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## SLUCare Division of Pediatric Gastroenterology and Hepatology Faculty Publications and Presentations (selected)

### Faculty Publications

**Patel D**, Principal Investigator, Absorption and Safety with Sustained Use of Relizorb™ Evaluation (ASSURE) Study in Patients with Cystic Fibrosis Receiving Enteral Feeding. July 1 2016- current

**Patel D**, Principal Investigator, CFFC - A Long-Term Prospective Observational Safety Study of the Incidence of and Risk Factors for Fibrosing Colonopathy in US Patients with Cystic Fibrosis Treated with Pancreatic Enzyme Replacement Therapy: A Harmonized Protocol Across Sponsors. July 1 2016- current

Mukhopadhyay A, Baligar P, Mohanty S, Arindkar S, Mukherjee S, **Teckman JH**, Patel S. Bone marrow stem cell therapy partially ameliorates pathological consequences of liver in mice expressing mutant human  $\alpha$ 1-antitrypsin. *Hepatology* 2017 Apr;65(4):1319-1335. doi: 10.1002/hep.29027. Epub 2017 Feb 25. PMID: 28056498.

**Jain AK**, Sharma A, Arora S, Blomenkamp KS, Jun I, Luong R, Westrich, DJ, Mittal A, Buchanan PM, Guzman MA, Long JP, Neuschwander-Tetri BA, **Teckman JH**. Preserved gut microbial diversity accompanies upregulation of TGR5 and hepatobiliary transporters in bile acid treated animals on Parenteral Nutrition. *JPEN J Parenter Enteral Nutr.* 2017 Feb;41(2):198-207. Premier Research Paper

Pastore N, Attanasio S, Granese B, Castello R, **Teckman J**, Wilson AA, Ballabio A, Brunetti-Pierri N. Activation of the c-Jun N-terminal kinase pathway aggravates proteotoxicity of hepatic mutant Z alpha1-antitrypsin. *Hepatology.* (2017) Jun;65(6):1865-1874. doi: 10.1002/hep.29035. Epub 2017 Apr 10. PMID: 28073160

Schwarzenberg SJ, Ling SC, Cloonan YK, Lin HS, Evon DM, Murray KF, Rodriguez-Baez N, Rosenthal P, **Teckman J**, Schwarz KB. Health-related Quality of Life in Pediatric Patients with Chronic Hepatitis B Living in the United States and Canada. *J Pediatr Gastroenterol Nutr.* 2017 May;64(5):760-769. PMID: 28169971.

Hazari YM, Bashir A, Habib M, Bashir S, Habib H, Qasim MA, Shah NN, Haq E, **Teckman JH**, Fazili KM. Alpha-1-antitrypsin Deficiency: Genetic Variations, Clinical Manifestations and Therapeutic Interventions. *Mutation Research.* Vol. 773, 1 July 2017, p14-25 PMID: 28927525

Ratchford Kirby A, Pinz H, **Patel D**. Intractable Diarrhea due to Undiagnosed DGAT1 Mutation Leading to Severe Electrolyte Derangements, Pneumatosis Intestinalis, Protein-Losing Enteropathy, and Rickets in a Female Infant (In production, accepted Sep 7 2017 in *Journal of Pediatric Gastroenterology, hepatology and Nutrition.*

**Teckman JH**. Chapter 8. Alpha-1-antitrypsin Deficiency Liver Disease. In *Alpha-1-antitrypsin Deficiency: Biology, Diagnosis, Clinical Significance and Emerging Therapies.* Noor Kalsheker, Robert Stockley, eds. Elsevier Publishing, 2017. p117-131.

**Jain AK, Teckman JH**. Wilson's Disease; in *Decision Support in Medicine: Pediatrics.* Barrett DJ, McMillan J, Jones D, Boney C. eds. (Textbook on-line app, 2017).

**Teckman JH**. Emerging Concepts and Human Trials in Alpha-1-Antitrypsin Deficiency Liver Disease. *Seminars in Liver Disease;* 37(02): 152-158. 2017. Gores G and Bruix J eds. PMID: 28564723

**Jain AK**, Le Roux C, Puri P, Tavakkoli A, Miller NG, Laferrere B, Kellermayer R, DiBase JK, Martindale, RG, Wolfe BM. Proceedings of the 2017 ASPEN Research Workshop— Gastric Bypass: Role of the Gut. *JPEN J Parenter Enteral Nutr.* 2018;Feb 42:279–295

Turner AM, Stolk J, Bals R, Lickliter J, Hamilton J, Christianson DR, Given BD, Burdon JG, Loomba R, Stoller JK, **Teckman JH**. Hepatic-targeted RNA interference provides persistent knockdown of alpha-1 antitrypsin levels in ZZ patients. *J Hepatology* (in press). 2018 Mar 21. pii: S0168-8278(18)30176-4. doi: 10.1016/j.jhep.2018.03.012. [Epub ahead of print] PMID: 29572094

Shen S, Sanchez ME, Blomenkamp K, Corcoran EM, Marco E, Yudkoff CJ, Bumcrot D, Jiang H, **Teckman JH**, Albright CF. Amelioration of Alpha-1 Antitrypsin Deficiency Diseases with Genome Editing in Transgenic Mice. *Human Gene Therapy.* 2018 Mar 16. doi: 10.1089/hum.2017.227. [Epub ahead of print] PMID: 29641323.

Seth R, Akinboyo I, Chhabra A, Qaiyum Y, Shet A, Gupte N, **Jain AK**, Jain SK. Mobile-Phone Incentives for Childhood Immunizations in Rural India. *Pediatrics.* 2018 Apr;141(4). pii: e20173455. doi: 10.1542/peds.2017-3455. Epub 2018 Mar 14

Villalona G, Price A, Blomenkamp KS, Manithody C, Ratchford T, Saxena S, Westrich M, Kakarla V, Pochampally S, Phillips W, Heafner N, Korremla N, Greenspon J, Guzman M, **Jain AK**. No gut no gain! Enteral bile acid treatment preserves gut growth but is not protective for parenteral nutrition associated liver injury in a novel extensive short bowel animal model. Premier Research Paper. *JPEN J Parenter Enteral Nutr.* 2018; Apr:1–14

Higher purpose. Greater good.

Karlow KE, Africa JA, Wells A, Hou J, Beth PH, Behling CA, **Jain AK**, Molleston JP, Newton KP, Rosenthal P, Vos MB, Xanthak SA, Lavine J, Schwimmer JB, for the Nonalcoholic Steatohepatitis Clinical Research Network (NASH CRN). Clinically Actionable Hypercholesterolemia and Hypertriglyceridemia in Children with Nonalcoholic Fatty Liver Disease. *J Pediatr*. 2018 Apr 13. j.jpeds.2018.02.038.

**Mangalat N, Teckman JH**. Pediatric Intestinal Failure Review. *Children* 2018, 5, 100; doi:10.3390/. PMID: 30037012

Compher C, **Jain AK**, Nichol Peter, Blackmer A, Earthman C, Evans D, McCarthy M, Taylor B, Mehta N. Research Agenda 2018: the American Society for Parenteral and Enteral Nutrition. *JPEN, J Parenter Enteral Nutr*. 2018 Jul;42(5):838-844

Loomes KM, Spino C, Goodrich NP, Hangartner TN, Marker AE, Heubi JE, Kamath BM, Schneider BJ, Rosenthal P, Hertel PM, Karpen SJ, Molleston JP, Murray KF, Schwarz KB, Squires RH, **Teckman JH**, Turmelle YP, Alonso EM, Sherker AH, Magee JC, Sokol RJ, Childhood Liver Disease Research Network (ChiLDReN). Bone Density in Children with Chronic Liver Disease Correlates with Growth and Cholestasis. *Hepatology*, 2018 Jul 31. doi: 10.1002/hep.30196. PMID: 30063078.

Tang Y, Blomenkamp KS, Fickert P, Trauner M, **Teckman JH**. norUDCA promotes degradation of  $\alpha$ 1-antitrypsin mutant Z protein by inducing autophagy through AMPK/ ULK1 pathway. *PLOS One*. August 1, 2018. 13(8): e0200897. <https://doi.org/10.1371/journal.pone.0200897>. PMID: 30067827

Ratchford TL, **Teckman JH, Patel D**. Gastrointestinal Pathophysiology and Nutrition in Cystic Fibrosis. *Expert Review in Gastroenterology and Hepatology*. 2018 Sep;12(9):853-862. doi: 10.1080/17474124.2018.1502663. Epub 2018 Aug 3. PMID: 30019967

Price A, Blomenkamp KS, Manithody C, Saxena S, Abraham S, Greenspon J, Villalona G, **Jain AK**. Developing a novel ambulatory total parenteral nutrition dependent short bowel syndrome animal model. *Journal of Surgical Research*. (Accepted, In Print, Aug 2018)

**Patel D**, Principal Investigator, - A long term Gastrointestinal Symptoms endpoint evaluation by CF foundation. GALAXY trial- August 2018- current

Denton C, Price A, Friend J, Manithody C, Blomenkamp K, Westrich M, Kakarla V, Philips W, Krebs J, Abraham S, Osei H, **Jain AK**. Role of the Gut-Liver Axis in Driving Parenteral Nutrition Associated Injury. *Children* (Accepted, In Print Sep 2018)

**Mangalat N MD, Patel D MD** Internal sphincter achalasia in a child with intestinal failure presenting with feeding intolerance and pseudo-obstruction. Accepted and to be presented at 10th International Pediatric Intestinal Failure Symposium at Pittsburg PA September 22 2018.

Shapiro WL, Yu E, Collins J, Murray K, Ali S, Desai N, Xanthakos S, Lin H, Alkhouri N, Abdou R, Abrahams S, Butler M, Faasse SA, Gills L, Hadley TA, **Jain AK**, Kavan M, Kordy K, Lee P, Panganiban J, Pohl J, Potter C, Rudolph B, Sundaram S, Joshi S, Proudfoot J, Goyal N, Harlow K, Newton K, Schwimmer J. Clinical Practice Approach to Nonalcoholic Fatty Liver Disease by Pediatric Gastroenterologists in the United States, JPGN (Accepted, JPGN Sep 2018)

Rosenthal P, Ling SC, Belle SH, Murray KF, Rodriguez-Baez N, Schwarzenberg SJ, **Teckman J**, Lin HS, Schwarz KB; Hepatitis B Research Network (HBRN). Combination of entecavir/peginterferon alfa-2a in children with HBeAg-positive immune tolerant chronic hepatitis B virus infection. *Hepatology* 2018 Oct 15. doi: 10.1002/hep.30312. [Epub ahead of print] PMID: 30318613

**Patel D, Teckman JH**. Alpha-1-antitrypsin Deficiency Liver Disease. *Clinics in Liver Disease*. Nov. 22 (2018) 643-655. Elsevier. Gitlin N and Rosenthal P eds. PMID: 30266154

Ratchford TL, **Teckman JH, Patel D**. Nutrition in Cystic Fibrosis. In: Gupte S, Gupte SB, Gupte M (Eds): *Recent Advances in Pediatrics (Special Vol 28: Child Nutrition in Practice)*. New Delhi: Jaypee 2018.

Steven D. Freedman, MD, PhD, Colby Wyatt, John Stevens, Dhiren Patel, Robert Gallotto, Danica Grujic, Perry Brown doi: 10.1097/MPG.0000000000002110, *Journal of Pediatric Gastroenterology and Nutrition* Absorption and Safety with Sustained Use of RELiZORB Evaluation (ASSURE) in Patients with CF Receiving Enteral Feeding

#### **Faculty Presentations**

Denton C MD, **Patel D**. An unusual case of dysphagia in an adolescent boy. Accepted to be presented at NASPGHAN annual meeting at Hollywood FL , October 23 2018

Stacy Kim, MD, Thomas Ratchford, MD, Paula Buchanan, MPH, PhD, **Dhiren Patel, MD**, Ting Tao, MD, PhD, **Jeffrey Teckman, MD**, Jeffrey J. Brown, MD, and Shannon G. Farmakis, Diagnostic accuracy of non-contrast MR Enterography in detecting active bowel inflammation in pediatric patients with diagnosed or suspected inflammatory bowel disease to determine necessity of gadolinium-based contrast agents. oral presentation at 104th Scientific Assembly and Annual Meeting, November 25-30 McCormick Place RSNA 2018, Chicago IL

Pediatricians at the SSM Health Cardinal Glennon Children's Hospital are faculty of Saint Louis University School of Medicine and members of SLUCare Physician Group, the clinical arm of Saint Louis University School of Medicine.