

DIGEST

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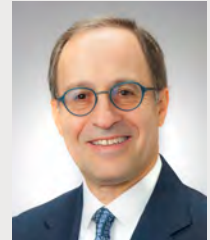
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Disclosures: Drs. Bataller, Behari, Bettner, Dugum, and Klinge have reported no relevant relationships with entities producing health care goods or services.

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Message From the Chief

*I grow old ... I grow old ...
I shall wear the bottoms of my trousers rolled.
Shall I part my hair behind? Do I dare to eat a peach?
I shall wear white flannel trousers, and walk upon the beach.
I have heard the mermaids singing, each to each.
I do not think that they will sing to me.*



The Love Song of J. Alfred Prufrock, by T. S. Eliot

Aging. As expressed so eloquently in this excerpt, aging is replete with “come downs,” including a marked inability to do what you did before — thinning hair, shrinking posture with pants that are too long, teeth and musculature that don’t permit the elegant gnashing of a piece of fruit, or ruefully and tellingly, the loss of appeal to mermaids.

The number of Americans ages 65 and older is projected to more than double from 46 million to over 98 million by 2060. Accompanying the aging of our society will be significant increased demand for nursing homes and ~~huge~~ increases in those living with dementia. The American Geriatrics Society estimates we will need about 30,000 geriatricians by 2030, yet there are only about 7,000 currently practicing. Geriatrics is among the ~~lowest~~ paying specialties and is distinctive in being the only medical specialty ~~I know of which~~ results in a decrement in income with more training – geriatricians make on average about \$20,000 LESS than internists every year. Why do extra training to earn less? It’s not a model primed for success. The aging of the GI tract is another mystery. There are certainly no outward manifestations of aging on endoscopic examination – the 100 year-old colon looks ~~similar~~ to the teenager’s. ~~And, even~~ function, it’s hard to identify a decrement in GI tract ~~fun~~ with time.

Fortunately, the indignity of aging is ~~counter~~-balanced by the repose and wisdom that comes with it. Also, there is no doubting that resources and commitment to studying aging will ~~necessarily and~~ rapidly expand in the future. It is my great pleasure to introduce another edition of Digest, which we hope will never grow old and will continue to inform and inspire. Thanks again for joining us.

To good health,

Robert E. Schoen, MD, MPH
Professor of Medicine and Epidemiology
Chief, Division of Gastroenterology, Hepatology and Nutrition



Affiliated with the University of Pittsburgh School of Medicine, UPMC Presbyterian Shadyside is ranked among America’s Best Hospitals by *U.S. News & World Report*.



Changing the Paradigm: Living-Donor Liver Transplantation

Compared to other nations, the United States performs very few living-donor liver transplants and a high percentage of deceased-donor transplants. This standard has continued for decades, despite a large body of evidence that speaks to the benefits of living donation. But as the discrepancy grows between the number of patients on the waiting list and the number of available deceased-donor livers, living donation is a safe and viable way to expand the donor pool.



Abhinav Humar, MD, is the clinical director of the Thomas E. Starzl Transplantation Institute and chief of the Division of Transplantation in the Department of Surgery at UPMC. His areas of research include partial liver transplants and studies of hepatic regeneration, clinical outcome studies in kidney, liver and pancreas transplants, cytomegalovirus (CMV) disease in transplant recipients, outcomes in pediatric kidney and liver transplants and small bowel transplantation.



Swaytha Ganesh, MD, is the medical director of the Living Donor Program at UPMC. She is renowned for her clinical expertise in treating a wide range of patients with liver disease with a specific interest in living-donor liver transplantation. Her areas of research include evaluating recipients and donors and assessing their ability for living donor surgery and developing disease management protocols in post-liver transplant patients with metabolic syndrome, hyperlipidemia, and systemic hypertension.

Changing the Paradigm: Living-Donor Liver Transplantation

First performed by Thomas E. Starzl, MD, in 1967, liver transplantation offers thousands of patients in the United States a second chance at life. Although this is a life-saving procedure, there is a large discrepancy between the number of available deceased-donor organs and the number of people on the waiting list.

This organ shortage continues to grow. The median waiting time for a patient with a MELD of 29 has gone from 201 days in 2014 to 691 days in 2014. Approximately 20 percent of the patients on the list succumb to their disease prior to receiving a transplant, creating high waiting list mortality.

Patients are listed based on their MELD score, which predicts mortality while on the wait list. However, the MELD score has several limitations. For patients listed with a low MELD score, a living-donor liver transplant may be their best and only option.

At UPMC, our mission is to offer early access to transplant for patients on the liver transplant waiting list. We consider living-donor liver transplantation a first-line option, rather than a last resort, for patients on our liver transplant waiting list. In both 2017 and 2018, we performed more than 50 percent of our liver transplants with living donors.

Benefits of Living Donation

Living-donor liver transplant has several benefits, including reduced time on the transplant waiting list, early access to transplant giving these patients the best chance of survival, immunological advantages, and a shorter length of stay at the hospital post liver transplant.

Due to that shorter wait and the elective, non-emergent nature of a living donor transplant, patients can be transplanted before their disease has progressed to a more severe state, leading to improved patient and graft survival and reduced recovery time when compared to deceased donor transplantation.

Other benefits include fewer instances of postoperative complications, the ability to overcome MELD limitations, and the incredible opportunity for family, friends, and the community to be part of saving a life. In many cases, because a living-donor liver transplant is a non-urgent procedure, the procedure can be scheduled well ahead of time, at the convenience of the patient and the living donor.

The most impactful benefit for the transplant community comes from the living donor themselves. When a living donor volunteers to donate to a loved one, they bring a new liver into the organ pool and reduce the demand for deceased-donor livers.

Why U.S. Hospitals Perform Few Living Donor Transplants

Internationally, and especially in East Asian countries like South Korea, China, and Taiwan, living donor transplantation is the highest standard of care. But U.S. hospitals have been slow to adopt this, due to a few barriers.

The primary obstacle to increasing the availability of living donor transplantation is lack of awareness among both patients and physicians. Many physicians are unfamiliar with living-donor liver transplant or only see it as an option after all other treatments have been exhausted. Unfortunately, at that point many patients have a MELD score so high that they will not experience the most salient benefits of a living donor transplant.

At UPMC, we believe in the power of living donation to save lives. Our main criteria for determining if a patient should pursue living donor transplantation are that they will experience a greater survival benefit with a liver transplant than any other treatment and that they have identified a suitable, willing donor.

Living donor surgery, just like any surgery, does present some level of risk. This risk, along with certain rare but highly-publicized tragedies, is another reason that U.S. transplant centers have been slow to adopt living donor transplantation. Minimizing risk for living donors is a key part of any living donor transplant program, and findings from the National Institutes of Health show that living-donor liver transplant outcomes improve with center experience. The experience of the UPMC transplant team has allowed us to successfully pursue living donation while protecting donors by investing in a rigorous pre-operation evaluation process.

Evaluating Potential Living-Liver Donors

At UPMC, we believe the safety of living donors is of paramount importance. Every potential donor undergoes a 1.5-day evaluation including blood work, physical examination, psycho social evaluation, EKG, chest x-ray, CT scan, and MRI of the abdomen along with other testing. They are seen and evaluated by a multidisciplinary team including a hepatologist, a transplant surgeon, an independent living donor advocate, a nutritionist, a social worker and a living donor advocate.

Living-liver donors should typically be between the ages of 18 and 55, have a BMI of less than 32, and be of generally good health. They should have no history of the following:

- Liver disease
- Heart disease
- HIV
- Ongoing substance abuse
- Cancers or other major health conditions or diseases that could complicate surgery

While the risks for donors are evident, just as they would be in any surgery, it's important that we allow our patients to decide how they want to move forward. Rather than eliminating the option of living-donor liver transplantation, it's important for physicians to decide the best course of action alongside their patient. That's why, at UPMC, we discuss living donor transplantation with every patient who will benefit from a liver transplant.

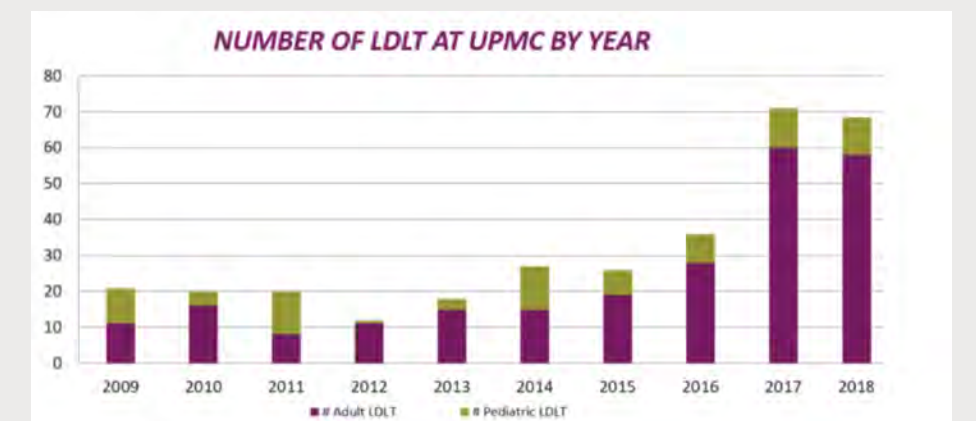
UPMC Expertise

With decades of experience performing living-donor liver transplants, the UPMC Liver Transplant Program continues to lead the field by performing a higher percentage of living-donor liver transplants than any other center in the United States. This allows us to address the particularly acute shortage of deceased-donor livers in our distressed region and to meet the needs of patients who travel to our center for treatment from all around the United States.

Our experienced team of transplant surgeons, hepatologists, nurse coordinators, and social workers collaborate at every stage of the process to ensure optimal outcomes for both patients and donors. We are dedicated to spreading awareness of and educating people about living donation.

We hope to change the paradigm in the United States, to educate patients and physicians about pursuing living donor transplantation not as a last resort, but as a first-line treatment option for every patient on the liver transplant waiting list.

For more information about living donation and how to begin the referral process at UPMC, please visit UPMC.com/LiverTransplantReferral.



Virtual Patient Software: Untapped Technology for Practicing Gastroenterologists

Awarded a Carnegie Science Award in 2018, James B. McGee, MD, has spent more than two decades developing technology to improve medical education for medical students and to make continuing medical education more efficient for physicians. One of his most striking technological innovations is vpSim, a virtual patient software that can help medical students and practicing gastroenterologists develop clinical decision-making skills in an engaging clinical context.



James B. McGee, MD, is director of the Laboratory for Educational Technology at the University of Pittsburgh School of Medicine and an associate professor of medicine with the University of Pittsburgh Division of Gastroenterology, Hepatology and Nutrition. He was awarded a Carnegie Science Award in 2018 for his twenty years of innovative endeavors to bring technology into medical education.

[an image/screengrab of the software itself]

In the early 2000s, online medical education was still in its infancy when Dr. McGee established and became the first editor for online education at the American Gastroenterology Association (AGA), identifying a need to curate, organize, and carefully supervise online educational content for gastroenterologists. "Today, online education is virtually taken for granted, a sign of how successful work in that area has been," says Dr. McGee.

Seeing similar success with recent improvements in technology, virtual patient software is being adopted by medical schools and professional societies around the country, but there are still untapped opportunities to use virtual-patient software in gastroenterology (GI) clinical environments.

Virtual Patient Software and vpSim

Even early iterations of virtual patient software, like the one developed by Dr. McGee at Harvard Medical School in 1996, displayed clear benefits of the simulated clinical environment for student learners.

"Virtual patient software allows a clinician or a student to develop clinical decision-making skills in a safe environment and to get constructive feedback on realistic on-screen patient-management," says Dr. McGee.

Today, because of advancements in technology, virtual patient software allows for even more complex and realistic patient scenarios. Dr. McGee's most recent iteration of this software is called vpSim, a cloud-based platform that enables custom case simulations on an easy-to-use interface. This software allows a clinician or medical student to interview a patient virtually, make decisions about diagnostic tests, determine a diagnosis, treat the patient, and then

receive immediate feedback from a virtual online expert. The main addition of vpSim to the area of simulation is branched decision making trees, which allow clinicians to explore several diagnostic therapeutic pathways and receive feedback on the efficacy of each option relative to other treatments. This software is now used in medical schools and professional settings all around the country because it provides a new way of learning, not through rote memorization and repetition, but by grounding learning in a clinical, patient-focused context.

Both the American Academy of Allergy, Asthma, and Immunology and the American College of Physicians use vpSim to test and practice applying the clinical knowledge of their members and to encourage them to integrate novel therapies and guidelines into their practice. By situating learning in a patient-centered context, simulation software creates a more realistic, engaging, and memorable experience, which physicians prefer because it helps them to learn at a faster pace. The addition of branched decision making creates new opportunities for use in the GI clinical environment.

Opportunities to Use Virtual Patient Software in GI Clinical Environment

Perhaps the most striking features of contemporary treatment of GI diseases is the incredibly wide range of available therapies and the diversity of patient responses to those therapies. For example, Dr. McGee recently treated a patient who was experiencing severe diarrhea with a celiac disease-like clinical picture. After his examination, Dr. McGee realized that this patient was actually suffering from an uncommon but serious side effect of a particular cholesterol lowering medication. Virtual patient software can be used to familiarize physicians with such uncommon occurrences.

The future growth of virtual patient software may have complementary success with genetic science and precision medicine. It is likely that the patient's medication reaction was caused by individual genetic factors. Every year, researchers learn more about how specific genetic profiles relate to the observed clinical state of a patient. This new genetic data allows for gastroenterologists to provide highly-personalized care that could result in more effective treatments.

With more dedicated use of virtual patient training with branching pathways, clinicians can fine-tune their clinical decision-making skills and, in time, may discover linkages to highly-personalized genetic profiles. To learn more about the vpSim and virtual patient simulation, visit Kynectiv.com.

What Is This?

CASE PRESENTATION



Jeffrey Dueker, MD, MPH, is a chief gastroenterology fellow, Year III in the Division of Gastroenterology, Hepatology, and Nutrition.

An 86-year-old male with past medical history for coronary artery disease, COPD, and prior heavy tobacco use presents to the emergency room with dysphagia to solids and liquids. Further history from the patient is notable for immediate regurgitation of all liquids for the past few days, identifying a concern for food impaction. He undergoes an esophagogastroduodenoscopy with removal of a food bolus, followed by computed tomography (CT) without IV contrast. An endoscopic picture from the middle third of the esophagus after the food bolus was removed is shown, as well as a relevant axial CT image.

What is the explanation for this patient's findings? What are the recommended treatment strategies?

Compare your answer to Dr. Dueker's on page 11.



Robotic Repair of Giant Paraesophageal Hernia

Hiatal hernias are common in people more than 50 years of age. Most are small and asymptomatic, but approximately 5 percent are giant paraesophageal hernias (GPEH) with migration of the gastroesophageal junction and portions of the stomach into the chest. When a significant volume of the stomach herniates into the chest space, concerning symptoms such as early satiety, vomiting, and pain cause the patient significant distress and can herald impending emergent obstruction and loss of blood supply to the stomach.



Inderpal S. Sarkaria, MD, FACS, is vice chairman of Clinical Affairs in the Department of Cardiothoracic Surgery and director of Thoracic Robotic Surgery at UPMC. He is a thoracic surgeon who specializes in malignant and benign diseases of the chest including the lungs, pleura, esophagus and mediastinum.

Some patients present in extremis, when the stomach is under torsion, infarcted, and at risk of losing its blood supply. This can cause death of the stomach, which can be life-threatening. Historically, patients with GPEH were referred for surgical repair only in the most urgent cases, because there was a poor perception of the benefits of GPEH repair as compared with the risks of the procedure. Pioneering work done at UPMC by James D. Luketich, MD, FACS, and his colleagues in the UPMC Department of Surgery changed this paradigm and established laparoscopic GPEH repair as the gold standard approach.¹ We are now using robot-assisted laparoscopic surgery for GPEH repair in an increasing number of patients and have found that this safe and effective repair results in excellent outcomes and safety.

Treating GPEH by Laparoscopy

The current standard of care for GPEH repair is to laparoscopically dissect the hernia sac, reduce the hernia, repair the diaphragmatic crura, and perform an anti-reflux procedure. To avoid recurrence, high mediastinal dissection to free as much of the esophagus as possible is very important. The hernia sac and stomach must be completely dissected away from the mediastinal structures to enable a tension-free reduction back into the abdomen. In the Division of Thoracic and Foregut Surgery at UPMC, we tend to avoid the use of mesh or other prosthetics at the diaphragmatic hiatus through aggressive and adept dissection in the mediastinum. A short esophagus, if present, must be recognized and managed with a Collis gastroplasty or other esophageal-lengthening techniques. During the Collis gastroplasty, we tubularize part of the stomach by stapling, so that the esophagus may be lengthened to reduce tension.

Dr. Luketich was one of the first surgeons to explore treating GPEH's laparoscopically. In a landmark, retrospective study of more than 650 GPEH laparoscopic repairs at UPMC, we found that laparoscopic repair provided excellent patient satisfaction, symptom improvement, and preservation of quality of life even when performed in patients with comorbidities. Outcomes were very similar to the outcomes of open surgical repair at an experienced center, harkening back to the gold standard at that time. This new laparoscopic procedure results in far less morbidity, low mortality (<2 percent), and low rates of recurrence.

To date, surgeons at UPMC have performed more than 1,000 laparoscopic GPEH repairs. Few centers can match our experience. Recently, an important study from our group linked performing GPEH repair in an elective setting with reduced morbidity (18 vs. 38 percent) and mortality (1 vs. 8 percent) as compared with non-elective GPEH repair. The need to repair asymptomatic or minimally symptomatic GPEH is often debated, because balancing the risks of surgery with the risks of observation can be difficult, especially in elderly patients. Our findings provide compelling evidence for the benefits of earlier surgical repair. If a patient is having GPEH symptoms, they should be seen by a surgeon. Patients fare better if they are referred for evaluation before their condition requires urgent care.

Benefits of Robotic-Assisted GPEH Repair

We are now working to further improve on this standard of care using robotics, which give expert surgeons the opportunity to work with an extremely precise tool. Robotic surgery — which is actually telemanipulation of an advanced instrument that the surgeon controls completely — has several potential advantages over laparoscopy. The surgical robot allows for better visualization and gives better control of more dexterous tools (Figure 1). The well-characterized benefits of minimally invasive surgery, such as faster recovery times and fewer postoperative complications, seem to carry over to minimally invasive robotic surgeries. The surgeon's experience, expertise, and

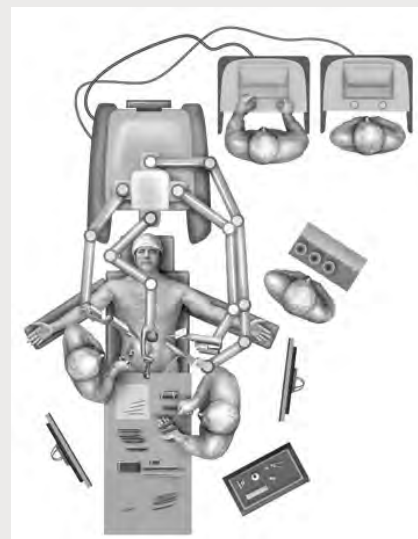


Figure 1. Room setup and patient positioning. Reprinted from Operative Techniques in Thoracic and Cardiovascular Surgery, 18(3), 204-214. Justin Karush and Inderpal S. Sarkaria, Robotic-Assisted Giant Paraesophageal Hernia Repair and Nissen Fundoplication. Copyright (2013), with permission from Elsevier.

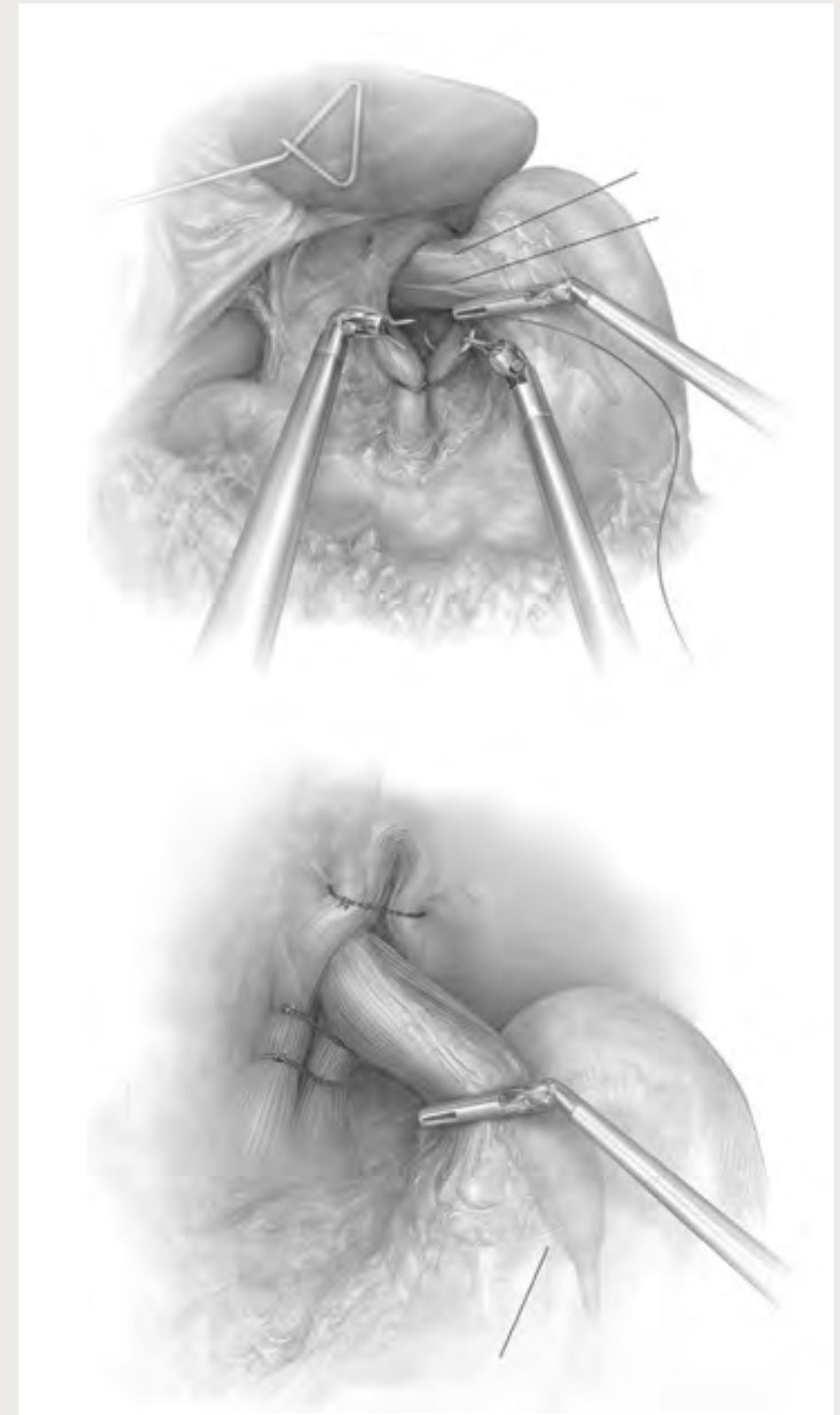


Figure 2. Cruroplasty. Reprinted from Operative Techniques in Thoracic and Cardiovascular Surgery, 18(3), 204-214. Justin Karush and Inderpal S. Sarkaria, Robotic-Assisted Giant Paraesophageal Hernia Repair and Nissen Fundoplication. Copyright (2013), with permission from Elsevier.

Continued...

judgment are critical in selecting appropriate patients for robot-assisted procedures. The surgeon also needs to conduct these procedures safely and to know when to abandon a robotic approach.

The procedures required during GPEH repair (i.e., gastropexy or gastropasty, hernia repair, and fundoplication) are highly suited to the advantages of robotic surgery. The GPEH repair operation requires visualization and manipulation over a long distance in a confined space and near several vital organs that must remain unharmed, such as the aorta, inferior pulmonary vein, and spine. The increased control afforded by the robotic system is particularly helpful under these conditions. The system has fantastic optics and the ability to change views intraoperatively at the touch of a button once the cameras are in place. Additionally, the robotic platform is a fantastic suturing machine (Figure 2). Short-term data from several early, single-institution case series, including a 24-patient series from UPMC, are encouraging and show at least comparable operative outcomes in contrast with traditional laparoscopic procedures.

Robotic GPEH repair can be safely adopted with comparable results by surgeons familiar with minimally invasive repair of paraesophageal hernias.

At UPMC, we have developed advanced stapling techniques using the robotic platform to enable the surgeon to “self-assist.” Because the robotic machine has more than two arms, a single surgeon can place more than two tools at the same time. The surgeon can set instruments for retraction and exposure, and then manipulate the robotic stapler. Small changes in instrument position can be difficult for a bedside assistant to avoid during traditional minimally invasive surgery. In contrast, the robotic arms are very stable. The robotic stapler is immobile once it is applied, resulting in a better staple line with less bleeding. Additionally, the surgeon can focus more thoroughly on the operation, because he or she does not have to “think” for another set of hands and then relay those expectations to the assistant.

Conclusion

Experts at the UPMC Department of Cardiothoracic Surgery, are spearheading pioneering advancements in robotic thoracic surgery, and robotic laparoscopic GPEH repair is an excellent application of this technology. The surgeon can “self-assist” through every step of the operation with excellent visualization in the tight spaces of the patient’s anatomy. Further assessment of robotic GPEH repair is ongoing. As with any complex procedure, we are exploring innovative ways to achieve improved outcomes.

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Join the Division of Gastroenterology, Hepatology, and Nutrition at Upcoming Meetings and National Conferences

2019 Update in Abdominal Transplantation Medicine and Surgery:

This conference will provide health care professionals with an overview of current trends, management techniques, and updates on various topics, including living donor transplants, trends in organ allocation, and caring for patients with end-stage liver and kidney disease.

Friday, May 10 at 8 a.m. to 4 p.m.
The University Club
123 University Place, Pittsburgh, PA 15260

For more information, please contact Susan Piccirilli at piccirillis@upmc.edu.

Digestive Diseases Week:

We look forward to seeing you at DDW in May. Stop by the exhibit hall to meet our experts and learn more about our program.

Saturday, May 18 to Monday, May 20
San Diego Convention Center
San Diego, CA

PancreasFest2019:

PancreasFest is a meeting of pancreas physicians and translational researchers during the last week of July. It is an umbrella conference that supports multiple smaller meetings with focused goals and an overarching pragmatic purpose: facilitating multicenter research.

[dates for PancreasFest]
William Pitt Union
3959 Fifth Avenue
Pittsburgh, PA 15213

For more information, contact Joy Merusi at joj2@pitt.edu.

AASLD 2019 Liver Meeting:

Please visit our booth at the exhibit hall to meet our experts and learn more about our program. We look forward to seeing you in November.

Friday, Nov. 8 to Tuesday, Nov. 12
John b. Hynes Memorial Convention Center
Boston, MA

Division of Gastroenterology, Hepatology, and Nutrition News & Updates



Ramon Bataller, MD, PhD, and Gavin Arteel, PhD, FAASLD, Co-Edit February 2019 Issue of *Journal of Hepatology*, a special issue on alcoholic hepatitis. This issue highlights alcoholic hepatitis advancements, and the co-editors introduce the issue in a compelling editorial introduction, “Alcohol-Related Liver Disease: A Time for Action.”



James B. McGee, MD, Selected as The 2018 Recipient of The Carnegie Science Award, in the University/Post-Secondary Educator award category in May 2018. This award was presented by the Carnegie Science Center in recognition of Dr. McGee’s excellent work and dedication in science and technology teaching. Dr. McGee is an associate professor of medicine with the University of Pittsburgh Division of Gastroenterology, Hepatology and Nutrition. He also serves as the assistant dean of Medical Education Technology and directs the Lab for Educational Technology for the University of Pittsburgh School of Medicine.



Naudia Jonassaint, MD, Appointed as the Department of Medicine’s First Vice Chair for Diversity and Inclusion. Dr. Jonassaint, an assistant professor of Medicine and a transplant hepatologist, will work to develop new initiatives and programs to enhance diversity and inclusion across the department.



David Binion, MD, Receives Sherman Prize for his analysis of “Big Data” to better understand IBD and to optimize treatment strategies for patients with Crohn’s disease and ulcerative colitis. Dr. Binion received this coveted award from the Sherman Foundation during the national Advances in Inflammatory Bowel Disease meeting in December 2018.



Eva Szigethy, MD, PhD, Will Be the Principle Investigator for a \$6.3 Million PCORI (Patient-Centered Outcomes Research Institute) Grant, “Specialty Medical Homes to Improve Outcomes for Patients with Inflammatory Bowel Disease (IBD) and Behavioral Health Conditions.” Dr. Szigethy submitted this grant in collaboration with the UPMC Center for High Value Health Care. She is a professor of Psychiatry, Medicine and Pediatrics and a faculty member with the Division of Gastroenterology, Hepatology and Nutrition, where she directs Total Care-IBD and the UPMC IBD medical home. She is the founder of the Division’s Visceral Inflammation & Pain (VIP) Center.



Randi Sigal and Pamela Wein-Levy Honored at Crohn’s & Colitis Annual Luncheon, by the WPA/WV chapter of the Crohn’s & Colitis Foundation for their outstanding support of IBD patients and patient advocacy, especially for their founding of UPMC’s IBD Connect.

Schistosomiasis: Diagnosis and Treatment

Most commonly found in sub-Saharan Africa, schistosomiasis infects more than 200 million people world-wide. When symptoms present in locales where the infection rarely appears, it can prove an elusive diagnosis. Diagnosis requires an extensive infectious evaluation and the procurement of multiple stool samples.



Hui-Wei (Doris) Chen, MD, is a UPMC Gastroenterology Fellow, Year II in the Division of Gastroenterology, Hepatology and Nutrition.

A 52-year-old female immigrant from Tanzania presented with a three-year history of worsening diffuse abdominal pain. She endorsed fatigue, nausea, early satiety, and weight loss. She denied fevers, vomiting, constipation, and diarrhea. She appeared cachectic and her abdomen was firm, distended, and diffusely tender with splenomegaly. Admission labs demonstrated pancytopenia, normal metabolic panel, mild alkaline phosphatase and total bilirubin elevation, INR of 1.4, and albumin of 3.2. Computed tomography (CT) from her admission is shown in Figure 1.

This patient was diagnosed with schistosomiasis, a disease caused by infection with parasitic blood flukes that live in freshwater snails.



Figure 1. Massive splenomegaly with evidence of portal hypertension and varices.

Diagnosing Schistosomiasis

The prevalence of schistosomiasis is highest in sub-Saharan Africa and more than 200 million people are infected worldwide. Infection occurs when the skin is penetrated by cercariae, the infectious form of the parasite, found in contaminated water. There are five schistosoma species known to cause human infection, though *schistosoma mansoni*, *S. japonicum*, and *S. haematobium* are the most notable. *S. mansoni* and *S. japonicum* cause intestinal tract and liver disease, while *S. haematobium* is associated with genitourinary tract disease.

Acute schistosomiasis syndrome commonly occurs in travelers to endemic regions and is a systemic hypersensitivity reaction to schistosome antigens secondary to deposited eggs. An elevated serum eosinophil count is almost universally present, and initial management consists of inflammation reduction with corticosteroids followed by praziquantel.

Residents of endemic regions develop chronic infection from repeated exposure. The severity of the disease depends on the duration and the number of eggs trapped in tissues. Clinical symptoms often begin insidiously. Chronic infection occurs from the host's immune response to schistosome egg antigens, which leads to granuloma formation at the site of maximal egg accumulation. Diarrhea and fecal blood loss are common.

Schistosomiasis is a leading cause of non-cirrhotic portal hypertension worldwide. Chronic infection leads to hepatic inflammation, portal vein sclerosis, and occasionally, a peri-portal fibrosis known as Symmers' pipe-stem fibrosis, which can be seen radiographically in advanced cases and is characteristic of schistosomiasis. Adult worms are rarely identified histologically, but eggs may be seen. Usually, hepatic function is preserved initially, but as portal fibrosis progresses, increased resistance to portal venous flow can result in portal hypertension, varices, and splenomegaly.

The diagnosis of schistosomiasis is established by detection of schistosome eggs in feces or urine, or antibody detection in individuals with low egg burdens. In some circumstances, rectal or bladder biopsies may be performed.

Treating Schistosomiasis

Praziquantel, the mainstay of treatment, cures schistosomiasis in 85 percent of patients, but re-examination of feces or urine one month after treatment is necessary to assess the treatment response. For those not cured, the parasite burden is generally reduced by 90 percent. In some cases, hepatosplenic schistosomiasis may not be completely reversed. Splenectomy can reduce portal hypertension and improve coagulation factor levels and liver function, and classic and distal splenorenal shunts have also been performed with some success.

Our patient had splenomegaly with CT evidence of portal hypertension and varices (Figure 1). Due to her recent immigration status, an extensive infectious evaluation was performed. A positive schistosomiasis antibody was eventually identified, despite multiple negative stool samples. We were highly suspicious for hepatosplenic schistosomiasis, so praziquantel therapy was administered, and a splenectomy of her significantly enlarged 1,500-gram spleen was performed. The patient experienced clinical improvement and a near resolution of pancytopenia.

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What Is This?

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This patient has a large, dilated thoracic aortic aneurysm, which led to extrinsic compression of the esophagus starting at the level of the mid esophagus. Further history from the patient was notable for dysphagia to solids and prior episodes of food sticking that previously resolved without intervention. He was evaluated by a vascular surgeon, who advised that the patient was too high risk to undergo aneurysm repair. The patient was managed with dietary modifications (i.e., eat soft foods and chew food well) and ongoing control of hypertension. He was counseled to present immediately for signs of recurrent food impaction. Although uncommon, this condition is referred to as dysphagia aortica. Patients who develop an aorto-esophageal fistula may have a prodromal symptom of dysphagia.

Reference:

- Hiller HG and Lagattolla NR. Thoracic aortic aneurysm presenting with dysphagia: A fatal delay in diagnosis. Thorac Surg Sci. 2007 Feb;4: Doc01.

ABOUT THE UPMC DIVISION OF GASTROENTEROLOGY, HEPATOLOGY AND NUTRITION

The Division of Gastroenterology, Hepatology and Nutrition is one of the leading centers for gastrointestinal clinical care and research in the country.

The UPMC Digestive Disorders Center is a comprehensive care program for patients that covers the full range of digestive health conditions including:

- Inflammatory Bowel Diseases
- Cancer Prevention and Treatment
- Functional Bowel Disorders
- Hepatic Disorders and Diseases
- Pancreatic and Biliary Diseases
- Nutrition Support

The Division also includes eight Centers of Excellence that provide specialized care for complex cases and conduct research on numerous fronts to better understand, and develop treatments for, disorders and diseases of the gastrointestinal and related systems.

Centers of Excellence

- Pancreas and Biliary Center
- Center for Liver Diseases
- Center for Intestinal Health and Nutrition Support
- Center for Women's Digestive Health
- IBD Center and UPMC Total Care-IBD
- GI Cancer Prevention and Treatment Center
- Neurogastroenterology and Motility Center
- Visceral Inflammation and Pain Center

To learn more about the UPMC Division of Gastroenterology, Hepatology and Nutrition, please visit UPMCPhysicianResources.com/GI.

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