Innovations in Gastroenterology & GI Surgery

Rady Children's Hospital San Diego

Rady Children's - A comprehensive system focused solely on children.

PEOPLE

New physician brings expertise in advanced procedure

Koorosh Kooros, M.D., an expert in the field of capsule endoscopy, recently joined the Division of Gastroenterology, Hepatology & Nutrition at Rady Children's Hospital-San Diego. He is also an associate physician diplomate at UC San Diego School of Medicine.

Dr. Kooros will use capsule endoscopy to evaluate suspected Crohn's disease, obscure GI bleeding, vascular malformations, Meckel's diverticulum, intestinal polyposis syndrome and other small bowel pathology. As the procedure does not require ionizing radiation, and deep sedation or general anesthesia, it is especially beneficial for pediatric patients.

Dr. Kooros earned his medical degree at the University of Pecs Medical School in Pecs, Hungary. He completed his internship and residency in pediatrics at Children's Hospital, University of Pecs Medical School and St. Luke's - Roosevelt Hospital Center in New York and his fellowship in pediatric gastroenterology, hepatology and nutrition at Floating Hospital for Children at Tufts Medical Center.

A fellow of the American Gastroenterological Association, Dr. Kooros is also an active member of NASPGHAN (North American Society of Pediatric Gastroenterology, Hepatology and Nutrition), where he has served on various committees, including the Endoscopy and Procedures committee and International committee. Along with capsule endoscopy, his clinical interests include therapeutic endoscopy, GI disorders in autism spectrum disorders, gastroesophageal reflux disease (GERD) and eosinophilic esophagitis.

PROGRAMS

Colorectal Surgery Clinic offers expert, coordinated care

The new Colorectal Surgery Clinic combines the expertise of the Gastroenterology and Pediatric Surgery divisions to provide integrated care for children with colon and rectal issues. Hayat Mousa, M.D., clinical director of pediatric gastroenterology and director of the Neurogastroenterology and Motility Center, and Rebecca Cherry, M.D., provide the gastroenterology consultations.

The team treats various types of functional and surgical colorectal conditions, including anorectal malformations/ imperforate anus, Hirschsprung's disease, motility disorders, cloacal malformations, encopresis/idiopathic constipation, fecal incontinence and inflammatory bowel disease. Among the procedures performed are the posterior sagittal

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innovation belongs in every moment

RESEARCH

Advanced stem cell technology for metabolic liver diseases

The laboratories of Ariel Feldstein, M.D., and Paulina Ordonez, M.D., are heavily invested in the development of human stem cell and human genetic approaches to the problem of metabolic diseases affecting the liver and to

anorectoplasty (PSAEP), also known as the pull-through procedure, anorectal manometry and pudendal nerve stimulation.

To help ensure that children regain as much bowel function and control as possible, the team pays particular attention to post-surgery recovery.

INNOVATIONS

International group creates guidelines for esophageal atresia

The GI working group of the International Network on Esophageal Atresia, comprised of members from the European Society for Paediatric Gastroenterology Hepatology and Nutrition (ESPGHAN) and the North American Society for Pediatric Gastroenterology, Hepatology and Nutrition (NASPGHAN), has developed uniform evidence-based guidelines for the management of GI complications in children with esophageal atresia-tracheoesophageal fistula.

The group, which includes Hayat Mousa, M.D., was tasked with the guideline development to address a fragmented approach to care. Esophageal atresia (EA) is one of the most common congenital digestive anomalies, and with improvements in surgical techniques and intensive care treatments, the focus of care has shifted from mortality to morbidity and quality-of-life issues. Children with EA face gastrointestinal (GI) problems not only in early childhood but also through adolescence and adulthood.

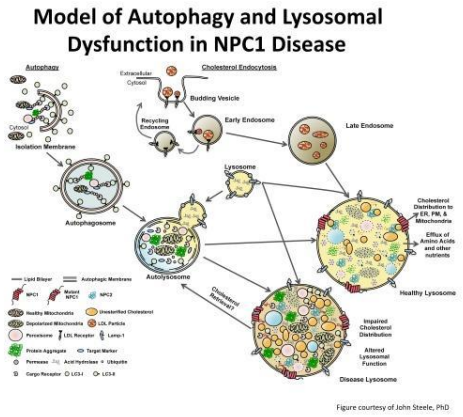
The group developed the guidelines by formulating 36 clinical questions addressing the diagnosis, treatment and prognosis of the common GI complications in patients with EA, including gastroesophageal reflux, cyanotic spells, dysphagia, feeding difficulties, anastomotic strictures and congenital esophageal stenosis. Eosinophilic esophagitis and associated GI anomalies in symptomatic patients with EA were excluded, and the reasons for this are explained. The quality of life of these patients and the importance of a systematic transition of care to adulthood are also discussed.

The guidelines were published in the November 2016 issue of the *Journal of Pediatric Gastroenterology and Nutrition*.
Read the abstract.

harnessing these approaches to find new therapeutic targets in live human hepatocytes.

The main goal of their research is to use human induced pluripotent stem cell (hiPSC) and gene editing technology to derive human hepatocytes that replicate metabolic diseases such as alpha-1 antitrypsin (AAT) deficiency, Niemann Pick type C1 (NPC1) and non-alcoholic fatty liver disease (NAFLD). These hepatocytes carry mutations similar to those that cause disease in patients and can be analyzed to find early phenotypes of disease that can be then rescued to accelerate the development of new therapies.

Using this powerful technology, Dr. Ordonez has discovered a new category of Food and Drug Administration-approved compounds that may offer an immediate therapeutic benefit to NPC1 patients. NPC1 is a rare but devastating lysosomal storage disease that causes neurodegeneration leading to death, often in childhood. Two of the most promising compounds identified are already approved for use in patients for other indications, underscoring the enormous potential of repurposing compounds for the treatment of rare diseases.



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In addition, Dr. Ordonez's team has used CRISPR gene editing technology to generate the first human stem cell derived model of NAFLD. Specifically, her lab has developed human hepatocytes carrying a polymorphism of PNPLA3 that is strongly associated with the development of fatty liver. PNPLA3 encodes a protein called adiponutrin, which is involved in processing of lipids in the liver and adipose tissue. Human hepatocytes carrying the risk variant of PNPLA3 accumulate triglycerides and generate a strong pro-inflammatory response when exposed to toxic fatty acids, similar to those present in diets rich in saturated fats.

Dr. Ordonez's team is now actively testing compounds that may decrease fat accumulation in human hepatocytes and can be further developed to generate new therapies for NAFLD.

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