therapy), hemodialysis access in children, and pediatric hypertension trials.

Moritz is also the chief editor of *Frontiers in Pediatrics*. In that capacity, he oversees 20 journal sections and almost 3,000 editorial board members. In 2016, he was selected for the Chief Editor's Award out of a field of 54 chief editors.

#### **ADVISORY COMMITTEE MEMBERSHIPS**

- Content Development Team, American Board of Pediatrics
- Section of Fluid and Electrolyte Therapy in Children, Clinical Practice Guideline Subcommittee, American Academy of Pediatrics
- Clinical Affairs Committee, American Society of Pediatric Nephrology
- Committee for "Making Dialysis Safer," Centers for Disease Control and Prevention
- Medical Advisory Board, National Kidney Foundation Serving the Alleghenies

#### **EDITORSHIPS**

- Field chief editor, Frontiers in Pediatrics
- Specialty chief editor, Pediatric Nephrology Section, Frontiers in Pediatrics
- Associate editor, Nephrology Section, Frontiers in Medicine
- Research topic editor, "Recent Advances in Hyponatremia," Frontiers in Medicine and Frontiers in Pediatrics
- Member, Editorial Board, Pediatric Nephrology

#### **MAJOR LECTURESHIPS AND SEMINARS**

 "Outpatient Pediatric Nephrology," meeting of the Three Rivers Chapter of the National Association of Pediatric Nurse Practitioners, Pittsburgh, Pa., December 2016

- "What Nurses Need to Know About Hyponatremia," American Nephrology Nurses Association Spring Ahead meeting, Pittsburgh, Pa., May 2017
- "Prevention and Treatment of Hospital-Acquired Hyponatremia," pediatric grand rounds, Loma Linda University Health, Loma Linda, Calif., June 2017
- Prevention and Treatment of Hospital-Acquired Hyponatremia," pediatric grand rounds, New York University School of Medicine, New York, N.Y., June 2017

#### PROFESSIONAL AFFILIATIONS/SOCIETY MEMBERSHIPS

- · American Academy of Pediatrics
- · American Society of Nephrology
- American Society of Pediatric Nephrology
- International Pediatric Nephrology Association
- National Kidney Foundation
- · Society for Pediatric Research

#### **HONORS**

- Field Chief Editor's Award, Frontiers Journals, 2016
- Chair Distinction Award for outstanding service, 2016
- Top Doctors, Castle Connolly, 2017
- Best Doctors in America, Woodward/White, Inc., 2017
- Best Doctors, Pittsburgh Magazine, 2017
- University of Pittsburgh Honors Convocation, faculty honoree, 2017
- Publons Peer Review Award, top 1 percent peer reviews in medicine, 2017
- Certificate of Expertise, International Consortium for Clinical Research Excellence, Education, and Ethics, 2017

#### Rannar Airik, PhD

#### RESEARCH

Rannar Airik's research focuses on deciphering the mechanisms of chronic kidney disease using mouse models of a genetic form of childhood chronic kidney disease, nephronophthisis (NPHP), an autosomal recessive chronic kidney disease characterized by tubulointerstitial fibrosis, tubular basement membrane disruption, and kidney cysts. Renal fibrosis is the primary determinant of end-stage kidney disease. The pathomechanisms underlying NPHP are only poorly understood. Although NPHP has long been considered a "ciliopathy," caused by a dysfunction in cilia, recent gene identification in humans has linked the pathogenesis of NPHP to defective DNA damage response signaling, resulting in genome instability and cell-cycle defects. To study the disease mechanisms of NPHP and to understand the cellular and molecular mechanisms of

DNA damage response in the renal pathology of NPHP, Airik has generated knockout mouse models of the human condition. He has employed primary and secondary cell-culture models of NPHP to interrogate the disease mechanisms. His research is supported by one NIH grant and several intramural funds.

#### STUDY SECTIONS

• Ad hoc reviewer, Kidney Research UK

#### MAJOR LECTURESHIPS AND SEMINARS

- "The Nephronophthisis Gene Anks6 Regulates Glomerular Development," internal medicine grand rounds, UPMC Presbyterian Hospital, University of Pittsburgh School of Medicine, June 2017
- Lecturer, Integrative Systems Biology Graduate School course Model Systems (#2035), University of Pittsburgh School of Medicine

#### PROFESSIONAL AFFILIATIONS/SOCIETY MEMBERSHIPS

- American Society of Nephrology
- American Society of Human Genetics

#### **Demetrius Ellis, MD**

#### RESEARCH

Demetrius Ellis's research interests include diabetic nephropathy, pediatric hypertension, and pediatric transplantation. His research is supported, in part, by an NIH grant in collaboration with Trevor Orchard (Epidemiology of Diabetes Complications, phase II, DK34818-20).

#### **ADVISORY COMMITTEE MEMBERSHIPS**

 Ad hoc, Promotion Committee, University of Pittsburgh School of Medicine

#### PROFESSIONAL AFFILIATIONS/SOCIETY MEMBERSHIPS

- American Society of Pediatric Nephrology
- Council of Pediatric Nephrology
- International Pediatric Nephrology Association
- National Kidney Foundation Serving the Alleghenies

#### **HONORS**

- Excellence in Pediatrics, Academic Pediatrician,
   Children's Hospital of Pittsburgh of UPMC physicians and Children's Hospital of Pittsburgh of UPMC alumni
- Highest overall teaching grade by medical students, nephrology fellows, and pediatric residents
- Best Doctors in America, Woodward/White, Inc.
- Best Doctors, Pittsburgh Magazine (33 consecutive years)

Dana Fuhrman, DO, MS

Dana Fuhrman serves as a pediatric intensivist and nephrologist with an overall research interest in predicting and preventing acute kidney injury (AKI) in children and young adults. Specifically, she works toward establishing a method to predict a kidney's response to stress and decline in glomerular filtration rate over time. She previously studied and published a method to quantify renal functional reserve in healthy young adults. She plans to determine whether renal functional reserve values and differences in baseline tubular biomarkers may serve to identify a kidney with favorable "renal fitness," defined by the ability to respond well to renal stress and, therefore, show a decreased risk of acute kidney injury and chronic kidney disease. She has studied young adult patients with congenital heart disease who are at risk for numerous kidney injuries across a lifetime and the development of chronic kidney disease. She was a co-investigator for the Prospective Pediatric Acute Kidney Injury Research Group through Cincinnati Children's Hospital, an international research consortium dedicated to advancing research in pediatric acute kidney injury.

#### **EDITORSHIP**

• Review editor, Pediatric Critical Care Medicine, 2015 to the present

#### MAJOR LECTURESHIPS AND SEMINARS

• "Renal Replacement Therapy on Extracorporeal Membrane Oxygenation (ECMO)," Children's Hospital of Pittsburgh's annual ECMO conference, August 2016

#### PROFESSIONAL AFFILIATIONS/SOCIETY MEMBERSHIPS

- American Society of Nephrology
- · American Society of Pediatric Nephrology
- Society of Critical Care Medicine

#### Jacqueline Ho, MD

#### **RESEARCH**

Jacqueline Ho studies requirements for miRNAs in kidney progenitor cells during renal development and in the regulation of podocyte structure and function during proteinuric kidney diseases. miRNAs represent a fundamentally new means of controlling gene expression and have been implicated in the regulation of diverse developmental processes. Her previous work demonstrated a critical role for miRNAs in the regulation of kidney progenitor survival during development and in the maintenance of podocyte structure. Work in her laboratory focuses on the

identification of specific miRNAs and their mRNA targets that mediate these effects. Her research is supported by an NIH R01 grant and intramural funds. She is a coinvestigator on another R01 and a U24.

#### STUDY SECTIONS

· Reviewer, Division of Kidney, Urologic, and Hematologic Diseases Fellowship, NIDDK

#### **ADVISORY COMMITTEE MEMBERSHIPS**

- Research Committee, American Society of Pediatric Nephrology
- · Program Committee, American Society of Pediatric Nephrology

#### **EDITORSHIP**

· Associate editor, section on pediatric nephrology, Frontiers in Pediatrics

#### **MAJOR LECTURESHIPS AND SEMINARS**

- · "Correlation of Prenatal Imaging of Kidney Malformations as Basis for Intervention, Including Recent Experience With Pediatric Outcomes," Developmental Renal Malformations, Oligo/Anhydramnios: Pathophysiology and Clinical Aspects, National Institute of Child Health and Human Development workshop, Bethesda, Md., August 2016
- "Small RNAs and Small Kidneys," pediatric nephrology grand rounds, Children's Hospital of Philadelphia, Philadelphia, Pa., December 2016
- "Small RNAs and Small Kidneys," renal grand rounds, Northwestern University, Chicago, Ill., December 2016
- "Small RNAs and Small Kidneys," experimental biology meeting, Chicago, Ill., April 2017
- · "Small RNAs and Small Kidneys: A Role for the miR-17~92 Cluster," Senior Vice Chancellor's Seminar Series, University of Pittsburgh School of Medicine, Pittsburgh, Pa., April 2017

#### PROFESSIONAL AFFILIATIONS/SOCIETY MEMBERSHIPS

- American Academy of Pediatrics
- American Society of Nephrology
- · American Society of Pediatric Nephrology
- · Canadian Society of Nephrology
- · Canadian Association of Pediatric Nephrologists
- International Pediatric Nephrology Association
- · Society for Pediatric Research

#### **HONORS**

 Renée Habib Young Investigator Award, International Pediatric Nephrology Association, 2013



#### Yosuke Miyashita, MD, MPH

#### **RESEARCH**

Yosuke Miyashita's research interests focus on pediatric hypertension and Shiga toxin-producing Escherichia coli, as well as atypical hemolyticuremic syndrome. He was the representative for Children's Hospital of Pittsburgh of UPMC in the Midwest Pediatric Nephrology Consortium (MWPNC). He continues to participate in multicenter clinical trials with this consortium, especially in the field of pediatric hypertension and the use of 24-hour ambulatory blood pressure monitoring. His most recent efforts are aimed at formulating an evidence-based practice guideline for white-coat hypertension in children and adolescents. For this effort, he recently completed and published a survey study of pediatric nephrologists in the MWPNC centers regarding whitecoat hypertension. He investigated the changes of 24-hour ambulatory blood pressure monitoring results over time for children and adolescents who were being followed for blood pressure concerns but were not on antihypertensive medication. Those were pilot studies for an eventual multicenter retrospective study to investigate the same issue in a larger MWPNC cohort. Lastly, he served as the faculty mentor for two trainees in their respective projects in the MWPNC.

#### **ADVISORY COMMITTEE MEMBERSHIPS**

- Clinical Affairs Committee, American Society of Pediatric Nephrology
- MWPNC

#### **EDITORSHIP**

 Review editor, Section on Pediatric Nephrology and Nephrology, Frontiers in Pediatrics and Frontiers in Medicine

#### **MAJOR LECTURESHIPS AND SEMINARS**

 "Evaluation and Management of White-Coat Hypertension," MWPNC meeting, Minneapolis, Minn., October 2016

#### PROFESSIONAL AFFILIATIONS/SOCIETY MEMBERSHIPS

- American Society of Pediatric Nephrology
- International Pediatric Hypertension Association
- International Pediatric Nephrology Association

#### Christina Nguyen, MD

#### RESEARCH

Christina Nguyen focuses her research in the areas of transplant outcomes, transition of care between pediatric and adult medicine, and infectious complications in kidney transplantation. She is the director of the Pediatric Kidney Transplant Program. She serves as a site sub-investigator in two multicenter, NIH-funded research projects in the field of transplantation. She is a co-investigator for an internal grant with the Hillman Foundation looking at neurocognitive outcomes in very young recipients of kidney transplants. She serves as the site principal investigator in two pharmaceutical trials and a co-investigator on one other pharmaceutical trial. Active projects include outcomes and epidemiology of donor-specific antibodies in pediatric kidney transplantation, a case series on the use of bortezomib to treat acute antibody-mediated rejection, and stratification of pre-transplant psychosocial risk factors that are associated with high-risk post-transplant behaviors in kidney transplant recipients.

#### **ADVISORY COMMITTEE MEMBERSHIPS**

- Pediatric to Adult Transition Subcommittee, American Society of Transplantation
- Education Committee, American Society of Pediatric Nephrology
- Outreach Committee, International Pediatric Transplant Association

#### **EDITORSHIP**

- Review editor, Frontiers in Pediatrics, 2014 to the present
- Editorial Board for Kidney, Pediatric Transplantation, 2017

#### PROFESSIONAL AFFILIATIONS/SOCIETY MEMBERSHIPS

- American Society of Pediatric Nephrology
- American Society of Transplantation
- International Pediatric Transplant Association

#### **HONORS**

 Chair Distinction Award, Children's Hospital of Pittsburgh of UPMC, 2016

#### Sunder Sims-Lucas, PhD

#### **RESEARCH**

Sunder Sims-Lucas is a basic research scientist whose focus is on the formation of the developing vasculature in the kidney and the predisposition to adult-onset disease when there is defective development. He is involved in a major initiative by the NIH in the growth of new kidney tissues. His laboratory also studies blood flow and hypoxia and the role they play in patterning the various renal lineages. He recently began exploring the role of the microvasculature in acute kidney injury. He has authored more than 45 publications and, despite being an early-stage investigator, has been supported by many NIH grants, including a K01, an R03, and U24 grants. He has acquired local awards,

#### STUDY SECTIONS

- Reviewer, ad hoc, P30 grants, NIDDK, NIH
- Reviewer, ad hoc, R03 grants, NIDDK, NIH
- Reviewer, (Re)Building a Kidney, Partnership Program grants, NIDDK
- Reviewer, Diabetic Complications Consortium (DiaComp) Pilot and Feasibility Program, NIDDK

#### **ADVISORY COMMITTEE MEMBERSHIPS**

- Faculty advisor for summer student interns, Children's Hospital of Pittsburgh
- Co-chair, Organizing Committee, Rangos Research Symposium, Children's Hospital of Pittsburgh
- Graduate Schools Admission Committee

#### **EDUCATIONAL ROLES**

- Appointed director of student research training at the Children's Hospital of Pittsburgh
- Member of the educational committee at the Children's Hospital of Pittsburgh
- Designed a new course titled, "Pediatrics: Bench to Bedside," which is run out of the Honors College

#### **EDITORSHIPS**

- Editorial Board, Frontiers in Pediatrics
- Special guest editor, Frontiers in Cell and Developmental Biology, Organogenesis: From Development to Disease

#### **MAJOR LECTURESHIPS AND SEMINARS**

- "Renal Endothelial Progenitors During Development and Injury," 13th International Workshop on Developmental Nephrology, Snowbird Ski and Summer Resort, Salt Lake City, Utah, July 2015
- "Blood Flow and Oxygenation During Kidney Development and Disease," Molecular Medicine Seminar Series, Department of Pediatrics, Children's Hospital of Pittsburgh, Pa., 2016

#### PROFESSIONAL AFFILIATIONS/SOCIETY MEMBERSHIPS

- American Heart Association
- · American Physiological Society
- · American Society of Nephrology
- North American Vascular Biology Organization

#### Agnieszka Swiatecka-Urban, MD

#### **RESEARCH**

Agnieszka Swiatecka-Urban studies regulation of cell-surface stability and intracellular trafficking of membrane proteins in epithelial cells. One focus of her research activity has been characterization of mechanisms and pathways that regulate the cystic fibrosis transmembrane conductance regulator (CFTR) in order to improve treatment strategies for cystic fibrosis. Swiatecka-Urban has also used her expertise to address a significant unmet need of patients with nephrotic syndrome, a severe form of kidney disease responsible for 20% of pediatric kidney failure. She has been studying the role of protein-protein interactions in the dynamic regulation of nephrin in the podocyte slit diaphragm—an intercellular junction, which is compromised in nephrotic syndrome. Swiatecka-Urban's work on both nephrin and CFTR has resulted in several successful research awards. Her research is supported by an NIH R56 award, an NIH P30 pilot and feasibility sub-award, an NIH cooperative agreement sub-award, and an American Heart Association grant. Finally, she leads the University of Pittsburgh site for the CureGlomerulopathy (CureGN) network cohort study—a multicenter, prospective cohort observational study for patients with nephrotic syndrome.

#### **STUDY SECTIONS**

- Research and Research Training Committee, Cystic Fibrosis Foundation
- Basic Cell MSO1 Study Section, American Heart Association
- Ad hoc reviewer, Digestive, Kidney, and Urological Systems, NIH
- Review Committee, University of Pittsburgh Competitive Medical Research Fund
- · Research Council United Kingdom
- University of Pittsburgh Competitive Research Fund
- Research Fellowship Grants Committee, Mallinckrodt Pharmaceuticals
- Innovative Research Grant Basic Science 3 Study Section, American Heart Association

#### **ADVISORY COMMITTEE MEMBERSHIPS**

- Competitive Medical Research Fund
- Research Council United Kingdom
- PhD Defense Committee, Center for Biodiversity, Functional, and Integrative Genomics, University of Lisbon, Lisbon, Portugal
- PhD Defense Committee, Geisel School of Medicine, Dartmouth College, Hanover, N.H.
- Mallinckrodt Pharmaceuticals, Inc.
- Alexion Pharmaceuticals, Inc.



- Chair, annual Nephrotic Syndrome Symposium: Evidence-Based Management and Personalized Approach, Children's Hospital of Pittsburgh
- Science Policy and Public Policy committees, Personalized Medicine Coalition
- Member, Program Committee, Pediatric Academic Societies/American Society of Pediatric Nephrology, 2016 to the present
- Workshop champion, The Future of Nephrology: Tissue Regeneration, 3D Printing, and the Wearable Kidney, scheduled for the 2017 Pediatric Academic Societies/ American Society of Pediatric Nephrology meeting
- Program Committee, Pediatric Academic Societies/ American Society of Pediatric Nephrology

#### **EDITORSHIPS**

- Editorial Board, American Journal of Physiology— Renal Physiology
- Editorial Board, Conference Papers in Cell Biology
- Editorial Board, Frontiers in Physiology
- Editorial Board, Journal of Visual Experimentation
- Frontiers in Pediatric Research
- Associate guest editor: nephrotic syndrome, Frontiers in Pediatric Research
- Associate editor, section on Pediatric Nephrology
- British Journal of Pharmacology

#### **MAJOR LECTURESHIPS AND SEMINARS**

 "Role of MicroRNA 145 in TGFβ1-Mediated Repression of CFTR in Primary Human Bronchial Epithelium," 131st Salt and Water Club Meeting, University of Pittsburgh School of Medicine, 2016  "Incidence of Pediatric Nephrotic Syndrome—Single Center Experience," National Kidney Foundation and Kidney and Urology Foundation of America Nephrotic Syndrome Symposium, New York, N.Y., 2016

#### PROFESSIONAL AFFILIATIONS/SOCIETY MEMBERSHIPS

- American Society of Nephrology
- American Society of Pediatric Nephrology
- International Pediatric Nephrology Association
- International Society of Nephrology
- · Salt and Water Club
- Society for Pediatric Research

#### **HONORS**

- Fellow, American Society of Nephrology
- Top Doctors, Castle Connolly, 2015
- Chair, Third Annual Symposium, Nephrotic Syndrome: Clinical Challenges and Evidence-Based Management, Pittsburgh, Pa., 2016
- Faculty, BioSys-PhD Program in Biological Systems, Functional and Integrative Genomics. BioSys-PhD is a multidisciplinary PhD program sponsored by the Portuguese Foundation for Science and Technology. Students receive 50% of their training in Swiatecka-Urban's lab, 2015.
- Tenured associate professor of pediatrics and cell biology, University of Pittsburgh School of Medicine
- UPMC's Excellence in Patient Experience, Physician and Medical Staff Honor Roll
- Award for Excellence in Patient Experience, Physician and Medical Staff, presented by the Wolff Center at UPMC

#### THREE-YEAR BIBLIOGRAPHY

# 2015

Acker SN, Mandell EW, **Sims-Lucas S**, Gien J, Abman SH, Galambos C. Histologic identification of prominent intrapulmonary anastomotic vessels in severe congenital diaphragmatic hernia. *Journal of Pediatrics*. 2015;166:178-83.

Agathis NT, Libman IM, **Moritz ML**. Hyponatremia due to severe primary hypothyroidism in an infant. *Front Pediatr*. 2015; 3(96):1-4.

Ashokkumar C, Ganguly B, Townsend R, White J, Levy S, **Moritz M**, Mazariegos G, Sun Q, Sindhi R. Alloreactive CD154-expressing T-cell subsets with differential sensitivity to the immunosuppressant

belatacept: Potential targets of novel belatacept-based regimens. *Sci Rep.* 2015; 5:15218.

Ayus JC, Caputo D, Bazerque F, Heguilen R, Gonzalez CD, **Moritz ML**. Treatment of hyponatremic encephalopathy with 3% sodium chloride protocol: A case series. *Am J Kidney Dis*. 2015; 65(3):435-42.

**Ellis D**, Lieb J. Hyperoxaluria and genitourinary disorders in children ingesting almond milk products. *J Pediatr*. 2015;167:1155-8.

Ellis D, Miyashita Y. Management of the hypertensive child. In: Avner E, Harmon W, Niaudet P, Yoshikawa N, eds. *Pediatric Nephrology.* Springer Berlin Heidelberg; 2015: 1-87.

Di Giovanni V, Walker KA, Bushnell D, Schaefer C, Sims-Lucas S, Puri P, Bates CM. Fibroblast growth factor receptor-Frs $2\alpha$  signaling is critical for nephron progenitors. *Dev Biol.* 2015;400(1):82-93.

Galambos C, **Sims-Lucas S**, Ali N, Gien J, Dishop MK, Abman SH. Intrapulmonary vascular shunt pathways in alveolar capillary dysplasia with misalignment of pulmonary veins. *Thorax*. 2015;70:84-5.

Joyce E, **Ho J**, El-Gharbawy A, Salgado CM, Ranganathan S, Reyes-Mugica M. The value of renal biopsy in diagnosing infantile nephropathic cystinosis associated with secondary nephrogenic diabetes insipidus. *Pediatr Dev Pathol*. 2015; 20(1):72-75.

Junttila S, Saarela U, Halt K, Manninen A, Parssinen H, Lecca MR, Brandli AW, **Sims-Lucas S**, Skovorodkin I, Vainio SJ. Functional genetic targeting of embryonic kidney progenitor cells *ex vivo. J Am Soc Nephrol.* 2015;26:1126-37.

**Moritz ML**, Ayus JC. Disorders of plasma sodium. *N Eng J Med*. 2015 Mar 26;372(13).

**Moritz ML**, Ayus JC. Hyponatraemia: Isotonic fluids prevent hospital-acquired hyponatremia. *Nat Rev Nephrol*. 2015;11(4):202-3.

**Moritz ML**, Ayus JC. Maintenance intravenous fluids in acutely ill patients. *N Engl J Med*. 2015;373(14):1350-60.

Phua YL, **Ho J**. MicroRNAs in the pathogenesis of cystic kidney disease. *Curr Opin in Pediatr.* 2015;27:219-26.

Pastor-Soler NM, Sutton TA, Mang HE, Kinlough CL, Gendler SJ, Madsen CS, Bastacky SI, **Ho J**, Al-Bataineh MM, Hallows KR, Singh S, Monga SP, Kobayashi H, Haase VH, Hughey RP. Muc1 is protective during kidney ischemia-reperfusion injury. *Am J Physiol Renal Physiol*. 2015;308(12):F1452-62.

Phua YL, Chu JYS, Marrone AK, Bodnar AJ, **Sims-Lucas S**, **Ho J**. Renal stromal miRNAs are required for normal nephrogenesis and glomerular mesangial survival. *Physiol Reports*. 2015;3(10).

Rymer CC, **Sims-Lucas S**. *In utero* intra-cardiac tomato-lectin injections on mouse embryos to gauge renal blood flow. *J Vis Exp.* 2015;96.

Stolz DB, **Sims-Lucas S**. Unwrapping the origins and roles of the renal endothelium. *Pediatric Nephrology*. 2015;30:865-72.

Walker KA, Ikeda Y, Zabbarova I, Schaefer CM, Bushnell DS, de Groat WC, Kanai AJ, **Bates CM**. Fgfr2 is integral for bladder mesenchyme patterning and function. *Am J Physiol Renal Physiol*. 2015;308(8):F888-98.

Webb TN, Griffith H, Miyashita Y, Bhatt R, Jaffe R, Moritz M, Hofer J, Swiatecka-Urban A. Atypical hemolytic uremic syndrome and chronic ulcerative colitis treated with eculizumab. *Int J Med Pharm Case Reports*. 2015;4(5):105-12.

# 2016

**Airik R\***, Schueler M, Airik M, Cho J, Porath JD, Mukherjee E, Sims-Lucas S, Hildebrandt F\*. A FANCD2/FANCI-associated nuclease 1-knockout model develops karyomegalic interstitial nephritis. *J Am Soc Nephrol.* 2016 27(12):3552-9. \*Cocorresponding authors

Airik R\*, Schueler M, Airik M, Cho J, Ulanowicz KA, Porath JD, Hurd TW, Bekker-Jensen S, Schrøder JM, Andersen JS, Hildebrandt F\*. SDCCAG8 interacts with RAB effector proteins RABEP2 and ERC1 and is required for hedgehog signaling. *PLoS One*. 2016;11(5):e0156081. \*Co-corresponding authors

Ayus JC, Fuentes NA, Negri AL, **Moritz ML**, Giunta DH, Kalantar-Zadeh K, Nigwekar SU, Thadhani RI, Go AS, De Quiros FG. Mild prolonged chronic hyponatremia and risk for hip fracture in the elderly. *Nephrol Dial Transplant*. 2016. Oct;31(10):1662-9.

Bebee TW, **Sims-Lucas S**, Park JW, Bushnell D, Cieply B, Xing Y, **Bates CM**\*, Carstens RP\*. Ablation of the epithelial-specific splicing factor Esrp1 results in ureteric branching defects and reduced nephron number. 2016. *Dev Dyn.* 245(10):991-1000. \*Cocorresponding authors

Casinelli G, LaRosa J, Sharma M, Cherok E, Banerjee S, Branca M, Edmunds L, Wang Y, **Sims-Lucas** S, et al. N-Myc overexpression increases cisplatin resistance in neuroblastoma via deregulation of mitochondrial dynamics. *Cell Death Discov.* 2016;2:16082.

Farinha CM, **Swiatecka-Urban A**, Brautigan DL, Jordan P. Regulatory crosstalk of protein kinases on CFTR trafficking and function. *Front Chem.* 2016;4:1.

**Fuhrman DY**, Kellum JA. Remote ischemic preconditioning in the PICU: A simple concept with a complex past. *Pediatr Crit Care Med*. 2016;17:e371-9.

**Fuhrman DY**, Kellum JA. Biomarkers for diagnosis, prognosis, and intervention in acute kidney injury. *Contributions to Nephrology*. 2016;187:47-54.

Gee HY, Sadowski CE, Aggarwal PK, Porath JD, Yakulov TA, Schueler M, Lovric S, Ashraf S, Braun DA, Halbritter J, Fang

H, Airik R, Vega-Warner V, Cho KJ, Chan TA, Morris LG, Ffrench-Constant C, Allen N, McNeill H, Büscher R, Kyrieleis H, Wallot M, Gaspert A, Kistler T, Milford DV, Saleem MA, Keng WT, Alexander SI, Valentini RP, Licht C, Teh JC, Bogdanovic R, Koziell A, Bierzynska A, Soliman NA, Otto EA, Lifton RP, Holzman LB, Sibinga NE, Walz G, Tufro A, Hildebrandt F. FAT1 mutations cause a glomerulotubular nephropathy. *Nat Commun.* 2016;7:10822.

Hemker SL, **Sims-Lucas S**, **Ho J**. The role of hypoxia in nephrogensis. *Pediatr Nephrol.* 2016;31(10):1571-7.

Joyce EL, Kane-Gill SL, **Fuhrman, DY**, Kellum JA. Drug-associated acute kidney injury: Who's at risk? *Pediatr Nephrol*. 2016;32:59-69.

Liu X, Edinger RS, Klemens CA, Phua, YL, Bodnar AJ, LaFramboise WA, **Ho J**, Butterworth MB. A microRNA cluster miR-3~24~27 is upregulated by aldosterone in the distal kidney nephron where it alters sodium transport. *J Cell Physiol*. 2016 Sept 16.

Maringer K, **Sims-Lucas S**. The multifaceted role of the renal microvasculature during acute kidney injury. *Pediatr Nephrol.* 2016;31(8):1231-40.

Maringer K, Yarbrough A, **Sims-Lucas S**, Saheb E, Jawed S, Bush J. Dictyostelium discoideum RabS and Rab2 colocalize with the Golgi and contractile vacuole system and regulate osmoregulation. *J Biosci*. 2016;41(2):205-17.

**Moritz ML**, Ayus JC. Misconceptions in the treatment of dehydration in children. *Pediatr Rev.* 2016;37(7):e29-31.

**Moritz ML**, Ayus JC. Maintenance intravenous fluids in acutely ill patients. *N Engl J Med*. 2016;374(3):290-1.

Phua YL, **Ho J**. Renal dysplasia in the neonate. *Curr Op Pediatr*. 2016;28:209-15.

Puri P, Bushnell D, Schaefer CM, **Bates CM**. Frs2α knockout mice are a novel model of renal cystogenesis. *Sci Rep*. 2016;6:36736.

Shukla S, Basu A, **Moritz ML**. Use of hypotonic maintenance intravenous fluids and hospital-acquired hyponatremia remain common in children admitted to a general pediatric ward. *Front Pediatr.* 2016 4:90.

Squires J, **Nguyen C**. Complexity of preemptive liver transplantation in children with primary hyperoxaluria type 1. *Pediatr Transplant*. 2016;20(5):604-6.

Song R, Preston G, Kidd L, Bushnell D, Sims-Lucas S, **Bates CM**, Yosypiv IV. Prorenin receptor is critical for nephron progenitors. *Dev Biol*. 2016;409(2):382-91.

Walker KA, **Sims-Lucas S, Bates CM**. Fibroblast growth factor receptor signaling in kidney and lower urinary tract development. *Pediatr Nephrol*. 2016;31(6):885-95.

# 2017

Cerqueira DM, Bodnar AJ, Phua YL, Freer R, Hemker SL, Walensky LD, Hukriede NA, **Ho J**. Bim gene dosage is critical in modulating nephron progenitor survival in the absence of microRNAs during kidney development. *FASEB J*. 2017;31(8):3540-54.

Dangle P, Salgado C, Reyes-Mugica M, Schneck F, Ost M, **Sims-Lucas S**. Testicular hypoplasia is driven by defective vascular formation. *Urology*. 2017;101:94-8.

**Fuhrman DY**, Crowley K, Vetterly C, Hoshitsuki K, Koval A. Medication use as a contributor to fluid overload in the PIUC: A prospective observational study. *J Pediatr Intensive Care*. In press.

**Fuhrman DY**, Kellum JA. Epidemiology and pathophysiology of cardiac surgery–Associated acute kidney injury. *Curr Opin Anaesthesiol*. 2017;30(1):60-5.

**Fuhrman DY**, Schneider MF, Dell KM, Blydt-Hansen TD, Mak R, Saland JM, Furth SL, Warady BA, Moxey-Mims MM, Schwartz GJ. Albuminuria, proteinuria, and renal disease progression in children with CKD. *Clin J Am Soc Nephrol*. In press.

Geramita MA, Hofer J, Cooper J, **Moritz ML**. Decreased severity of Shiga toxin-producing *Escherichia coli* haemolytic uraemic syndrome (STEC-HUS) in a child with type 1 von Willebrand disease. *BMJ Case Rep*. In press.

Ikeda Y, Zabbarova I, Schaefer CM, Bushnell DS, de Groat WC, Kanai AJ, **Bates CM**. Fgfr2 is integral for bladder mesenchyme patterning and function. *Am J Physiol Renal Physiol*. 2017;312(4):F607-18.

Joyce E, **Ellis D**, **Miyashita Y**. Chapter 14: Nephrology. In: Zitelli BJ, McIntire SC, Nowalk AJ (Eds.), *Zitelli and Davis' Atlas of Pediatric Physical Diagnosis* (7<sup>th</sup> ed.). Philadelphia, PA: Saunders; 2017.

Joyce E, Glasner P, Ranganathan S, **Swiatecka-Urban A**. Tubulointerstitial nephritis: Diagnosis, treatment, and monitoring. *Pediatr Nephrol*. 2017;32(4):577-87.

Joyce E, **Ho J**, El-Gharbawy A, Salgado CM, Ranganathan S, Reyes-Mugica M. The value of renal biopsy in diagnosing infantile nephropathic cystinosis associated with secondary nephrogenic diabetes insipidus. *Pediatr Dev Pathol*. 2017;20(1):72-5.

**Miyashita Y**, Flynn JT, Hanevold CD. Diagnosis and management of white-coat hypertension in children and adolescents: A Midwest Pediatric Nephrology Consortium study. *J Clin Hypertens*. 2017;19:884-9.

**Moritz ML**, Ayus JC. How to improve intravenous fluid prescribing practices in bronchiolitis. *Hosp Pediatr*. 2017;7(5):300-2.

Mukherjee, E, Maringer KV, Papke E, Bushnell DS, Schaefer CM, Kramann R, Ho J, Humphreys BD, Bates CM, Sims-Lucas S. Endothelial markers expressing stromal cells are critical for kidney formation. *Am J Physiol Renal Physiol*. In press.

Narla D, Slagle S, Schaefer CM, Bushnell DS, Puri P, **Bates CM**. Loss of peri-Wolffian duct stromal Frs2α expression in mice leads to abnormal ureteric bud induction and vesicoureteral reflux. *Pediatr Res.* In press.

Puri P, Schaefer CM, Bushnell D, Taglienti ME, Kreidberg JA, Yoder BK, **Bates CM**. Ectopic phosphorylated Creb marks dedifferentiated proximal tubules in cystic kidney disease. *Am J Pathol*. In press.

Peterson CG, **Miyashita Y**. The use of ambulatory blood pressure monitoring as standard of care in pediatrics. *Front Pediatr*. 2017;5:153.

Rodenbach KE, **Fuhrman DY**, Maier PS, Schwartz GJ. Renal response to a protein load in healthy young adults as determined by iohexol infusion clear-

ance, cimetidine-inhibited creatinine clearance, and cystatin C estimated glomerular filtration rate. *J Ren Nutr.* 2017;4:275-81.

Sims-Lucas S, Good M, Vainio SJ. Editorial: Organogenesis: From development to disease. *Front Cell Dev Biol*. 2017;5:85.

Sindhi R, Higgs BS, Venkataramanan R, **Moritz ML**. Synergistic immunosuppression and unintended consequences. *Pediatr Transplant*. In press.

**Swiatecka-Urban, A**. Endocytic trafficking at the mature podocyte slit diaphragm. *Front Pediatr.* 2017;5:32.

Vermeren M, Lyraki R, Wani S, **Airik R**, Albagha O, Mort R, Hildebrandt F, Hurd T. Osteoclast stimulation factor 1 (Ostf1) KNOCKOUT increases trabecular bone mass in mice. *Mamm Genome*. In press.

Woroniecki R, **Swiatecka-Urban A**, Kaskel FJ. Editorial: Nephrotic syndrome in pediatric patients. *Front Pediatr*. 2017;3(5):167.

Zhang Y, Bharathi SS, Rardin MJ, Lu J, Maringer KV, **Sims-Lucas S**, Prochownik EV, Gibson BW, Goetzman ES. Lysine desuccinylase SIRT5 binds to cardiolipin and regulates the electron transport chain. *J Biol Chem.* 2017;292(24):10239-49.