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DIVISION OF GASTROENTEROLOGY, HEPATOLOGY, AND NUTRITION

Mission

The mission of the Division of Gastroenterology, Hepatology, and Nutrition is:

- To provide up-to-date and compassionate care to children with gastrointestinal (GI) and liver disorders
- To educate the next generation of gastroenterologists and pediatricians
- To advance knowledge of pediatric gastroenterology through research

FACULTY

David Keljo, MD, PhD
Interim Chief, Division of Gastroenterology, Hepatology, and Nutrition (April 1, 2017, to the present)
Professor of Pediatrics
Codirector, Inflammatory Bowel Disease (IBD) Program

Mark E. Lowe, MD, PhD
Chief, Division of Gastroenterology, Hepatology, and Nutrition (until March 31, 2017)
Professor of Pediatrics
Vice Chair of Graduate Medical Education

Feras Alissa, MD
Assistant Professor of Pediatrics
Director, Endoscopy Center

Riha Bhatt, MD
Assistant Professor of Pediatrics
Codirector, Pediatric Gastroenterology Fellowship Program

Kristy Boggs, PhD
Research Assistant Professor

Maria I. Clavell, MD
Associate Professor of Pediatrics

John F. Eisses, MD, PhD
Assistant Professor of Pediatrics

Tamara Feliciano Alvarado, MD
Assistant Professor of Pediatrics

Arjumand Ghazi, PhD
Assistant Professor of Pediatrics, Developmental Biology, and Cell Biology and Physiology

Sohail Z. Husain, MD
Associate Professor of Pediatrics
Director, Exocrine Pancreas Center (EPC)

Zahida Khan, MD, PhD
Assistant Professor of Pediatrics and Pathology

Sandra C. Kim, MD
Associate Professor of Pediatrics
Codirector, IBD Center

Dale King, MD
Assistant Professor of Pediatrics

Douglas Lindblad, MD
Assistant Professor of Pediatrics

Patrick J. McKiernan, MD
Professor of Pediatrics
Director, Pediatric Hepatology Program

Amitava Mukherjee, PhD
Research Instructor

Jeffrey A. Rudolph, MD
Associate Professor of Pediatrics

Wednesday Marie A. Sevilla, MD, MPH, CNSC
Assistant Professor of Pediatrics

Sapana Shah, MD
Assistant Professor of Pediatrics

Leah Siebold, MD
Assistant Professor of Pediatrics
Clinical Director of Gastroenterology

Amina Smajlovic, MD
Clinical Assistant Professor of Pediatrics

James E. Squires, MD, MS
Assistant Professor of Pediatrics
Director, Pediatric Transplant Hepatology Fellowship Program

Robert H. Squires Jr., MD
Professor of Pediatrics
Medical Director, Liver Transplant Program

Arvind Srinath, MD
Assistant Professor of Pediatrics
Codirector, Pediatric Gastroenterology Fellowship Program

Veena Venkat, MD
Associate Professor of Pediatrics

Xunjun Xiao, PhD
Research Assistant Professor (until March 31, 2017)

OVERVIEW OF DIVISION

Over the past year, the division has experienced changes and improvements.

PERSONNEL CHANGES

- Divisional leadership changed at the end of March 2017, when Mark Lowe moved to St. Louis, Mo., and David Keljo took over as interim chief.
- New physician arrivals included the following.
  - Wednesday Sevilla moved from Memphis, Tenn., and is participating in the care of children with intestinal failure.
  - Sandra Kim moved from Nationwide Children's Hospital in Columbus, Ohio, to become the codirector of the IBD Center.

PROGRAM DEVELOPMENT

- Under the leadership of Jeffrey Rudolph and in its seventh year, the Intestinal Care Service continued to increase its patient care activities.
- The EPC, led by Mark Lowe and Sohail Husain, increased its patient numbers and participated in the evaluation and care of children undergoing total pancreatectomy and islet cell auto-transplantation for chronic pancreatitis.
- Under the leadership of Patrick McKiernan and Robert Squires, the inpatient hepatology service now includes joint rounds with Transplant Surgery on patients with liver or small-bowel transplants.
- The IBD Program, codirected by David Keljo and Sandra Kim, cares for almost 1,000 children with Crohn's disease or ulcerative colitis. The center is joining the Improve Care Now initiative and developing a Transition to Adulthood Program in collaboration with the IBD Center at Presbyterian Hospital of UPMC.
- Under the leadership of Arvind Srinath and Riha Bhatt, the fellowship program remains strong and continues to attract applicants who are competitive for the major training programs. Two of the fellows were supported on National Institutes of Health (NIH) T32 training grants.

CLINICAL ACTIVITIES

The following table shows data for the clinical activities of the division. Inpatient visits increased by more than 40%, driven by the hepatology consult service. The outpatient patient volume and procedure volume have been relatively constant, with variations attributable to the number of clinicians. The division staff sees outpatients in nine locations. The total number of outpatient visits was second among all the subspecialty divisions (excluding Emergency Medicine) of the Department of Pediatrics. The division staffs three inpatient services: the general GI service, the intestinal failure service, and the newly launched hepatology consult service.

PATIENT ACTIVITY BY YEAR AND DATE

Activity	Fiscal Year (FY)12	FY13	FY14	FY15	FY16	FY17
Outpatients	17,046	18,535	15,478	16,830	17,622	16,023
Inpatients	5,508	6,699	5,865	6,572	6,899	9,313
Procedures	4,279	4,381	4,045	4,264	4,439	4,337

Because of outstanding clinical and research activities, the division was ranked #5 in pediatric gastroenterology and surgery in the recent *U.S. News & World Report* ratings.

RESEARCH OVERVIEW

Research remains an important facet of the division; four faculty members have NIH funding, and the division continues to attract new research funding from the NIH and other organizations. It participates in multiple national studies for liver disease, pancreatic disease, and IBD.

LABORATORY-BASED RESEARCH

The research laboratories made major advances in the most recent academic year, and they have an even brighter future ahead with an influx of talented researchers.

- Sohail Z. Husain, MD, founded and directs the EPC. Members of the EPC include John F. Eisses, MD, PhD; Amitava Mukherjee, PhD; Kristy Boggs, PhD; and postdoctoral students Li Wen, MD, PhD, and Nayyar Ahmed, PhD. The EPC laboratory manager is Tanveer Javed. This year, the team welcomed a visiting scholar from Chengdu, China: Lihui Deng, MD. The EPC hosted three summer students in 2017. Under Husain's mentorship, five members wrote grants. Last year, EPC members were awarded an R01 competitive renewal (to Husain), a startup grant from the Children's Hospital of Pittsburgh Foundation (to Boggs), and a mid-sized pharmaceutical grant (to Husain).

- Arjumand Ghazi, PhD, and her laboratory joined the division. The Ghazi Laboratory studies the genetics of aging, reproduction, and lipid metabolism using the model system *Caenorhabditis elegans*. The following projects are the focus of the laboratory's research: balancing lipogenesis and lipolysis in health and aging, the role of TCER-1/TCERG1 in immunity and reproductive health, and the role of germline-intrinsic meiotic genes in somatic aging. The Ghazi Laboratory includes postdoctoral students Francis RG Armit, PhD, and Hyeljin Hwang, PhD; graduate students Julia Loose and Nikki Naim; and undergraduate student Thayjas Patil.

CLINICAL AND TRANSLATIONAL RESEARCH

Twenty clinical and clinical/translational research activities are under way in hepatology, IBD, and pancreatic diseases. Eleven pharmaceutical trials are going on within the division.

CHALLENGES

Clinical: The clinical productivity of the division is at capacity and is not adequate to meet the health system mandate of same-day scheduling. The division is addressing this by hiring more advanced practice providers (APPs), so that physicians can focus most of their energies on the most medically advanced aspects of their subspecialty training. To facilitate recruitment and optimize performance and retention of APPs, Srinath and nurse practitioner Leslie Coda are developing a curriculum for a gastroenterology APP fellowship program.

Research: Research is the future of medicine. Research funding is increasingly difficult to obtain. Eisses and Khan have promising K award applications submitted. Transitioning fellows to K awards has not been successful for a few years. One MD/PhD fellowship candidate has been accepted to the fellowship program, and another has been recruited for the combined pediatric residency/GI physician-scientist training program. Two NIH-funded researchers have been recruited for next year. Two fellows are developing new clinical research programs in IBD. Efforts are being made to participate in a larger number of pharmaceutical studies, particularly in IBD.

RESEARCH AND OTHER SCHOLARLY ACTIVITIES

David Keljo, MD, PhD

David Keljo began serving as interim division chief on April 1, 2017.

RESEARCH

David Keljo's research focuses on clinical aspects of pediatric IBD and involves collaborative arrangements with multiple centers.

Sex Differences in Statural Growth Impairment in Pediatric Crohn's Disease. Growth impairment is common in pediatric Crohn's disease and appears to be more marked in males than in females. This study aims to identify the causes of that discrepancy.

Risk Stratification and Identification of Immunogenetic and Microbial Markers of Rapid Disease Progression in Children With Crohn's Disease. An inception cohort study is gathering clinical, genetic, serologic, tissue, and microbiologic samples to try to predict which patients will have rapid progression to stricture or perforation. Enrollment is complete, and 36-month follow-up has been completed. The primary outcome paper has been published. Follow-up continues.

Predicting Response to Standardized Pediatric Colitis Therapy: The PROTECT Study. This multicenter (25 centers), open-label study funded by the National Institute of Diabetes and Digestive and Kidney Diseases (NIDDK) is designed to evaluate the safety and efficacy of standardized initial therapy using either mesalamine or corticosteroids then mesalamine for the treatment of children and adolescents newly diagnosed with ulcerative colitis. The study investigates whether response to the initial four weeks of therapy combined with clinical, genetic, and immune parameters determined during the initial course of therapy will predict severe disease as reflected by need for escalation of medical therapy or surgery. Enrollment and follow-up are complete for this study, and Pittsburgh is one of the top six enrolling centers. The primary outcome paper has been accepted for publication.

A Multidisciplinary Human Study on the Genetic, Environmental, and Microbial Interactions That Cause IBD: The GEM Study. This multicenter study sponsored by the Crohn's and Colitis Foundation of Canada aims to recruit unaffected siblings and offspring of patients with Crohn's disease. Siblings and offspring are 100-fold more likely to develop Crohn's disease than the rest of the population. The study will prospectively measure environmental exposures, changes in enteric microbial flora, and changes in immune responses in relation to the barrier function of the intestine and genetic makeup of these individuals to identify the changes that determine who develops Crohn's disease. Enrollment is ongoing.

David Keljo, MD, PhD
Interim Division Chief, Gastroenterology, Hepatology, and Nutrition

ADVISORY COMMITTEE MEMBERSHIPS

- Information Systems Physician Advisory Committee, Children's Hospital of Pittsburgh of UPMC

PROFESSIONAL AFFILIATIONS/SOCIETY MEMBERSHIPS

- American Gastroenterological Association (AGA)
- North American Society for Pediatric Gastroenterology, Hepatology, and Nutrition (NASPGHAN)
- Crohn's and Colitis Foundation of America (CCFA)
- Alpha Omega Alpha

HONORS

- *Best Doctors in America*, Woodward/White, Inc.
- Best Doctors, *Pittsburgh Magazine*

Mark Lowe, MD, PhD

Mark Lowe vacated his position as division chief on March 30, 2017.

RESEARCH

Molecular Mechanisms of Dietary Fat Digestion by Pancreatic Lipases. The long-term goal of Lowe's research is to elucidate the molecular mechanisms of dietary fat digestion by pancreatic lipases. This project focuses on dietary fat digestion by lipases in human newborns. The results will advance knowledge of dietary fat digestion by pancreatic lipases and facilitate bench-to-bedside development of novel nutritional therapies, such as more effective formulas or enzyme replacement that will transform care and improve outcomes of premature infants and critically ill newborns.

Proteotoxicity in the Pathophysiology of Chronic Pancreatitis. This project focuses on a novel mechanism for pancreatitis. Herein, the researchers address the hypothesis that carboxyl ester lipase (CEL) mutants associated with chronic pancreatitis activate adaptive cell signaling pathways and increase susceptibility of cells to injury by metabolic stress. The knowledge gained by the proposed studies will improve the overall understanding of pancreatic injury and provide insight into potential pharmacological interventions directed at a new therapeutic target, protein homeostasis.

International Study Group of Pediatric Pancreatitis: In Search for a Cure 2 (INSPPIRE2) to Study Chronic Pancreatitis. The objective of this application is to determine the natural history of pediatric chronic pancreatitis, identify risk factors and genetic modifiers for its onset and sequelae, and develop approaches to improve clinical outcomes. The long-term goal is to develop diagnostic modalities, prognostic factors, and better treatment approaches for pediatric chronic pancreatitis.

STUDY SECTIONS

- Special Emphasis Panel for the Extramural Loan Repayment Program, NIDDK, NIH
- NIH Special Emphasis Panel for Silvio O. Conte Digestive Diseases Research Core Centers, NIDDK, NIH

ADVISORY COMMITTEE MEMBERSHIPS

- Board of Directors, Western Pennsylvania Chapter of the National Pancreas Foundation
- Board of Directors, National Pancreas Foundation
- Fellow and Resident Leadership Initiative, Department of Pediatrics, University of Pittsburgh
- Ambulatory Care Steering Committee, Children's Hospital of Pittsburgh of UPMC
- Executive Committee, Department of Pediatrics, University of Pittsburgh
- Perioperative Committee, Children's Hospital of Pittsburgh of UPMC
- Resident Performance and Evaluation Committee, Department of Pediatrics, University of Pittsburgh
- Research Task Force, Department of Pediatrics, University of Pittsburgh
- Graduate Medical Education Full Committee, University of Pittsburgh School of Medicine
- Planning Committee, PancreasFest
- Promotions Committee, Department of Pediatrics, University of Pittsburgh
- Visit Coordination Program, Children's Hospital of Pittsburgh of UPMC
- Finance Committee, NASPGHAN
- Strategic Planning Committee, Children's Hospital of Pittsburgh of UPMC
- Pediatric Intern Selection Committee, Department of Pediatrics, University of Pittsburgh
- Center for Rare Disease Therapy, Children's Hospital of Pittsburgh of UPMC

EDITORSHIPS

- Associate editor, *Journal of Pediatric Gastroenterology and Nutrition*

HONORS

- *Best Doctors in America*, Woodward/White, Inc.
- Best Doctors, *Pittsburgh Magazine*
- Carol Ann Kraumer Endowed Chair for Pediatric Research

MAJOR LECTURESHIPS AND SEMINARS

- "Assessing Acinar and Duct Cell Function: Gaps and Opportunities," Chronic Pancreatitis in the 21st Century, NIH/NIDDK Conference, Pittsburgh, Pa., 2016
- "Steatorrhea: What If It's Not Cystic Fibrosis?" World Congress of Pediatric Gastroenterology, Hepatology, and Nutrition, Montreal, Canada, 2016
- "Pancreatitis," learning lunch, World Congress of Pediatric Gastroenterology, Hepatology, and Nutrition, Montreal, Canada, 2016
- "Recurrent and Chronic Pancreatitis: Natural History, Prevention, and Treatment in Pediatrics," American Pancreatic Association annual meeting, Boston, Mass., 2016
- "Pancreatic Lipase-Related Protein 2: From the Lab to the Nursery," Infant Brain annual meeting, Elsinore, Denmark, 2016
- "Recent Advances in Pediatric Acute Pancreatitis," Lviv Children's Hospital Conference, Lviv, Ukraine, 2017
- "Nutrition and Pancreatitis: What To Do When?" fellows symposium, National Pancreas Foundation, Grapevine, Texas, 2017
- "Mechanisms of Lipase Mutation in Chronic Pancreatitis," European Pancreas Club, Budapest, Hungary, 2017

PROFESSIONAL AFFILIATIONS/SOCIETY MEMBERSHIPS

- American Pancreatic Association
- American Pediatric Society
- NASPGHAN
- American Association for the Advancement of Science
- AGA
- Society for Pediatric Research
- American Society for Biochemistry and Molecular Biology

Feras Alissa, MD

Feras Alissa's research includes examining the dose-effect relationship of methotrexate and hepatic fibrosis in patients with IBD.

PROFESSIONAL AFFILIATIONS/SOCIETY MEMBERSHIPS

- NASPGHAN

MAJOR LECTURESHIPS AND SEMINARS

- "Line Dancing: Learning How to Obtain Venous Access, Maintain It, and Minimize Complications. Is Loss of Access the Ultimate Failure? And How to Overcome It," Ninth International Pediatric Intestinal Failure and Rehabilitation Symposium, Los Angeles, Calif., 2016

Riha Bhatt, MD

Riha Bhatt is a sub-investigator in several studies within the division.

ADVISORY COMMITTEE MEMBERSHIPS

- Associate program director, Children's Hospital of Pittsburgh Pediatric Gastroenterology Fellowship Program
- Education Committee, NASPGHAN
- Curriculum Committee, University of Pittsburgh School of Medicine
- Codirector of Theme Subcommittee, University of Pittsburgh School of Medicine
- Pediatric Gastroenterology Fellow Clinical Competency Committee, Children's Hospital of Pittsburgh
- Pediatric Gastroenterology Fellow Selection Committee, Children's Hospital of Pittsburgh

PROFESSIONAL AFFILIATIONS/SOCIETY MEMBERSHIPS

- NASPGHAN
- AGA
- AGA Academy of Educators
- Medical Education Research Certificate, Association of American Medical Colleges

Kristy Boggs, PhD

RESEARCH

Kristy Boggs' research focuses on defining the genomic landscape of pancreatic recovery and regeneration after injury. Through the utilization of next-generation sequencing protocols and molecular biology techniques, her aim is to characterize the transcriptome, genome, and epigenome of the pancreas following pancreatic injury. The exploitation of pancreatic recovery mechanisms may offer therapeutic targets for the treatment of pancreatitis.

Utilizing the murine caerulein hyperstimulation model of mild acute pancreatitis and recovery, she has sequenced the transcriptome of the non-injured and the histologically recovered pancreas by RNA-sequencing (RNA-seq). The RNA-seq data clearly demonstrate differentially expressed genes (DEGs) between the non-injured and the recovered pancreas, although histologically, the pancreas resembles that of the non-injured pancreas. This work has resulted in the preparation and submission to *Scientific Reports* of a manuscript titled "Pancreatic Gene Expression During Recovery After Pancreatitis Reveals Unique Transcriptome Profiles." The manuscript received favorable reviews and following revisions has been re-submitted to *Scientific Reports* (September 2017).

Whether the DEGs identified in the recovered pancreas allow the pancreas to thwart repeated events of stress or injury is currently under investigation. Boggs is assessing whether subsequent attacks of acute pancreatitis in mice are less severe than the initial attack, utilizing the caerulein hyperstimulation model of mild acute pancreatitis and recovery. The severity of recurrent acute pancreatitis is being determined both biochemically and histologically.

In addition to the identification of DEGs in the recovered pancreas, there is the possibility of poised chromatin states in the recovered pancreas. In this case, the gene is not expressed in the recovered pancreas, as determined by RNA-seq, but the chromatin structure is in a transcriptionally permissive state that is primed for rapid gene expression. An altered chromatin state in the recovered pancreas may confer a molecular memory that regulates severity of future organ injury. In future studies, Boggs will assess chromatin accessibility in the recovered pancreas by ATAC-seq (assay for transposase-accessible chromatin sequencing), with a specific focus on identifying poised regions in the chromatin of a recovered pancreas.

Also, in future work, she will determine the epigenetic mechanism(s), focusing on histone modifications, that induce a differential chromatin signature in the recovered

pancreas. Differentially modified histones between the non-injured and recovered pancreas will be assessed by ChIP-seq (chromatin immunoprecipitation and DNA sequencing) to identify specific post-transcriptional modifications of histone tails and the region of DNA associated with these transcriptionally active or repressive modifications. The identification of epigenetic modifications in the recovered pancreas will provided insight into the identified altered gene expression and chromatin structure in the recovered pancreas.

Boggs is affiliated with the Health Sciences Sequencing Core at Children's Hospital of Pittsburgh. She contributes to investigations of the Husain Laboratory and the larger scientific community by developing and optimizing next-generation sequencing technologies.

HONORS AND AWARDS

- Children's Hospital of Pittsburgh of UPMC Research Advisory Committee Grant, "Pancreatic Injury Induces a Unique Chromatin Footprint That Protects the Recovering Pancreas Against Repetitive Injury," 100% effort, $80,000 direct funds over two years, 2017–2019

Maria I. Clavell, MD

PROFESSIONAL AFFILIATIONS/SOCIETY MEMBERSHIPS

- American Society for Parenteral and Enteral Nutrition
- NASPGHAN

John F. Eisses, MD, PhD

RESEARCH

John Eisses' research focuses on understanding the role of epigenetic modifiers in regulating injury and recovery of the pancreas as it relates to pancreatic disease. He studies epigenetic regulation of pancreatitis in distinct cell types within the pancreas. Pancreatitis is a life-threatening inflammatory disorder that lacks targeted therapies. Injury and inflammation activate pancreatic stellate cells (PSCs), resulting in the remodeling of the pancreatic microenvironment, which allows pancreatic recovery to occur. Prolonged injury or aberrant regulation of PSCs has been implicated in the development or promotion of several pancreatic diseases, including chronic pancreatitis and pancreatic cancer. The overall goal of the current proposal is to examine novel epigenetic mechanisms by which pancreatic recovery is regulated in response to injury, particularly in the context of PSCs. The hypothesis is that the epigenetic regulation of gene expression in PSCs is crucial for activation and repression of gene expression necessary to promote a microenvironment suitable for pancreatic regeneration. The research is significant because it elucidates for the first time an epigenetic mechanism for regenerative signals important in regulating the pancreatic microenvironment to allow pancreatic recovery. On a much broader level, it opens a new paradigm of possible therapeutic strategies that may enhance the ability of the pancreas to recover, particularly in the context of remodeling the extracellular matrix and the pancreatic microenvironment by PSCs.

ADVISORY COMMITTEE MEMBERSHIPS

- Member, Pancreas Committee, NASPGHAN
- Member, Pediatric Pancreatology Consortium, INSPPIRE

EDITORSHIPS

- Associate editor, *GI Scholarly Newsletter*, Children's Hospital of Pittsburgh

PROFESSIONAL AFFILIATIONS/SOCIETY MEMBERSHIPS

- NASPGHAN
- European Pancreas Club
- AGA
- American Pancreatic Association
- Pediatric Pancreatology Consortium, INSPPIRE
- Collaborative Alliance for Pancreatic Education and Research

Tamara Feliciano Alvarado, MD

PROFESSIONAL AFFILIATIONS/SOCIETY MEMBERSHIPS

- NASPGHAN

Arjumand Ghazi, PhD

RESEARCH

The Ghazi Laboratory studies the genetics of aging, reproduction, and lipid metabolism using the model system *Caenorhabditis elegans*. The following projects were undertaken in the past year and are currently the focus of the laboratory's research.

Balancing Lipogenesis and Lipolysis in Health and Aging. Lipid imbalances are characteristic of obesity and a feature of many age-related ailments and reproductive pathologies in humans. Yet the relationship among lipid metabolism, aging, and reproduction remains poorly studied. This project investigates how the balance between lipid synthesis and breakdown influences an organism's rate of aging and health. In *C. elegans*, eliminating the germline extends lifespan, removes fertility, and is a major challenge to lipid metabolism because the animal needs to stop fat deposition into eggs and reorganize its lipid profile. Thus, germline-less worms provide a unique platform to understand how lipid balance is established in the cells and tissues of complex multicellular animals in the

face of major physiological changes. We discovered that fat production and degradation increase simultaneously in response to germline loss. In germline-less adults, a conserved transcription factor, NHR-49, upregulates fatty-acid β-oxidation and desaturation—processes that contribute to lipid breakdown and build-up, respectively. These two, and many other processes involved in lipid synthesis and degradation, are elevated in response to germline removal by the conserved transcription factors DAF-16 and TCER-1. These data suggest that the coordinated enhancement of lipid synthesis and breakdown facilitate adaptation to germline loss by mediating lipid homeostasis. The knowledge obtained from these studies is likely to reveal fundamental insights into obesity and the relationships among lipid metabolism, reproduction, and aging.

Role of TCER-1/TCERG1 in Immunity and Reproductive Health. Reproduction and stress resistance in metazoans are poorly described. We discovered a role for TCER-1, the worm homolog of human transcription elongation, and splicing factor TCERG1 in enhancing reproductive capacity in the face of pathogen attack. In *C. elegans*, eliminating germline stem cells increases lifespan and elevates stress resistance. Previously, we demonstrated that TCER-1/TCERG1 specifically promotes the longevity of germline-less *C. elegans* but is critical for reproductive health in normal, fertile animals. Surprisingly, we have discovered that TCER-1/TCERG1 inhibited stress tolerance in adult worms. *tcer-1* mutants exhibited elevated resistance against multiple biotic and abiotic stressors. TCER-1/TCERG1 impairs stress resistance by inhibiting PMK-1, a conserved innate immunity-promoting kinase. PMK-1-target genes are upregulated in *tcer-1* mutants, and the immunoresistance of *tcer-1* mutants is dependent upon PMK-1. The data suggest that TCER-1/TCERG1 promotes reproductive fitness and represses stress resilience under normal conditions. Under stressful conditions, TCER-1/TCERG1 is repressed, resulting in enhanced stress resilience and reduced reproductive capacity. Unlike most pro-longevity genes, TCER-1/TCERG1 appears to have distinct regulatory effects on lifespan, stress resistance, and fertility, suggesting that the protein may function as a molecular rheostat for coordinating major life-history traits. The phenotypic uncoupling observed in *tcer-1* mutants provides a unique platform to dissect the pathway that governs resource allocation among procreation, stress response, and somatic maintenance in metazoans.

Role of Germline-Intrinsic Meiotic Genes in Somatic Aging. Meiotic chromosomal defects increase dramatically with age and are a major cause of miscarriages, birth defects, and age-related fertility loss in women. Correlative evidence from human studies has indicated that germline fidelity has a role in overall health. This research used the nematode *C. elegans* to address the mechanisms of and cause-effect relationships between germline health and somatic aging. Specifically, the researchers have focused on genes involved in *C. elegans* meiosis, an intrinsically germline-restricted phenomenon that is fundamental to genetic diversity. They have found that mutations in multiple genes that act at different stages of worm meiosis cause lifespan reduction and demonstrate signs of premature aging. Besides lifespan, the researchers have examined the effects of these mutations directly on the rate of organismal aging by testing various measures of healthspan. Preliminary results suggest that meiotic mutants not only exhibit reduced lifespans but also show physiological and molecular signs of accelerated aging. The data suggest that genes that govern meiotic fidelity in germ cells of *C. elegans* impact the rate of aging of the whole animal.

ADVISORY COMMITTEE MEMBERSHIPS

- Chair, Admissions Committee, Interdisciplinary Biomedical Graduate Program, 2016–2017
- Member, Admissions Committee, Interdisciplinary Biomedical Graduate Program, 2017–2018

EDITORSHIPS

- *Scientific Reports*
- *Frontiers in Endocrinology and Aging*

HONORS

- Invited to participate in the National Institute on Aging, NIH, Division of Aging Biology New Investigators Forum, July 2017
- Publication recommended by Faculty of 1000 (Amrit et al., *PLoS Genetics*, 2014)

MAJOR LECTURESHIPS AND SEMINARS

- "Finding Human Longevity Genes in a Spineless Worm," Honors College Health Sciences Undergraduate Summer Fellowship Program, Pittsburgh, Pa., July 2016
- "Aging in Worms … Clinical Relevance?" Breakfast with Mentors, Summer Research Internship Program, Children's Hospital of Pittsburgh, Pittsburgh, Pa., July 2016
- "Concomitant Modulation of Somatic Lipid Production and Breakdown in Response to Germline Signals," Molecular Genetics of Aging meeting, Cold Spring Harbor Laboratory, Cold Spring Harbor, N.Y., September 2016
- "Genes That Link Fat, Fertility, and Aging," Long-Life Family Study Group, Department of Epidemiology, University of Pittsburgh, Pittsburgh, Pa., November 2016

- "Lipid Metabolism and Aging," Division of Endocrinology, Department of Pediatrics, University of Pittsburgh, Pittsburgh, Pa., April 2017
- "Concomitant Modulation of Somatic Lipid Production and Breakdown in Response to Germline Signals," second Interventions in Aging meeting, Cancun, Mexico, March 2017
- "Fat, Fertility, and Aging in *C. elegans*," featured speaker for conference held once every four years, 18th International Society for Developmental Biology meeting, National University, Singapore, June 2017

Sohail Z. Husain, MD

RESEARCH

The overarching goal of Sohail Husain's research is to determine the mechanisms that initiate and propagate pancreatitis, a painful inflammatory disease that accounts for more than 300,000 hospitalizations and 3,200 deaths in the United States annually. Specifically, current advances in the Husain Laboratory are focused on: (1) examining the aberrant calcium signals and calcium targets within the main pancreatic cell, the acinar cell, which are critical to both the initiation and propagation of pancreatitis; (2) determining the role of epigenetic factors in pancreatic recovery and regeneration after injury; and (3) deciphering the mechanisms underlying drug-induced pancreatitis. Husain's work is anticipated to lead to effective treatment or preventive strategies for pancreatitis that target aberrant calcium signaling, enhance pancreatic recovery, or compensate for toxic drug exposures.

STUDY SECTIONS

- Standing member, Clinical, Integrative, and Molecular Gastroenterology Study Section, NIH
- Section member, Peer-Reviewed Medical Research Program, U.S. Department of Defense
- Standing member, Research Advisory Committee, Children's Hospital of Pittsburgh of UPMC

ADVISORY COMMITTEE MEMBERSHIPS

- Chair, Pancreatitis: Inflammation, Fibrogenesis, and Immunology Review Section, AGA
- Chair, AGA Abstract Review for Pancreatitis: Inflammation, Fibrogenesis, and Immunology
- Chair, Pancreas Committee, NASPGHAN
- Councilor, Pancreatic Disorders Section, AGA Institute Council
- Organizing member, National Pancreas Foundation, Western Pennsylvania Chapter
- Site principal investigator, INSPIRRE2: Pediatric Acute and Chronic Pancreatitis Multi-Center, NIH-funded, R01 Consortium

EDITORSHIPS

- Editorial Board, *Pancreapedia*
- Editorial Board, *Journal of Biological Chemistry*
- Editor-in-chief, *GI Scholarly Newsletter*, Children's Hospital of Pittsburgh

MAJOR LECTURESHIPS AND SEMINARS

National/International:

- "Targeting the Calcium Effector Calcineurin in Pancreatitis," invited lecture, APA annual meeting, San Diego, Calif., 2017
- "Why Do Some Drugs Cause Pancreatitis?" invited lecture, Frontiers in Pediatric Pancreatology, NASPGHAN Single Topic Symposium, Las Vegas, Nev., 2017
- "Turning Investigational Therapies into Clinical Reality: Pancreatitis in the Spotlight," visiting professor, pediatrics seminar (host: Teresa Quattrin), University at Buffalo, Buffalo, N.Y., 2017
- "Epithelial Cell Calcineurin Signaling in Pancreatitis," visiting professor, Mayo Clinic Cancer Seminar Series (host: Baoan Ji), Mayo Clinic, Jacksonville, Fla., 2017
- "Pancreatitis," "Pancreatic Anatomy and Anomalies," and "Hereditary Disorders of the Pancreas," invited lectures, NASPGHAN Essentials Pediatric GI Review (course director: Chris Liacouras), Scottsdale, Ariz., 2017

Regional/Local:

- "Advances in Preventing Radiocontrast-Induced ERCP Pancreatitis by Targeting Calcium and Calcineurin," Molecular Medicine Research Seminar (host: Jay Kolls), Pittsburgh, Pa., 2016
- "Preventing Post-ERCP Pancreatitis by Targeting Calcineurin," faculty lecture, internal medicine gastroenterology grand rounds, Pittsburgh, Pa., 2016
- "Getting Published," faculty lecture, Unified Fellows Conference, Pittsburgh, Pa., 2016

PROFESSIONAL AFFILIATIONS/SOCIETY MEMBERSHIPS

- NASPGHAN
- American Pancreatic Association
- AGA
- INSPPIRE
- National Pancreas Foundation

HONORS

- *Best Doctors in America*, Woodward/White, Inc.
- Best Doctors, *Pittsburgh Magazine*

Zahida Khan, MD, PhD

RESEARCH

Zahida Khan took a sabbatical absence the academic year of 2016–2017 to complete fellowship training in advanced pediatric transplant hepatology at Texas Children's Hospital through Baylor University. Khan completed her advanced pediatric transplant hepatology fellowship in August of 2017. She then returned to Children's Hospital of Pittsburgh of UPMC and the University of Pittsburgh School of Medicine, where she continues to provide her services clinically and research liver diseases, focusing on cellular approaches to liver regeneration and transplantation.

More than 15,000 patients are awaiting liver transplantation, but only 6,729 liver transplants were performed last year. Alpha-1 antitrypsin deficiency (ATD) is the most common hereditary liver disease in children, and it is the most frequent genetic reason for liver transplantation. In classical ATD, the PiZ mutation results in misfolded ATZ protein monomers, which cause proteotoxicity, often leading to chronic liver injury, cirrhosis, and cancer. Not all patients develop severe liver disease, and it is unclear why only some patients need a liver transplant. Khan's research investigates the basic cell biology behind this question, using the PiZ transgenic mouse model. To understand how liver regeneration is impaired in ATD, she is studying intracellular mechanisms of protein processing and cell proliferation in PiZ hepatocytes. She is also studying how bile acid signaling can be utilized as a therapeutic target for ATD-related liver disease. Her research investigates largely unexplored pharmacologic pathways in ATD.

PROFESSIONAL AFFILIATIONS/SOCIETY MEMBERSHIPS

- Hepatology Committee, NASPGHAN Research Awards Committee, American Association for the Study of Liver Diseases (AASLD)

HONORS

- Research Presentation Award (Top Abstract of 144), Baylor College of Medicine, Texas Children's Hospital Pediatric Research Symposium, March 2017

MAJOR LECTURESHIPS

- "Bile Duct Ligation Induces ATZ Globule Clearance in a Mouse Model of Alpha-1 Antitrypsin Deficiency," NIH Child Health Research Center annual retreat, Bethesda, Md., September 2016
- "Liver Cell Regeneration in Alpha-1 Antitrypsin Deficiency," Alpha-1 Foundation investigators' meeting, Miami, Fla., October 2016

Sandra C. Kim, MD

RESEARCH

Sandra Kim focuses on clinical care of children with IBD. She has research expertise in basic, translational, and clinical/quality-improvement initiatives in pediatric and adolescent IBD. She is involved in clinical research and quality-improvement initiatives on regional and national levels for children and adolescents with IBD and has authored numerous studies on pediatric and adolescent IBD. She is a member of the Children's Hospital of Pittsburgh's Fellowship Clinical Competency Committee for the Pediatric Gastroenterology Program.

ADVISORY COMMITTEE MEMBERSHIPS

- Crohn's and Colitis Foundation
  - Co-chair, Government/Industry Affairs and Advocacy, National Scientific Advisory Committee
  - Member and past chair, Pediatric Affairs Committee, National Scientific Advisory Committee
- ImproveCareNow (ICN) Pediatric IBD Quality-Improvement Collaborative
  - Chair, Clinical Practice Committee
  - Co-chair, Transition of Care Innovation Community
  - Member, Physician Leadership Group
  - Member, Strategy Council
  - Member, Collaborative Steering Committee, ICN/Crohn's and Colitis Foundation

- NASPGHAN
  - Member, IBD Committee
  - Faculty, NASPGHAN First Year Fellows' Conference
  - NASPGHAN Fellows' Mentoring Program
  - Member, Home Infusion Task Force

PROFESSIONAL AFFILIATIONS/SOCIETY MEMBERSHIPS

- AGA
- Crohn's and Colitis Foundation
- NASPGHAN
- Alpha Omega Alpha
- Gold Humanism Society

HONORS

- University of Pittsburgh School of Medicine Advanced Faculty Leadership Academy, 2017
- *Best Doctors in America*, Woodward/White, Inc., 2015–2017

MAJOR LECTURESHIPS AND SEMINARS

- "Biosimilars in IBD: What Does the Pediatric Gastroenterologist Need to Know?" invited speaker, NASPGHAN annual meeting, November 2017
- "IBD 2020 at CHP: What Does the Future Hold?" Department of Pediatrics grand rounds, Children's Hospital of Pittsburgh of UPMC, Pittsburgh, Pa., October 2017
- "Rheumatologic Issues in Pediatric IBD," and "How Do We Discuss Surgery With Our Pediatric Patients With IBD?" invited speaker, CCFA Advances in IBD, Orlando, Fla., December 2016
- "Diet and IBD: Food for Thought," invited speaker, World Congress for Pediatric Gastroenterology, Hepatology, and Nutrition, postgraduate course, Montreal, Canada, October 2016

Dale E. King, MD

RESEARCH

Dale King participates in several clinical research projects in the department, including studies related to IBD, intestinal failure, parenteral lipid supplements (Omegaven and SMOFlipid), and bowel-preparation regimens.

PROFESSIONAL AFFILIATIONS/SOCIETY MEMBERSHIPS

- NASPGHAN
- AGA
- CCFA

Douglas Lindblad, MD

RESEARCH

Douglas Lindblad is involved with a research proposal (Cystic Fibrosis Foundation) to study the relationship between cystic fibrosis–associated liver disease and cystic fibrosis–related diabetes.

PROFESSIONAL AFFILIATIONS/SOCIETY MEMBERSHIPS

- American Academy of Pediatrics

Patrick J. McKiernan

RESEARCH

Patrick McKiernan's research interests cover the spectrum of pediatric liver disease but have fallen into three main areas.

- Liver transplantation: His published contributions span clinical outcomes, pharmacokinetics of immunosuppressants, long-term histological outcomes, and guidelines for transition to adult care.
- Portal hypertension: He has published on the use of endoscopic techniques, including variceal band ligation and endoscopic ultrasound, and he has contributed to the development of international guidelines.
- Metabolic liver disease: He has published on the clinical management of metabolic disease, especially tyrosinemia type I, and the evolving role of liver transplantation in metabolic liver disease. He has been involved in clinical trials of stem cell treatment for metabolic disease.

In addition, McKiernan is the local principal investigator for the following first-in-humans multicenter trial: a phase I/II, randomized, open-label, ascending-dose, delayed-treatment concurrent control clinical study to evaluate the safety and preliminary efficacy of AT342, an AAV8-delivered gene transfer therapy in Crigler-Najjar syndrome subjects aged 1 year and older.

Furthermore, he is the local principal investigator for a five-year, multicenter, longitudinal observational study of patients with nonalcoholic fatty liver or nonalcoholic steatohepatitis.

ADVISORY COMMITTEE MEMBERSHIPS

- Center for Rare Disease Therapy, Children's Hospital of Pittsburgh of UPMC

MAJOR LECTURESHIPS AND SEMINARS

- "Liver Transplantation for Metabolic Disease," Society for the Study of Inborn Errors of Metabolism annual scientific meeting, Rome, Italy, September 2016

- "Mitochondrial Hepatopathies in Children: State-of-the-Art of Diagnosis and Treatment," American Association for Study of Liver Disease annual meeting, Boston, Mass., November 2016
- "Improving Outcomes for Metabolic Disease," International Pediatric Medical Congress, Dubai, United Arab Emirates, November 2016
- "Update in Pediatric Liver Disease," Community Liver Alliance, Pittsburgh, Pa., December 2016
- "Current Approach to Neonatal Cholestasis," European Society for Paediatric Gastroenterology, Hepatology, and Nutrition (ESPGHAN) postgraduate course, Prague, Czech Republic, May 2017
- "Liver Transplantation in Mitochondrial Disorders," ESPGHAN annual scientific meeting, Prague, Czech Republic, May 2017
- "Monitoring the Liver Over the Long Term: Structural Assessment," International Pediatric Transplant Association scientific meeting, Barcelona, Spain, May 2017

PROFESSIONAL AFFILIATIONS/SOCIETY MEMBERSHIPS

- ESPGHAN
- American Association for the Study of Liver Disease
- British Society of Paediatric Gastroenterology, Hepatology, and Nutrition
- British Association for the Study of the Liver
- Society for the Study of Inborn Errors of Metabolism
- British Inherited Metabolic Diseases Group
- Studies of Pediatric Liver Transplantation (SPLIT)

Amitava Mukherjee, PhD

RESEARCH

Amitava Mukherjee's overarching goal is to uncover key molecular signaling pathways for exocrine pancreatic disorders, particularly drug-induced pancreatitis. Currently, his research focuses on deciphering the mechanisms underlying asparaginase-associated pancreatitis. The research suggests that pancreatic asparagine synthetase (ASNS) maintains acinar cell homeostasis and that its upregulation is required to mitigate asparaginase-induced pancreatic cell injury. Therefore, therapies that selectively augment pancreatic ASNS could be used to alleviate asparaginase-associated pancreatitis. He has generated sufficient preliminary data for grant submissions. Mukherjee has published work in collaboration with the Mark Lowe Laboratory on a mutant lipase that is responsible for chronic pancreatitis in the *Journal of Biological Chemistry*. He has written a chapter for *Liver Disease in α1-Antitrypsin Deficiency*.

Mukherjee has been assisted by his research team, consisting of a postdoctoral fellow, Nayyar Ahmed, PhD; a medical fellow, Fateema Rose, MD; and a visiting researcher, Li-Hui Deng. Mukherjee has participated in training two summer students, Chaitanya Srinivasan and Mahedah Rehman.

HONORS

- PancreasFest Travel Scholarship Award: Collaborative Alliance for Pancreatic Education and Research, University of Pittsburgh, June 2017

Jeffrey Rudolph, MD

RESEARCH

Intestinal Care and Rehabilitation Center Database. This retrospective database is designed to collect data that describe the natural history and management practices of intestinal failure and rehabilitation at Children's Hospital.

Compassionate Use of Omegaven in the Treatment of Parenteral Nutrition–Associated Liver Disease. This is a compassionate-use protocol for the use of omega-3 fatty acids as an alternative lipid source in children with total parenteral nutrition–associated liver disease.

A Prospective, Randomized, Controlled, Double-Blind, Parallel-Group, Phase III Study to Compare the Safety and Efficacy of Smoflipid 20% to Intralipid 20% in Hospitalized Neonates and Infants Requiring 28 Days of Parenteral Nutrition. Jeffrey Rudolph is site investigator for a multicenter study comparing the effects of standard parenteral lipid therapy to a novel lipid emulsion.

Biomarkers in Transplant Recipients. This entails obtaining biological samples from intestinal transplant patients during routine endoscopy for assessment of markers that predict the immunological state or organ rejection.

ADVISORY COMMITTEE MEMBERSHIPS

- Codirector, Pediatric Intestinal Failure, Rehabilitation, and Transplant Symposium, Pittsburgh, Pa., 2018
- Nutrition Committee, NASPHAN

MAJOR LECTURESHIPS AND SEMINARS

- "Trace Elements in TPN," invited speaker, Sydra Medical Corporation, Doha, Qatar, April 2017

PROFESSIONAL AFFILIATIONS/SOCIETY MEMBERSHIPS

- NASPGHAN
- American Society for Parenteral and Enteral Nutrition

HONORS

- Best Doctors, *Pittsburgh Magazine*

Wednesday Marie A. Sevilla, MD, MPH, CNSC

RESEARCH

American Society for Parenteral and Enteral Nutrition New Opportunities for Verification of Enteral Tube Location Project. This project addresses the safety of feeding tube placement. Wednesday Marie A. Sevilla collaborates with the task force on a variety of projects, including development of a position paper on best practices for nasogastric tube placement.

Soluble Fiber Utilization in Pediatric Short Bowel Syndrome: A Survey of Current Practice Trends. This project is a collaboration with a team at Le Bonheur Children's Hospital. The data from the survey will provide an overview of the clinical practice of administering supplemental enteral fiber in pediatric patients with short bowel syndrome. Data have been collected and analyzed. A manuscript is being prepared for submission for publication. Sevilla participated in developing the survey tool, monitoring data collection and analysis, and authoring the manuscript.

ADVISORY COMMITTEE MEMBERSHIPS

- Member, Nutrition Advisory Group, Children's Hospital of Pittsburgh

PROFESSIONAL AFFILIATIONS/SOCIETY MEMBERSHIPS

- Nutrition Committee, NASPGHAN
- AGA
- Pediatrics Section and Pediatric Intestinal Failure Section, American Society for Parenteral and Enteral Nutrition
- Intestinal Transplant and Rehabilitation Association

Sapana Shah, MD

RESEARCH

Predicting Response to Standardized Pediatric Colitis Therapy: The PROTECT Study. This multicenter (25 centers), NIDDK-funded, open-label study is designed to evaluate the safety and efficacy of standardized initial therapy using either mesalamine or corticosteroids then mesalamine for the treatment of children and adolescents newly diagnosed with ulcerative colitis. The study will investigate the hypothesis that response to the initial four weeks of therapy as well as specific clinical, genetic, and immune parameters determined during the initial course of therapy will predict severe disease as reflected by need for escalation of medical therapy or surgery. Enrollment and follow-up are complete, and the Pittsburgh location is one of the top six enrolling centers. The study is in the data-analysis phase.

An Efficacy and Safety Study of Infliximab in Pediatric Participants With IBD. This multicenter study sponsored by Janssen Scientific Affairs, LLC, is designed to evaluate whether trough serum infliximab concentrations at the time of loss of clinical response will identify pediatric participants with IBD who would benefit from dose escalation above the currently approved dose of 5 mg/kg.

Improve Care Now. Improve Care Now is a multicenter, quality-improvement collaborative for patients with pediatric IBD that enables patients, families, clinicians, and researchers to improve knowledge and outcomes related to Crohn's disease and ulcerative colitis. It engages all stakeholders in a learning health network that provides real-time quality improvement, research, and community-building for children with these conditions.

PROFESSIONAL AFFILIATIONS/SOCIETY MEMBERSHIPS

- NASPGHAN

Leah Siebold, MD

RESEARCH

Leah Siebold is principal investigator for the following.

- PRO-KIIDS and NEOPICS Retrospective Cohort Study of Very Early Onset IBD
- The international Early Onset Paediatric IBD Cohort Study
- A Study of Fecal Microbiota Transplantation in Pediatric Patients With Relapsed IBD

ADVISORY COMMITTEE MEMBERSHIPS

- Clinical Care and Quality Committee, NASPGHAN Physical Leadership Team, Children's Hospital of Pittsburgh
- Physician Compensation Committee, Children's Hospital of Pittsburgh
- 8B GI liaison, Children's Hospital of Pittsburgh
- Improve Care Now
- Taskforce for Home Infusions, NASPGHAN

Amina Smajlovic, MD

PROFESSIONAL AFFILIATIONS/SOCIETY MEMBERSHIPS

- American Academy of Pediatrics

James E. Squires, MD, MS

RESEARCH AND OTHER SCHOLARLY ACTIVITIES

Potential of Human Induced Pluripotent Stem Cells (iPSCs) in Studies of Progressive Intrahepatic Cholestasis. The hepatocyte canalicular membrane serves as the gateway through which hepatocyte metabolic products pass to form bile.

Disruption of canalicular function, therefore, is expected to have detrimental effects on liver and human health. Autosomal recessive disorders are known to disrupt hepatocyte canalicular function and bile secretion. Progressive familial intrahepatic cholestasis (PFIC) is a group of rare diseases clinically manifested by intrahepatic cholestasis in children and progresses to end-stage liver disease and death or liver transplantation.

The ability to derive patient-specific iPSCs allows opportunities to model hepatic disorders, gain mechanistic insights, and explore cell-based therapies. Briefly, human fibroblasts or lymphocytes are isolated and reprogrammed into hepatocyte-like cells, which bear significant similarities to primary hepatocytes in terms of transcriptional profile and functional properties but retain the unique, patient-specific genetic defects. These patient-derived iPSC cell lines enable the study of individual phenotypes under highly controlled conditions and allow linking the observed defects directly to disease-causing genetic alterations. Liver-specific disease models include ATD, familial transthyretin amyloidosis, glycogen storage disease, Wilson's disease, familial hypercholesterolemia, and Niemann-Pick disease. The present study hypothesizes that patient-specific human iPSCs can be generated and accurately recapitulate the main characteristics of PFIC. Squires is principal investigator.

Graft Injury Group Observing Late-Term Outcomes. This is a multicenter, international collaboration of pathologists, hepatologists, and transplant surgeons looking to better understand and characterize late graft injury in children who receive liver transplants. Squires is site investigator.

Biomarkers of Acute Kidney Injury in Pediatric Liver Transplant Recipients. Acute kidney injury (AKI) is a rapid loss of kidney function, and AKI following liver transplant adversely affects outcomes. Improved detection and management of AKI in the perioperative period would have long-term benefits in reducing the development of chronic kidney disease. Serum creatinine, the current gold-standard marker for kidney injury, is a late and insensitive marker of AKI. Newer biomarkers such as urine IGFBP7, TIMP-2, and NGAL have proven to be superior in the timely detection of renal injury and impairment in critically ill adults, and the ability to sample urine is advantageous. This research will study novel biomarkers of AKI in the pediatric liver transplant population to determine their diagnostic and predictive capabilities relating to the development of AKI in pediatric populations. Squires is principal investigator.

Childhood Liver Disease Research Network. Cholestatic liver diseases are a major cause of morbidity and mortality in children. The most common of these diseases, biliary atresia, is the leading indication for liver transplantation in children. The cause(s), optimal diagnosis, and management of biliary atresia remain unknown. Thus, the NIH has funded a cooperative consortium to conduct a comprehensive study of biliary atresia. The Children's Hospital of Pittsburgh of UPMC is one of 10 clinical centers participating in this collaborative investigation. Squires is co-investigator.

SPLIT. This project is a community of pediatric hepatologists, transplant surgeons, research coordinators, nurse coordinators, and other health professionals across the United States and Canada working together to advance knowledge in pediatric liver transplantation. SPLIT was started in 1995 and has evolved from a research registry into a multifaceted organization focused on improving outcomes for children receiving liver transplantation. Squires is co-investigator.

Clinical Trials in Organ Transplantation in Children (CTOTC): Biomarkers for Post-Transplant Lymphoproliferative Disorders (PTLDs) in Children. This is a clinical trial investigating biomarkers associated with the development of Epstein-Barr virus (EBV)-related PTLDs in pediatric liver, heart, heart with liver, small intestine, and liver with small intestine transplant candidates and recipients. Squires is co-investigator.

ADVISORY COMMITTEE MEMBERSHIPS

- Idiopathic Neonatal Cholestasis Subcommittee, Childhood Liver Disease Research Network (ChiLDReN) Alpha-1-Antitrypsin Deficiency Subcommittee, ChiLDReN
- Mitochondrial Hepatopathy Subcommittee, ChiLDReN
- Education Committee, SPLIT Hepatology Committee, NASPGHAN
- American Board of Pediatric Intensive Care Unit Liver Board Review

EDITORSHIPS

- Associate editor, *GI Scholarly Newsletter*, Children's Hospital of Pittsburgh

HONORS

- Co-author, "Neurocognitive Status in Alagille Syndrome: Results of a Multicenter Prospective Observational Study," AASLD Presidential Poster of Distinction
- AASLD Young Investigators Travel Award for "Liver Transplantation in Pediatric Acute Liver Failure: Practices and Patient Characteristics"

MAJOR LECTURESHIPS AND SEMINARS

- "Liver Dysfunction in Patients With IBD," invited lecture, Cincinnati Children's Hospital Medical Center Pediatric Autoimmune Liver Disease Symposium, Cincinnati, Ohio, October 2017

- "Post-Transplant Lymphoproliferative Disease," International Transplant Nurses Society local chapter symposium, August 2017
- "Anticipating Gene Therapy for Crigler-Najjar Syndrome," invited lecture, Fifth Annual Translational Medicine in Plain Populations Conference, Pittsburgh, Pa., August 2017
- "PFIC: The Clinical Perspective and Modeling Using Patient-Derived iPS Cells," invited lecture, Pittsburgh Liver Research Center Pediatric Cholestatic Liver Disease Focus Group, Pittsburgh, Pa., June 2017
- "Potential of Human Induced Pluripotent Stem Cells in Studies of Progressive Intrahepatic Cholestasis," invited lecture, Pittsburgh Liver Research Center Seminar Lecture Series, Pittsburgh, Pa., March 2017

Robert H. Squires Jr., MD

RESEARCH

A Multicenter Group to Study Acute Liver Failure in Children (NIH/NIDDK). Acute liver failure is a rare condition in which loss of liver cell function occurs rapidly (often less than two weeks), with devastating results. Members of this consortium continue to publish, and important clinical advances have also been made. Ongoing projects utilizing stored biomedical samples coupled with detailed clinical data include: characterizing the role of $CD8^+$ T-cell infiltration and indeterminate pediatric acute liver failure, identifying HMGB1 as an important cytokine mediator and acetaminophen toxicity, characterizing markers of immune activation, identifying genetic causes of indeterminate pediatric acute liver failure, and utilizing biomedical samples for viral discovery. A few recent achievements include: characterization of neurodevelopmental deficits in children following acute liver failure, exploration of liver transplant decisions in the setting of acute liver failure, and identification of a dynamic model using clinical and biochemical markers associated with clinical outcomes. The study is currently in its final no-cost extension phase. Squires is principal investigator.

The Pittsburgh Cholestatic Liver Disease Consortium (NIH/NIDDK). This is a renewal application that is a continuation, expansion, and merging of the Biliary Atresia Research Consortium and the Cholestatic Liver Consortium to form the Childhood Liver Disease Research and Education Network (ChiLDREN). Following enrollment, detailed longitudinal clinical data are collected as well as biomedical samples. The research team is currently enrolling participants with chronic cholestatic conditions and analyzing patient populations. There are two associated clinical trials to assess the efficacy of an apical sodium–dependent bile acid transporter inhibitor in the treatment of cholestatic liver disease in children with progressive familial intrahepatic cholestasis (sponsor: Lumena Pharmaceuticals Inc.) and Alagille syndrome (sponsor: NIH/NIDDK). Squires is principal investigator.

Hepatitis B Clinical Research Network—Data Coordinating Center (NIH/NIDK). This application proposes a multicenter group to study chronic hepatitis B infection to examine natural history and immunopathogenesis. Squires serves as a funded pediatric advisor at the data coordinating center.

ATD Treated by Carbamazepine (CBZ). CBZ decreased the hepatic load of AT granules and hepatic fibrosis in the PiZ mouse model of ATD-associated liver disease. Squires is investigating the effect of CBZ on severe liver disease due to ATD in adolescents and adults. Sponsor: Perlmutter Laboratory. Squires is co-investigator.

Creating Models of Rare Childhood Liver Diseases Using the Human Liver-on-a-Chip. Researchers are focusing initially on *POLG* mutations that cause Alpers Huttenlocher syndrome, one of many rare childhood diseases that involve the liver. This study will expand to include mutations involving the canalicular membrane. Sponsor: NIH. Squires is co-investigator.

Hepatitis C Clinical Trial. The trial has enrollment and is currently monitoring three patients in an open-label, multicenter study to evaluate the pharmacokinetics, safety, and efficacy of glecaprevir/pibrentasvir in pediatric subjects with genotype 1–6 chronic hepatitis C virus infection. Sponsor: AbbVie Inc. Squires is site principal investigator.

Pediatric Intestinal Failure Consortium. Although funding for this project has ended, ongoing sub-analysis of the data continues. A recent publication by Javid et.al. highlighted that the extent of intestinal failure–related liver disease is associated with increased mortality. Sponsor: NIH/NIDDK. Squires is principal investigator.

ADVISORY COMMITTEE MEMBERSHIPS

- 2011-16 International Classification of Disease (ICD)-11 revision process, Pediatric Topic Advisory Group, Gastroenterology Working Group, World Health Organization
- Gastroenterology Project Team, "Best Children's Hospitals," *U.S. News & World Report*
- American Academy of Pediatrics
  - Pediatrics Review and Education Program GI Editorial Board

  - Expert learning community meeting on "Innovative Financing for Health Care Services for Children With Special Health Care Needs: Value-Based Purchasing," American Academy of Pediatrics (Chicago, Ill.) and the Catalyst Center (Boston, Mass.)
  - Subspecialty advisor, Executive Board, Pennsylvania Chapter
- AASLD
  - Steering Committee, Pediatric Liver Disease Special Interest Group
  - Abstract Review Committee, Acute Liver Failure and Artificial Liver Support
- Finance Committee, NASPGHAN
- Advisor to the Executive Committee, INSPPIRE

HONORS

- Ashbel Smith Distinguished Alumnus Award, University of Texas Medical Branch
- Best Doctors, P*ittsburgh Magazine*, annually since 2005
- *Best Doctors in America*, Woodward/White, Inc., annually since 1993

MAJOR LECTURESHIPS AND SEMINARS

- "Acute Liver Failure in Children: A Brief History of Time," World Congress, Pediatric Gastroenterology, Hepatology, and Nutrition, Montreal, Canada, October 2016
- "Acute Liver Failure," breakfast session, World Congress, Pediatric Gastroenterology, Hepatology, and Nutrition, Montreal, Canada, October 2016
- "Acute Liver Failure," Third Annual James Ted Engle Pediatric Liver Transplant Lectureship, Cleveland Clinic, Cleveland, Ohio, October 2016
- "Childhood Liver Disease Research Network (ChiLDReN) and Pittsburgh Liver Research Center (PLRC): Opportunities for Collaboration," University of Pittsburgh, Pittsburgh, Pa., June 2017

Arvind Srinath, MD

RESEARCH

Arvind Srinath's research focuses on subspecialty medical education, pediatric gastroenterology curriculum development, and functional GI disorders.

- A multicenter, randomized, double-blind, placebo-controlled, parallel-group, safety and efficacy study of a range of linaclotide doses administered orally to children ages 7 to 17 years who have IBS with constipation (i.e., Fulfill Rome III Criteria for Child/Adolescent IBS and Fulfill Modified Rome III Criteria for Child/Adolescent Functional Constipation)
- A phase III, multicenter, long-term, safety, efficacy, and pharmacokinetics study of lubiprostone in pediatric subjects aged ≥ 6 years to < 18 years who have functional constipation
- Impact of a nurse practitioner–led teaching program on patient and caregiver knowledge of pediatric IBD, NASPGHAN/Association for Pediatric Gastroenterology and Nutrition Nurses, Susan Moyer 2017 Nursing Research Grant (co-principal investigator)

ADVISORY COMMITTEE MEMBERSHIPS

- NASPGHAN
- Training Committee, NASPGHAN, September 2015 to the present
- Resident Selection Committee, Children's Hospital of Pittsburgh
- Residency Program Leadership, Children's Hospital of Pittsburgh
- Pediatric Gastroenterology Fellow Clinical Competency Committee, Children's Hospital of Pittsburgh
- Pediatric Gastroenterology Fellow Selection Committee, Children's Hospital of Pittsburgh

PROFESSIONAL AFFILIATIONS/SOCIETY MEMBERSHIPS

- NASPGHAN
- AGA
- CCFA
- American Academy of Pediatrics

HONORS

- *Best Doctors in America*, Woodward/White, Inc.

MAJOR LECTURESHIPS AND SEMINARS

- "Pediatric Gastroesophageal Reflux," general pediatric podcast with Candace Jones, 2017
- "Good Teaching: What Works and What We Can Do," invited presentation, Children's Mercy Hospital, Kansas City, Mo., April 2017

Veena Venkat, MD

RESEARCH

The Pittsburgh Cholestatic Liver Disease Consortium (ChiLDReN). This NIH-funded network conducts detailed clinical and translational investigations of cholestastic liver diseases in children. Veena Venkat is a co-investigator.

A Phase I/IIa Trial of Intravenous Immunoglobulin Therapy Following Portoenterostomy in Infants With Biliary Atresia. This study aims to determine the feasibility, acceptability, tolerability, and safety profile of intravenous immunoglobulin treatment after hepatic portoenterostomy for biliary atresia. Venkat is site principal investigator.

Immunosuppression Withdrawal for Stable Pediatric Transplant Recipients. The long-term objective of this NIH-funded network is to improve outcomes for pediatric liver transplant recipients through discoveries that will guide clinical decision-making related to immunosuppression withdrawal and simultaneously ensure excellent allograft function while minimizing complications of immunosuppression. Venkat is a co-investigator.

Improving Medication Adherence in Children Who Had a Liver Transplant (U34). This is the next phase of a now-completed R01 grant (MALT: Medication Adherence in Children Who Had a Liver Transplant) that validated the use of a marker of medication nonadherence, the Medication Level Variability Index (MLVI), in predicting rejection in children who have had a liver transplant. The researchers are developing a multisite trial in which pediatric liver transplant centers will use the MLVI to identify nonadherent recipients and a telemetric intervention to improve adherence.

A Pilot Study of Frailty in Children With End-Stage Liver Disease. The overall objectives are to validate the concept of frailty in children awaiting liver transplantation and to arrive at an accurate and efficient instrument to gauge the morbidity of transplant patients and to improve post-transplant outcomes beyond survival alone. Venkat is site principal investigator.

Pediatric Primary Sclerosing Cholangitis Consortium. This is a multicenter study with the objective of describing the natural history of primary sclerosing cholangitis in children. Venkat is the site principal investigator.

A Multicenter Collaboration to Study Primary Sclerosing Cholangitis in Children: Primary Sclerosing Cholangitis Partners Seeking a Cure. The objective of this multicenter study is to collect retrospective data on children with primary sclerosing cholangitis that can be used to build a clinical trial and ultimately find an effective therapy. Venkat is the site principal investigator.

CTOTC: Biomarkers for PTLDs in Children. This is a clinical trial investigating biomarkers associated with the development of EBV-related PTLDs in pediatric liver, heart, heart with liver, small intestine, and liver with small intestine transplant candidates and recipients. Venkat is a co-investigator.

ADVISORY COMMITTEE MEMBERSHIPS

- Member, Steering Committee, Pediatric Special Interest Group, AASLD
- Co-chair, Biliary Atresia Natural History and Therapeutics Working Group, ChiLDReN Research Network
- Protocol Committee: A Prospective Database of Older Children with Biliary Atresia, ChiLDReN
- Member, Education Committee, SPLIT, November 2014 to the present

Xunjun Xiao, PhD

Xunjun Xiao served as research assistant professor until March 31, 2017.

RESEARCH

Xiao has been extensively involved in ongoing efforts to better understand how newborns digest dietary fats. His work aims to understand how PLRP2 functions in human newborns. First, he is characterizing the properties of purified PLRP2 in the laboratory. Second, he is translating his findings in newborn mice, demonstrating that PLRP2 is critical for fat digestion. The investigation may assist in finding nutritional therapies to treat the 10% of newborns who have slow weight gain at birth.

Xiao also works to elucidate the mechanism behind chronic pancreatitis. Most children with chronic pancreatitis have genetic mutations that increase their risk, but the contributing mechanisms are not well known. The researchers are using model systems to test the novel hypothesis that expression of the mutant proteins in the pancreas leads to a stress response and cell death. The findings stand to increase understanding of pancreatic injury and provide guidance for developing new pharmacological interventions to prevent the ongoing damage from progressing to chronic changes in the pancreas.

PROFESSIONAL AFFILIATIONS/SOCIETY MEMBERSHIPS

- American Society of Nutrition Sciences
- American Society of Biochemistry and Molecular Biology

HONORS

- Western Pennsylvania National Pancreas Foundation Research
- Grant Award: "Carboxyl Ester Lipase Hybrid Confers Susceptibility to Chronic Pancreatitis Through Proteotoxicity"

TEACHING ACTIVITIES

NEW TEACHING PROGRAMS AND COURSES (AND OTHER EDUCATIONAL INNOVATIONS)

During the 2016–2017 academic year, the Division of Gastroenterology, Hepatology, and Nutrition has been implementing new educational programs to enhance its faculty and fellows. Under the fellowship direction of Arvind Srinath and Riha Bhatt, as well as the division chiefs, David Keljo (interim) and Mark Lowe (past), the division has augmented and enhanced the curriculum for its fellows, as well as provided more educational opportunities for faculty to increase their knowledge in the many subspecialties that make up the division. Mentoring activities occur in the division's research laboratories and across clinical research teams.

PEDIATRIC EXOCRINE PANCREATIC SEMINAR SERIES

Husain was awarded an unrestricted educational grant from Abbvie to launch a new educational series for fellows and faculty, titled "The Pediatric Exocrine Pancreatic Disorders Seminar Series." The seminar series hosted authorities in the field of clinical and research pancreatology. It included the following visiting professors.

NAME	INSTITUTION	PRESENTATION	DATE OF VISIT
Miklos Sahin-Toth, MD, PhD	Boston University	"A Spotlight on the Genetic Basis for Hereditary Pancreatitis"	November 18, 2016
Vijay P. Singh, MBBS	Mayo Clinic, Scottsdale, Ariz.	"Nutrition in Pancreatitis"	December 16, 2016
Bradley Barth, MD, MPH	University of Texas Southwestern	"Pediatric Pancreatic Interventions"	January 13, 2016
Tanja Gonksa, MD	Hospital for Sick Children and University of Toronto	"CFTR and Pancreatitis"	February 17, 2017
Vikesh Singh, MD, MS	Johns Hopkins University	"Fluids, Pancreatitis Severity, and What's in the Pipeline for Pancreatitis Treatments"	March 17, 2017

The series received overwhelmingly positive feedback, and Husain looks forward to renewed funding.

PEDIATRIC GASTROENTEROLOGY FELLOWSHIP PROGRAM

Srinath and Bhatt, directors of the Pediatric Gastroenterology Fellowship Program, through requested feedback from fellows and faculty, enhanced the program as follows.The directors, along with the division's hepatologists, developed an inpatient rotation specifically to enhance the care of hepatology and transplant hepatology patients. The curriculum was augmented with targeted didactic objectives to be covered in monthly case-based conferences and weekly service patient conferences.

- Sevilla joined the faculty in 2016 and has since codeveloped and implemented (with the fellowship program directors) an advanced nutrition curriculum for the pediatric gastroenterology fellows. Presentations for fellows and faculty occur quarterly.
- A chief pediatric gastroenterology fellow program was piloted. The goal of the program is to offer leadership and mentoring opportunities to the two senior fellows. Feedback from the fellows has indicated success, which has led to its continuation next year.

PEDIATRIC TRANSPLANT HEPATOLOGY FELLOWSHIP PROGRAM

In November 2016, James Squires began the application process to restart the Pediatric Transplant Hepatology Fellowship Program. He received accreditation from the Accreditation Council for Graduate Medical Education (ACGME) for its establishment. He will lead the transplant hepatology team of McKiernan, Robert Squires, Khan, and Venkat and work directly with the pediatric transplant surgeon team to build the program.

RESEARCH LABORATORY TEACHING ACTIVITIES

Ghazi has been very active teaching courses and giving lectures for PhD students, fellows, residents, and medical students in a variety of courses.

TEACHING HONORS AND AWARDS

Hilary Michel was awarded First-Year Fellow of the Year by the UPMC Residency Program. This is the second year in a row that a pediatric gastroenterology fellow has received the award.

QUALITY MEASURES FOR TEACHING ACTIVITES

In addition to continuous bidirectional feedback from faculty, fellows, and staff, the division conducts the following quality measures for teaching activities: fellow evaluations (quarterly), which have been aligned with the ACGME milestones; Clinical Competency Committee evaluations, which include evaluations of fellow teaching activities; resident evaluations (continuous throughout the year for fellows and faculty); and yearly fellow training exam scores.

THREE-YEAR BIBLIOGRAPHY

2015

Alonso EM, James LP, Zhang S, **Squires RH**; Pediatric Acute Liver Failure Study G. Acetaminophen adducts detected in serum of pediatric patients with acute liver failure. *J Pediatr Gastroenterol Nutr.* 2015;61(1):102-7.

Cummings EE, O'Reilly LP, **King DE**, Silverman RM, Miedel MT, Luke CJ, **Perlmutter DH**, Silverman GA, Pak SC. Deficient and null variants of SERPINA1 are proteotoxic in a *Caenorhabditis elegans* model of alpha1-antitrypsin deficiency. *PLOS One*. 2015;10(10):e0141542.

Cutler DJ, Zwick ME, Okou DT, Prahalad S, Walters T, Guthery SL, Dubinsky M, Baldassano R, Crandall WV, Rosh J, Markowitz J, Stephens M, Kellermayer R, Pfefferkorn M, Heyman MB, LeLeiko N, Mack D, Moulton D, Kappelman MD, Kumar A, Prince J, Bose P, Mondal K, Ramachandran D, Bohnsack JF, Griffiths AM, Haberman Y, Essers J, Thompson SD, Aronow B, **Keljo DJ**, Hyams JS, Denson LA, Group P-KR, Kugathasan S. Dissecting allele architecture of early onset IBD using high-density genotyping. *PLOS One.* 2015;10(6):e0128074.

Eisses JF, Criscimanna A, Dionise ZR, Orabi AI, Javed TA, Sarwar S, Jin S, Zhou L, Singh S, Poddar M, Davis AW, Tosun AB, Ozolek JA, **Lowe ME**, Monga SP, Rohde GK, Esni F, **Husain SZ**. Valproic acid limits pancreatic recovery after pancreatitis by inhibiting histone deacetylases and preventing acinar redifferentiation programs. *Am J Pathol.* 2015 Dec;185(12):3304-15.

Grossi V, Lerer T, Griffiths A, LeLeiko N, Cabrera J, Otley A, Rick J, Mack D, Bousvaros A, Rosh J, Grossman A, Saeed S, Kay M, Boyle B, Oliva-Hemker M, **Keljo D**, Pfefferkorn M, Faubion W, Kappelman MD, Sudel B, Markowitz J, Hyams JS. Concomitant use of immunomodulators affects the durability of infliximab therapy in children with Crohn's disease. *Clin Gastroenterol Hepatol*. 2015;13(10):1748-56.

Haberman Y, Tickle TL, Dexheimer PJ, Kim MO, Tang D, Karns R, Baldassano RN, Noe JD, Rosh J, Markowitz J, Heyman MB, Griffiths AM, Crandall WV, Mack DR, Baker SS, Huttenhower C, **Keljo DJ**, Hyams JS, Kugathasan S, Walters TD, Aronow B, Xavier RJ, Gevers D, Denson LA. Corrigendum. Pediatric Crohn disease patients exhibit specific ileal transcriptome and microbiome signature. *J Clin Invest*. 2015;125(3):1363.

Hidvegi T, Stolz DB, Alcorn JF, Yousem SA, Wang J, Leme AS, Houghton AM, Hale P, Ewing M, Cai H, Pastore N, Annunziata P, Kaminski N, Pilewski J, Shapiro SD, Pak SC, Silverman GA, Brunetti-Pierri N, **Perlmutter DH**. Enhancing autophagy with drugs or lung-directed gene therapy reverses the pathological effects of respiratory epithelial cell proteinopathy. *J Biol Chem.* 2015.

Huprikar S, Danziger-Isakov L, Ahn J, Naugler S, Blumberg E, Avery RK, Koval C, Lease ED, Pillai A, Doucette KE, Levitsky J, Morris MI, Lu K, McDermott JK, Mone T, Orlowski JP, Dadhania DM, Abbott K, Horslen S, Laskin BL, Mougdil A, **Venkat VL**, Korenblat K, Kumar V, Grossi P, Bloom RD, Brown K, Kotton CN, Kumar D. Solid organ transplantation from hepatitis B virus-positive donors: Consensus guidelines for recipient management. *Am J Transplant.* 2015;15(5):1162-72.

Jericho HS, Kaurs E, Boverhof R, Knisely A, **Shneider BL**, Verkade HJ, Whitington PF. Bile acid pool dynamics in progressive familial intrahepatic cholestasis with partial external bile diversion. *J Pediatr Gastroenterol Nutr*. 2015;60(3):368-74.

Jin S, Orabi AI, Le T, Javed TA, Sah S, **Eisses JF**, Bottino R, Molkentin JD, **Husain SZ**. Exposure to radiocontrast agents induces pancreatic inflammation by activation of nuclear factor-kappaB, calcium signaling, and calcineurin. *Gastroenterology.* 2015;149(3):753-64 e711.

Khan FA, **Squires RH**, Litman HJ, Balint J, Carter BA, Fisher JG, Horslen SP, Jaksic T, Kocoshis S, Martinez JA, Mercer D, Rhee S, Rudolph JA, Soden J, Sudan D, Superina RA, Teitelbaum DH, Venick R, Wales PW, Duggan C; Pediatric Intestinal Failure C. Predictors of enteral autonomy in children with intestinal failure: A multicenter cohort study. *J Pediatr.* 2015;167(1):29-34 e21.

Le T, **Eisses JF**, Lemon KL, Ozolek JA, Pociask DA, Orabi AI, **Husain SZ**. Intraductal infusion of taurocholate followed by distal common bile duct ligation leads to a severe necrotic model of pancreatitis in mice. *Pancreas*. 2015;44(3):493-9.

Lowe ME, Whitcomb DC. Next generation of pancreatic enzyme replacement therapy: Recombinant microbial enzymes and finding the perfect lipase. *Gastroenterology.* 2015 Dec;149(7):1678-81.

Oliva-Hemker M, Hutfless S, Al Kazzi ES, Lerer T, Mack D, LeLeiko N, Griffiths A, Cabrera J, Otley A, Rick J, Bousvaros A, Rosh J, Grossman A, Saeed S, Kay M, Carvalho R, **Keljo D**, Pfefferkorn M, Faubion W Jr., Kappelman M, Sudel B, Schaefer ME, Markowitz J, Hyams JS.

Clinical presentation and five-year therapeutic management of very early-onset inflammatory bowel disease in a large North American cohort. *J Pediatr.* Sep 2015;167(3):527-32 e521-3.

Orabi AI, Sah S, Javed TA, Lemon KL, Good ML, Guo P, Xiao X, Prasadan K, Gittes GK, Jin S, **Husain SZ**. Dynamic imaging of pancreatic nuclear factor kappaB (NF-kappaB) activation in live mice using adeno-associated virus (AAV) infusion and bioluminescence. *J Biol Chem.* 2015;290(18):11309-20.

Perlmutter DH, Chopra K, **Chu A**. Two new considerations for improving the diagnosis of alpha1-antitrypsin deficiency-associated liver disease. *Dig Dis Sci.* 2015;60(6):1511-3.

Schwarzenberg SJ, Bellin M, **Husain SZ**, Ahuja M, Barth B, Davis H, Durie PR, Fishman DS, Freedman SD, Gariepy CE, Giefer MJ, Gonska T, Heyman MB, Himes R, Kumar S, Morinville VD, **Lowe ME**, Nuehring NE, Ooi CY, Pohl JF, Troendle D, Werlin SL, Wilschanski M, Yen E, Uc A. Pediatric chronic pancreatitis is associated with genetic risk factors and substantial disease burden. *J Pediatr.* 2015;166(4):890-6 e891.

Silverman RM, Cummings EE, O'Reilly LP, Miedel MT, Silverman GA, Luke CJ, **Perlmutter DH**, Pak SC. The aggregation-prone intracellular serpin SRP-2 fails to transit the ER in *Caenorhabditis elegans*. *Genetics.* May 2015;200(1):207-19.

Sorensen LG, Neighbors K, Zhang S, Limbers CA, Varni JW, Ng VL, **Squires RH**, Alonso EM; Pediatric Acute Liver Failure Study G. Neuropsychological functioning and health-related quality of life: Pediatric acute liver failure study group results. *J Pediatr Gastroenterol Nutr.* 2015;60(1):75-83.

Squires JE. 50 years ago in the *Journal of Pediatrics*: Primary liver cell carcinoma associated with biliary cirrhosis due to congenital bile duct atresia. *J Pediatr.* 2015;167(1):63.

Squires JE, Shivakumar P, Mourya R, Bessho K, Walters S, Bezerra JA. Natural killer cells promote long-term hepatobiliary inflammation in a low-dose rotavirus model of experimental biliary atresia. *PLOS One.* 2015;10(5):e0127191.

Sunseri WM, Kugathasan S, **Keljo DJ**, Greer JB, Ranganathan S, Cross RK, Siegel CA, Regueiro MD. IBD LIVE Case Series-Case 3: Very early-onset inflammatory bowel disease: When genetic testing proves beneficial. *Inflamm Bowel Dis.* 2015;21(12):2958-68.

Szabo A, **Xiao X**, Haughney M, Spector A, Sahin-Toth M, **Lowe ME**. A novel mutation in PNLIP causes pancreatic triglyceride lipase deficiency through protein misfolding. *Biochim Biophys Acta.* 2015;1852(7):1372-9.

Haberman Y, Tickle TL, Dexheimer PJ, Kim MO, Tang D, Karns R, Baldassano RN, Noe JD, Rosh J, Markowitz J, Heyman MB, Griffiths AM, Crandall WV, Mack DR, Baker SS, Huttenhower C, **Keljo DJ,** Hyams JS, Kugathasan S, Walters TD, Aronow B, Xavier RJ, Gevers D, Denson LA. Corrigendum. Pediatric Crohn disease patients exhibit specific ileal transcriptome and microbiome signature. *J Clin Invest*. 2015 Mar 2;125(3):1363.

Szigethy E, Youk AO, Gonzalez-Heydrich J, Bujoreanu SI, Weisz J, Fairclough D, Ducharme P, Jones N, Lotrich F, **Keljo D**, **Srinath A**, Bousvaros A, Kupfer D, DeMaso DR. Effect of 2 psychotherapies on depression and disease activity in pediatric Crohn's disease. *Inflamm Bowel Dis.* 2015;21(6):1321-8.

Grossi V, Lerer T, Griffiths A, LeLeiko N, Cabrera J, Otley A, Rick J, Mack D, Bousvaros A, Rosh J, Grossman A, Saeed S, Kay M, Boyle B, Oliva-Hemker M, **Keljo D**, Pfefferkorn M, Faubion W, Kappelman MD, Sudel B, Markowitz J, Hyams JS. Concomitant Use of Immunomodulators Affects the Durability of Infliximab Therapy in Children with Crohn's Disease. *Clin Gastroenterol Hepatol*. 2015 Oct; 13(10):1748-56.

Hommel KA, Gray WN, Hente E, Loreaux K, Ittenbach RF, Maddux M, Baldassano R, Sylvester F, Crandall W, Doarn C, Heyman MB, **Keljo D**, Denson LA. The Telehealth Enhancement of Adherence to Medication (TEAM) in pediatric IBD trial: Design and methodology. *Contemp Clin Trials*. 2015 May 21;43:105-113.

Cutler DJ, Zwick ME, Okou DT, Prahalad S, Walters T, Guthery SL, Dubinsky M, Baldassano R, Crandall WV, Rosh J, Markowitz J, Stephens M, Kellermayer R, Pfefferkorn M, Heyman MB, LeLeiko N, Mack D, Moulton D, Kappelman MD, Kumar A, Prince J, Bose P, Mondal K, Ramachandran D, Bohnsack JF, Griffiths AM, Haberman Y, Essers J, Thompson SD, Aronow B, **Keljo DJ**, Hyams JS, Denson LA; PRO-KIIDS Research Group, Kugathasan S. Dissecting Allele Architecture of Early Onset IBD Using High-Density Genotyping. *PLoS One*. 2015 Jun 22;10(6):e0128074.

Sunseri WM, Kugathasan S, **Keljo DJ**, Greer JB, Ranganathan S, Cross RK, Siegel CA, Regueiro MD. IBD LIVE Case Series-Case 3: Very Early-Onset Inflammatory Bowel Disease: When Genetic Testing Proves Beneficial. *Inflamm Bowel Dis*. 2015 Dec;21(12):2958-68.

Tafaleng EN, Chakraborty S, Han B, Hale P, Wu W, Soto-Gutierrez A, Feghali-Bostwick CA, Wilson AA, Kotton DN, Nagaya M, Strom SC, Roy-Chowdhury J, Stolz DB, **Perlmutter DH**, Fox IJ. Induced pluripotent stem cells model personalized variations in liver disease resulting from alpha1-antitrypsin deficiency. *Hepatology.* 2015;62(1):147-57.

Taylor CJ, Chen K, Horvath K, Hughes D, **Lowe ME**, Mehta D, Orabi AI, Screws J, Thomson M, Van Biervliet S, Verkade HJ, **Husain SZ**, Wilschanski M. ESPGHAN and NASPGHAN report on the assessment of exocrine pancreatic function and pancreatitis in children. *J Pediatr Gastroenterol Nutr.* 2015;61(1):144-53.

Venkat VL, Ranganathan S, Sindhi R. The challenges of liver transplantation in children with primary sclerosing cholangitis. *Expert Rev Gastroenterol Hepatol*. 2015;9(3):289-94.

Winkler EA, Nishida Y, Sagare AP, Rege SV, Bell RD, **Perlmutter D**, Sengillo JD, Hillman S, Kong P, Nelson AR, Sullivan JS, Zhao Z, Meiselman HJ, Wenby RB, Soto J, Abel ED, Makshanoff J, Zuniga E, De Vivo DC, Zlokovic BV. GLUT1 reductions exacerbate Alzheimer's disease vasculo-neuronal dysfunction and degeneration. *Nature Neuroscience.* Apr 2015;18(4):521-30.

Xiao X, **Lowe ME**. The beta5-loop and lid-domain contribute to the substrate specificity of pancreatic lipase related protein 2 (PNLIPRP2). *J Biol Chem.* 2015; 290(48): 28847-56.

2016

Qaddoumi I, Orisme W, Wen J, Santiago T, Gupta K, Dalton JD, Tang B, Haupfear K, Punchihewa C, Easton J, Mulder H, **Boggs K**, Shao Y, Rusch M, Becksfort J, Gupta P, Wang S, Lee RP, Brat D, Peter Collins V, Dahiya S, George D, Konomos W, Kurian KM, McFadden K, Serafini LN, Nickols H, Perry A, Shurtleff S, Gajjar A, Boop FA, Klimo PD Jr, Mardis

ER, Wilson RK, Baker SJ, Zhang J, Wu G, Downing JR, Tatevossian RG, Ellison DW. Genetic alterations in uncommon low-grade neuroepithelial tumors: BRAF, FGFR1, and MYB mutations occur at high frequency and align with morphology. *Acta Neuropathol*. 2016 Jun; 131(6): 833-45.

Zhang J, McCastlain K, Yoshihara H, Xu B, Chang Y, Churchman ML, Wu G, Li Y, Wei L, Iacobucci I, Liu Y, Qu C, Wen J, Edmonson M, Payne-Turner D, Kaufmann KB, Takayanagi SI, Wienholds E, Waanders E, Ntziachristos P, Bakogianni S, Wang J, Aifantis I, Roberts KG, Ma J, Song G, Easton J, Mulder HL, Chen X, Newman S, Ma X, Rusch M, Gupta P, **Boggs K**, et al. Deregulation of DUX4 and ERG in acute lymphoblastic leukemia. *Nat Genet*. 2016;48(12):1481-9.

Faber ZJ, Chen X, Gedman AL, **Boggs K**, Cheng J, Ma J, et al. The genomic landscape of core-binding factor acute myeloid leukemias. *Nat Genet*. 2016;48(12):1551-6.

Chu AS, Chopra KB, **Perlmutter DH**. Is severe progressive liver disease caused by alpha-1-antitrypsin deficiency more common in children or adults? *Liver Transpl*. 2016;22(7):886-94.

Eisses JF, **Husain SZ**. Molecular understanding of chronic pancreatitis. In Beger H (Ed.), *The Pancreas* (3rd ed.). Ulm, Germany: Wiley-Blackwell; 2016.

Rajan D, Greer JB, Regueiro MD, Baidoo L, Binion DG, Herfarth HH, Siegel CA, Hartman DJ, Farraye FA, Koutroubakis IE, Brand MH, Williams ED, **Goyal A**, Cross RK. IBD LIVE Case Series-Case 6: Persistent skin lesions in a patient with Crohn's disease: You hear hoof beats and discover a zebra. *Inflamm Bowel Dis*. 2016;22(11):2754-64.

Huang Y, Liu C, Eisses JF, **Husain SZ**, Rohde GK. A supervised learning framework for pancreatic islet segmentation with multi-scale color-texture features and rolling guidance filters. *Cytometry A*. 2016;89(10):893-902.

Husain S, Silveira FP, Azie N, Franks B, Horn D. Epidemiological features of invasive mold infections among solid organ transplant recipients: PATH Alliance® registry analysis Med Mycol. 2017 Apr 1;55(3):269-277.

Husain S, Sole A, Alexander BD, Aslam S, Avery R, Benden C, et al. The 2015 International Society for Heart and Lung Transplantation guidelines for the management of fungal infections in mechanical circulatory support and cardiothoracic organ transplant recipients: Executive summary. *J Heart Lung Transplant*. 2016;35(3):261-82.

Kumar S, Ooi CY, Werlin S, Abu-El-Haija M, Barth B, Bellin MD, Durie PR, Fishman DS, Freedman SD, Gariepy C, Giefer MJ, Gonska T, Heyman MB, Himes R, **Husain SZ**, Lin TK, **Lowe ME**, Morinville V, Palermo JJ, Pohl JF, Schwarzenberg SJ, Troendle D, Wilschanski M, Zimmerman MB, Uc A. Risk factors associated with pediatric acute recurrent and chronic pancreatitis: Lessons from INSPPIRE. *JAMA Pediatr*. 2016;170(6):562-9.

Pohl J, Morinville V, **Husain SZ**, Uc A. Toxic-metabolic risk factors are uncommon in pediatric chronic pancreatitis. *J Pediatr Gastroenterol Nutr*. 2016;62(6):e66-7.

Ting J, Wilson L, Schwarzenberg SJ, Himes R, Barth B, Bellin MD, Durie PR, Fishman DS, Freedman SD, Gariepy CE, Giefer MJ, Gonska T, **Husain SZ**, Kumar S, Morinville VD, **Lowe ME**, Ooi CY, Pohl JF, Troendle D, Usatin D, Werlin SL, Wilschanski M, Heyman MB, Uc A. Direct costs of acute recurrent and chronic pancreatitis in children in the INSPPIRE registry. *J Pediatr Gastroenterol Nutr*. 2016;62(3):443-9.

Husain SZ, Morinville V, Pohl J, Abu-El-Haija M, Bellin MD, Freedman S, et al. Toxic-metabolic risk factors in pediatric pancreatitis: Recommendations for diagnosis, management, and future research. *J Pediatr Gastroenterol Nutr*. 2016;62(4):609-17.

Levine A, **Keljo D**, DeMaso D, Bousvaros A, **Szigethy E**. O-017 YI inflammatory versus non-inflammatory predictors of specific depressive symptoms in a large pediatric cohort with IBD. *Inflamm Bowel Dis*. 2016;22 Suppl 1:S6.

Abdul-Baki H, Greer JB, Binion DG, Kermanshahi TR, Brand MH, Williams ED, Bensen SP, Holubar SD, **Keljo DJ,** Regueiro MD. IBD LIVE Case Series-Case 5: The Many Faces of Cytomegalovirus in Inflammatory Bowel Disease. *Inflamm Bowel Dis*. 2016 Sep;22(9):2245-54.

Keljo DJ. 2015 AAP Murray Davidson Award—Special feature. *J Pediatr Gastroenterol Nutr*. 2016;63(3):311.

Feld LD, Rodriguez D, Margolese J, Rosh JR, Rubin M, **Kim SC**, Rubin DT. National survey of inflammatory bowel disease patients identifies risk factors for emergency department visits. *Inflamm Bowel Dis.* 2016;23:224-32.

Lipstein EA, Lovell DJ, Denson LA, **Kim SC**, Spencer C, Britto MT. Parents' information needs and influential factors when making decision about TNF-a inhibitors. *Pediatric Rheumatology.* 2016;14:53.

Lipstein EA, Lovell DJ, Denson LA, **Kim SC**, Spencer C, Ittenbach RF, Britto MT. High levels of decisional conflict and decision regret when making decisions about biologics. *J Pediatr Gastroenterol Nutr.* 2016;63:e176-81.

Serpico M, Boyle B, Kemper KJ, **Kim SC**. Complementary and alternative medicine use in children with inflammatory bowel diseases: A single center survey. *JPGN*. 2016;63:651-7.

Szabo FK, Hornung L, Oparaji JA, Alhosh R, Husain SZ, Liu QY, Palermo J, Lin TK, Nathan JD, Podberesky DJ, **Lowe M**, Fei L, Abu-El-Haija M. A prognostic tool to predict severe acute pancreatitis in pediatrics. *Pancreatology*. 2016;16(3):358-64.

Uc A, Andersen DK, Bellin MD, Bruce JI, Drewes AM, Engelhardt JF, Forsmark CE, Lerch MM, **Lowe ME**, Neuschwander-Tetri BA, O'Keefe SJ, Palermo TM, Pasricha P, Saluja AK, Singh VK, **Szigethy EM**, Whitcomb DC, Yadav D, Conwell DL. Chronic pancreatitis in the 21st Century—Research challenges and opportunities: Summary of a National Institute of Diabetes and Digestive and Kidney Diseases workshop. *Pancreas*. 2016;45(10):1365-75.

Molven A, Fjeld K, **Lowe ME**. Lipase genetic variants in chronic pancreatitis: When the end is wrong, all's not well. *Gastroenterology*. 2016;150(7):1515-8.

Kelly D, Verkade HJ, Rajanayagam J, **McKiernan P**, Mazariegos G, Hubscher S. Late graft hepatitis and fibrosis in pediatric liver allograft recipients: Current concepts and future developments. *Liver Transpl*. 2016;22(11):1593-602.

McKiernan P, Ball S, Santra S, Foster K, Fratter C, Poulton J, et al. Incidence of primary mitochondrial disease in children younger than 2 years presenting with acute liver failure. *J Pediatr Gastroenterol Nutr*. 2016;63(6):592-7.

McKiernan PJ, Greene CM. High-throughput profiling for discovery of non-coding RNA biomarkers of lung disease. *Expert Rev Mol Diagn*. 2016;16(2):173-85.

Ranawaka R, Lloyd C, **McKiernan PJ**, Hulton SA, Sharif K, Milford DV. Combined liver and kidney transplantation in children: Analysis of renal graft outcome. *Pediatr Nephrol.* 2016;31(9):1539-43.

Sentner CP, Hoogeveen IJ, Weinstein DA, Santer R, Murphy E, **McKiernan PJ**, et al. Glycogen storage disease type III: Diagnosis, genotype, management, clinical course, and outcome. *J Inherit Metab Dis.* 2016;39(5):697-704.

Shneider BL, de Ville de Goyet J, Leung DH, Srivastava A, Ling SC, Duche M, **McKiernan P**, Superina R, **Squires RH**, Bosch J, Groszmann R, Sarin SK, de Franchis R, Mazariegos GV. Primary prophylaxis of variceal bleeding in children and the role of MesoRex Bypass: Summary of the Baveno VI Pediatric Satellite Symposium. *Hepatology*. 2016;63(4):1368-80.

Sreekantam S, Nicklaus-Wollenteit I, Orr J, Sharif K, Vijay S, **McKiernan PJ**, et al. Successful long-term outcome of liver transplantation in late-onset lysosomal acid lipase deficiency. *Pediatr Transplant*. 2016;20(6):851-4.

van Ginkel WG, Jahja R, Huijbregts SC, Daly A, MacDonald A, De Laet C, Cassiman D, Eyskens F, Korver-Keularts IM, Goyens PJ, **McKiernan PJ**, van Spronsen FJ. Neurocognitive outcome in tyrosinemia type 1 patients compared to healthy controls. *Orphanet J Rare Dis*. 2016;11(1):87.

Staufner C, Haack TB, Kopke MG, Straub BK, Kolker S, Thiel C, Freisinger P, Baric I, **McKiernan PJ**, Dikow N, Harting I, Beisse F, Burgard P, Kotzaeridou U, Lenz D, Kuhr J, Himbert U, Taylor RW, Distelmaier F, Vockley J, Ghaloul-Gonzalez L, Ozolek JA, Zschocke J, Kuster A, Dick A, Das AM, Wieland T, Terrile C, Strom TM, Meitinger T, Prokisch H, Hoffmann GF. Recurrent acute liver failure due to NBAS deficiency: Phenotypic spectrum, disease mechanisms, and therapeutic concepts. *J Inherit Metab Dis*. 2016;39(1):3-16.

Mukherjee A, Chakraborty S, Wang Y, Chu A, Perlmutter D. Liver disease in α1-antitrypsin deficiency (pages 111-40). Switzerland: Springer International Publishing; 2016.

Sevilla WMA, McElhanon B. Optimizing transition to home: Enteral nutrition for pediatric patients. *Nutrition in Clinical Practice.* 2016;31(6):762-8.

Savoie KB, Bachier-Rodriguez M, Jones TL, Jeffrey K, Papraniku D, **Sevilla WMA**, et al. Standardization of feeding advancement after neonatal gastrointestinal surgery: Does it improve outcomes? *Nutrition in Clinical Practice*. 2016;31(6):810-8.

Srinath AI, Gupta N, **Husain SZ**. Probing the association of pancreatitis in inflammatory bowel disease. *Inflamm Bowel Dis*. 2016;22(2):465-75.

Keerthy D, Youk A, **Srinath AI**, Malas N, Bujoreanu S, Bousvaros A, **Keljo D**, Demaso DR, Szigethy EM. Effect of Psychotherapy on Healthcare Utilization in Children with Inflammatory Bowel Disease and Depression. *J Pediatr Gastroenterol Nutr*. 2016 Dec;63(6):658-664.

Pulcini CD, Zettle S, **Srinath A.** Refeeding syndrome. *Pediatr Rev*. 2016;37(12):516-23.

Boiko JR, **Srinath AI**, Cooper JD. Portomesenteric venous thrombosis in previously healthy adolescents presenting with subacute abdominal pain. *Clin Pediatr (Phila)*. 2016 Sep;55(10):975-8.

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

Squires JE. Protecting the allograft following liver transplantation for PFIC1. *Pediatr Transplant*. 2016;20(7):882-3.

Stiehl SR, **Squires JE**, Bucuvalas JC, Hemmelgarn TS. Tacrolimus interaction with dexmedetomidine-a case report. *Pediatr Transplant.* 2016;20(1):155-7.

Squires JE. Protecting the allograft following liver transplantation for PFIC1. *Pediatr Transplant*. 2016;20(7):882-3.

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

Celik N, **Squires RH**, Vockley J, Sindhi R, Mazariegos G. Liver transplantation for maple syrup urine disease: A global domino effect. *Pediatr Transplant*. 2016;20(3):350-1.

Shneider BL, Magee JC, Karpen SJ, Rand EB, Narkewicz MR, Bass LM, Schwarz K, Whitington PF, Bezerra JA, Kerkar N, Haber B, Rosenthal P, Turmelle YP, Molleston JP, Murray KF, Ng VL, Wang KS, Romero R, **Squires RH**, Arnon R, Sherker AH, Moore J, Ye W, Sokol RJ, Childhood Liver Disease Research Network. Total serum bilirubin within three months of hepatoportoenterostomy predicts short-term outcomes in biliary atresia. *J Pediatr*. 2016;170:211-7,e1-2.

Mouzaki M, Bass LM, Sokol RJ, Piccoli DA, Quammie C, Loomes KM, Heubi, JE, Hertel PM, Scheenstra R, Furuya K, Kutsch E, Spinner NB, Robbins KN, **Venkat V**, Rosenthal P, Beyene J, Baker A, Kamath BM. Early-life predictive markers of liver disease outcome in an international, multicenter cohort of children with Alagille syndrome. *Liver International*. 2016;36(5):755-60.

Xiao X, Fischbach S, Fusco J, Zimmerman R, Song Z, Nebres P, et al. PNA lectin for purifying mouse acinar cells from the inflamed pancreas. *Scientific Reports*. 2016;6:21127.

Xiao X, Jones G, Sevilla WA, Stolz DB, Magee KE, Haughney M, et al. A carboxyl ester lipase (CEL) mutant causes chronic pancreatitis by forming intracellular aggregates that activate apoptosis. *J Biol Chem*. 2016;291(44):23224-36.

Veena L. Venkat. Liver Transplantation. In Christine Waasdorp-Hurtado, B U.K. Li (Eds) *The NASPGHAN Fellows Concise Review of Pediatric Gastroenterology, Hepatology and Nutrition, Second Edition. 2016 Elsevier.*

Xiao X, Jones G, Sevilla WA, Stolz DB, Magee KE, Haughney M, **Mukherjee A**, Wang Y, **Lowe ME.** A Carboxyl Ester Lipase (CEL) Mutant Causes Chronic Pancreatitis by Forming Intracellular Aggregates That Activate Apoptosis. *J Biol Chem*. 2017 May 12;292(19):7744.

2017

Orabi AI, **Wen L**, **Javed TA**, Le T, Guo P, Sanker S, Ricks D, **Boggs K**, **Eisses JF**, Castro C, **Xiao X**, Prasadan K, Esni F, Gittes GK, **Husain SZ**. Targeted inhibition of pancreatic acinar cell calcineurin is a novel strategy to prevent post-ERCP pancreatitis. *Cell Mol Gastroenterol Hepatol*. 2017;3(1):119-28.

Abu-El-Haija M, Kumar S, Quiros JA, Balakrishnan K, Barth B, Bitton S, **Eisses JF**, Foglio EJ, Fox V, Francis D, Freeman AJ, Gonska T, Grover AS**, Husain SZ**, et al. The management of acute pancreatitis in the pediatric population: A clinical report from the NASPGHAN Pancreas Committee. *J Pediatr Gastroenterol Nutr*. 2017.

King CD, Singh D, Holden K, Govan AB, Keith SA, **Ghazi A**, Robinson RASR. Dataset of proteomics analysis of aging *C. elegans* exposed to *Pseudomonas aeruginosa*. *Data Brief*. 2017;11:245-51.

Amrit FRG, **Ghazi A**. Analysis of *C. elegans* transcriptomic data by the Tuxedo suite of the Galaxy pipeline. *J Vis Exp*. 2017;122.

Amrit FRG, **Ghazi A**. Influences of germline cells on organismal lifespan and healthspan. In: Gill MS, Olsen A (Eds.), C. elegans *and Its Contribution to Our Understanding of Aging and Longevity*. Springer; 2017.

Husain SZ, **Srinath A**. What's unique about acute pancreatitis in children? *Nature Reviews Gastroenterology*. 2017;14(6):366-72.

Oparaji J, Rose F, Okafor D, Howard A, Turner RL, Orabi AI, Byersdorfer C, Mi Q, Ritchey K, **Lowe ME**, **Husain SZ**. Risk factors for asparaginase-associated pancreatitis: A systematic review. *J Clin Gastroenterol*. 2017;51(10):907-13.

Scheers I, Gonska T, Barth B, Fishman DS, Freedman SD, Gariepy CE, Giefer MJ, Heyman MB, Himes R, **Husain SZ**, **Lowe ME**, et al. Autoimmune pancreatitis in children: Characteristic features, diagnosis and management. *Am J Gastroenterology*. 2017;112(10):1604-11.

Troendle DM, Fishman DS, Barth BA, Giefer MJ, Lin TK, Liu QY, Abu-El-Haija M, Bellin MD, Durie PR, Freedman SD, Gariepy C, Gonska T, Heyman MB, Himes R, **Husain SZ**, Kumar S, **Lowe ME**, et al. Therapeutic endoscopic retrograde cholangiopancreatography in pediatric patients with acute recurrent and chronic pancreatitis: Data from the INSPPIRE (INternational Study group of Pediatric Pancreatitis: In search for a cuRE) Study. *Pancreas*. 2017;46(6):764-9.

Giefer MJ, Lowe ME, Werlin SL, Zimmerman B, Wilschanski M, Troendle D, Schwarzenberg SJ, Pohl JF, Palermo J, Ooi CY, Morinville VD, Lin TK, **Husain SZ**, et al. Early-onset acute recurrent and chronic pancreatitis is associated with PRSS1 or CTRC gene mutations. *J Pediatr*. 2017;186:95-100.

Kugathasan S, Denson LA, Walters TD, Kim MO, Marigorta UM, Schirmer M, Mondal K, Liu C, Griffiths A, Noe JD, Crandall WV, Snapper S, Rabizadeh S, Rosh JR, Shapiro JM, Guthery S, Mack DR, Kellermayer R, Kappelman MD, Steiner S, Moulton DE, **Keljo D**, et al. Prediction of complicated disease course for children newly diagnosed with Crohn's disease: A multicentre inception cohort study. *Lancet*. 2017;389(10080):1710-8.

Khan Z, Yokota S, Ono Y, Bell AW, Oertel M, Stolz DB, Michalopoulos GK. Bile duct ligation induces ATZ globule clearance in a mouse model alpha-1 antitrypsin deficiency. *Gene Exp*. 2017;17(2)115-27.

Khan Z, Orr A, Michalopoulos GK, Ranganathan S. Immunohistochemical analysis of the stem cell marker LGR5 in pediatric liver disease. *Pediatr Dev Pathol*. 2017;20(1):16-27.

Khan Z, **Venkat VL**, Soltys KA, Stolz DB, Ranganathan S. A challenging case of severe infantile cholestasis in alpha-1 antitrypsin deficiency. *Pediatr Dev Pathol*. 2017;20(2):176-81.

Mitchell EM, **Khan Z**. Liver disease in alpha-1 antitrypsin deficiency: Current approaches and future directions. *Curr Pathobiol Rep*. 2017;5:243.

Khan Z, Strom SC. Hepatocyte transplantation in special populations: Clinical use in children. *Methods Mol Biol*. 2017;1506:3-16.

Stollon N, Zhong Y, Ferris M, Bhansali S, Pitts B, Rak E, Kelly M, **Kim S**, van Tilburg M. Chronological age when healthcare transition skills are mastered in adolescents/young adults with inflammatory bowel disease. *World J Gastro.* 2017;23:3349-55.

Watson KA, **Kim SC**, Boyle BM, Saps M. The prevalence and impact of functional abdominal pain disorders in children with inflammatory bowel diseases (IBD-FAPD). *J Pediatr Gastroenterol Nutr.* 2017;65(2):212-7.

Brooks AJ, Smith PJ, Cohen R, Collins P, Douds A, Forbes V, Gaya DR, Johnston BT, **McKiernan PJ**, et al. UK guideline on transition of adolescent and young persons with chronic digestive diseases from paediatric to adult care. *Gut.* 2017;66:988-1000.

Coad J, Toft A, Claridge L, Ferguson J, Hind J, Jones R, Mcclean P, **McKiernan P**, et al. Using mobile phone technology to support young liver transplant recipients moving to adult services: A systematic rapid review. *Progress in Transplantation.* 2017;27:207-18.

Droge C, Bonus M, Baumann U, Klindt C, Lainka E, Kathemann S, Brinkert F, Grabhorn E, Pfister ED, Wenning D, Fichtner A, Gotthardt DN, Weiss KH, **McKiernan P**, et al. Sequencing of FIC1, BSEP, and MDR3 in a large cohort of patients with cholestasis revealed a high number of different genetic variants. *J Hepatol.* 2017;67(6):1253-64.

Ekong UD, **McKiernan P**, Martinez M, Lobritto S, Kelly D, Ng VL, et al. Long-term outcomes of *de novo* autoimmune hepatitis in pediatric liver transplant recipients. *Pediatr Transplant*. 2017;21(6).

Grammatikopoulos T, **McKiernan PJ**, Dhawan A. Portal hypertension and its management in children. *Arch Dis Child.* 2017.

Hadzic N, Baumann U, **McKiernan P**, Mclin V, Nobili V. Long-term challenges and perspectives of pre-adolescent liver disease. *Lancet Gastroenterol Hepatol*. 2017;2(6):435-45.

Jahnel J, Zohrer E, Fischler B, D'antiga L, Debray D, Dezsofi A, Haas D, Hadzic N, Jacquemin E, Lamireau T, Maggiore G, **McKiernan PJ**, et al. Attempt to determine the prevalence of two inborn errors of primary bile acid synthesis: Results of a European survey. *J Pediatr Gastroenterol Nutr*. 2017;64(6):864-8.

Mclin VA, Allen U, Boyer O, Bucuvalas J, Colledan M, Cuturi MC, D'antiga L, Debray D, Dezsofi A, Goyet JV, Dhawan A, Durmaz O, Falk C, Feng S, Fischler B, Franchi-Abella S, Frauca E, Ganschow R, Gottschalk S, Hadzic N, Hierro L, Horslen S, Hubscher S, Karam V, Kelly D, Maecker-Kolhoff B, Mazariegos G, **McKiernan P**, et al. Early and late factors impacting patient and graft outcome in pediatric liver transplantation: Summary of an ESPGHAN Monothematic Conference. *J Pediatr Gastroenterol Nutr.* 2017;65:e53-9.

Van Dam E, Daly A, Venema-Liefaard G, Van Rijn M, Derks TG, **McKiernan PJ**, et al. What is the best blood sampling time for metabolic control of phenylalanine and tyrosine concentrations in tyrosinemia type 1 patients? *JIMD Rep*. 2017;36:49-57.

McKiernan PJ. Recent advances in liver transplantation for metabolic disease. *J Inherit Metab Dis.* 2017;40:491-5.

McKiernan P. Liver transplantation for hereditary tyrosinaemia type 1 in the United Kingdom. *Adv Exp Med Biol.* 2017;959:85-91.

McKiernan P. Pathobiology of the hepatic glycogen storage diseases. *Current Pathobiology Reports.* 2017;5:253-7.

McKiernan PJ. The acutely ill baby. In: Kelly DA (Ed.), *Diseases of the Liver and Biliary System in Children* (4th ed.). Chichester, England: Wiley-Blackwell; 2017.

Kerecuk L, **McKiernan PJ**. Congenital and structural abnormalities of the liver. In: Kelly DA (Ed.), *Diseases of the Liver and Biliary System in Children* (4th ed.). Chichester, England: Wiley-Blackwell; 2017.

McKiernan PJ, Hoffmann G. Metabolic liver disease. In: Hoffmann et al. (Eds.), *Inherited Metabolic Diseases* (2nd ed.). Springer; 2017.

Szydlowski EG, **Rudolph JA**, Vitale MA, Zuckerbraun NS. Bloodstream infections in patients with intestinal failure presenting to a pediatric emergency department with fever and a central line. *Pediatr Emerg Care*. 2017;33(12):e140-5.

Turay F, **Rudolph JA**. Nutrition and gastroenterology. In: Zitelli BJ, McIntire SC, Nowalk AJ (Eds.), *Zitelli and Davis' Atlas of Pediatric Physical Diagnosis* (7th ed.). Philadelphia, PA: Elsevier Saunders; 2017.

Squires JE, Balistreri WF, Bezerra JA. Pediatric cholestatic syndromes. In: Sanyal AJ, Boyer T, Terrault N, Lindor K (Eds.), *Zakim & Boyer's Hepatology: A Textbook of Liver Disease* (7th ed.). Elsevier; 2017.

Rees MA, **Squires JE**, Tadros S, Squires JH. Canal of Nuck hernia: A multimodality imaging review. *Pediatr Radiol*. 2017;47(8):893-8.

Braun T, Di Segni A, BenShoshan M, Asaf R, **Squires JE**, Farage Barhom S, et al. Fecal microbial characterization of hospitalized patients with suspected infectious diarrhea shows significant dysbiosis. *Sci Rep*. 2017;7(1):1088.

Squires JE, Balistreri WF. Hepatitis C virus infection in children and adolescents. *Hepatology Communications*. 2017;1(2):87-98.

Squires JE, Celik N, Morris A, Soltys K, Mazariegos G, Shneider B, **Squires RH**. Clinical variability following partial external biliary diversion in familial intrahepatic cholestasis 1 deficiency. *J Pediatr Gastroenterol Nutr*. 2017;64(3):425-30.

Squires JH, Bill A, Thieret J, **Squires JE**. Identification of suspected hepatocellular carcinoma with contrast-enhanced ultrasound. *J Pediatr*. 2017;182:398.

Squires JE, Wallihan D, Bradley K, Serai S, Rattan M, Miethke A. Hepatocyte-specific contrast-enhanced MRI for quantitative assessment of liver function in children. *Int J Rare Dis and Orphan Drugs*. 2017;2(1):1002.

Liang J, Alfano DN, **Squires JE**, Riley MM, Parks WT, Kofler J, et al. Novel NLRC4 mutation causes a syndrome of perinatal autoinflammation with hemophagocytic lymphohistiocytosis, hepatosplenomegaly, fetal thrombotic vasculopathy, and congenital anemia and ascites. *Pediatr Dev Pathol*. 2017.

Narkewicz M, Horslen S, Belle SH, Rudnick D, Ng V, Rosenthal R, Romero R, Loomes KM, Zhang S, **Squires RH**; for the Pediatric Acute Liver Failure Study Group. Prevalence and significance of auto-antibiodies in children with acute liver failure. *J Pediatr Gastroenterol Nutr*. 2017;64:210-7.

Soltys KA, Sindhi R, Soto-Gutiérrez A, Sada R, Haberman K, Gramignoli R, Crowley J, Quader MA, Deutsch M, Ashokkumar C, Tafaleng E, Shneider BL, **Squires RH**, et al. Host conditioning and rejection monitoring in clinical hepatocyte transplantation. *J Hepatology*. 2017;66:987-1000.

Feldman AG, Sokol RJ, Hardison RM, Alonso EM, **Squires RH**, Narkewicz MR; Pediatric Acute Liver Failure Study Group. Lactate and lactate:pyruvate ratio in the diagnosis and outcomes of pediatric acute liver failure. *J Pediatr*. 2017;182:217-22.

Shneider BL, Moore J, Kerkar N, Magee J, Ye W, Karpen SJ, Kamath B, Molleston J, Bezerra JA, Murray K, Loomes K, Whitington PF, Rosenthal P, **Squires RH**, et al., for the Childhood Liver Disease Research Network. Initial assessment of the infant with neonatal cholestasis—Is this biliary atresia? *PLoS One.* 2017;12:e0176275.

Javid PJ, Oron AP, Duggan C, **Squires RH**, Horslen SP; on behalf of the Pediatric Intestinal Failure Consortium. The extent of intestinal failure–associated liver disease in patients referred for intestinal rehabilitation is associated with increased mortality: An analysis of the Pediatric Intestinal Failure Consortium Database. *J Pediatr Surg*. 2017. pii: S0022-3468(17)30546-8.

Makadia P, **Srinath A**, Madan-Khetarpal S, McGuire M, Infante E, Zhang J, et al. Aplastic anemia and cytotoxic T lymphocyte antigen-4 haploinsufficiency treated with bone marrow transplantation. *J Allergy Clin Immunol Pract*. 2017;5(5):1445-7.

Deneau MR, El-Matary W, Valentino PL, Abdou R, Alqoaer K, Amin M, Amir AZ, Auth M, Bazerbachi F, Broderick A, Chan A, Cotter J, Doan S, El-Youssef M, Ferrari F, Furuya KN, Gottrand M, Gottrand F, Gupta N, Homan M, Kamath BM, Mo Kim K, Kolho KL, Konidari A, Koot B, Iorio R, Ledder O, Mack C, Martinez M, Miloh T, Mohan P, O'Cathain N, Papadopoulou A, Ricciuto A, Saubermann L, Sathya P, Shteyer E, Smolka V, Tanaka A, Varier R, **Venkat V**, et al. The natural history of primary sclerosing cholangitis in 781 children: A multicenter, international collaboration. *Hepatology*. 2017;66(2):518-27.

Xiao X, Fischbach S, Zhang T, Chen C, Sheng Q, Zimmerman R, Patnaik S, Fusco J, Ming Y, Guo P, Shiota C, Prasadan K, Gangopadhyay N, **Husain SZ**, et al. SMAD3/Stat3 signaling mediates β-cell epithelial-mesenchymal transition in chronic pancreatitis-related diabetes. *Diabetes*. 2017;66(10):2646-58.