JOHNS HOPKINS MEDICINE

Physician Update

NEWS FOR PHYSICIANS FROM JOHNS HOPKINS MEDICINE

SUMMER 2016

Making Cancer Patients Whole

Reconstruction efforts are key to successful complex surgeries.

Cancer patients sometimes take a double hit: Not only have they recently received diagnosis of a life-threatening disease, but often their only chance of survival involves surgery that takes something vital away from them—a breast, a knee, even an esophagus or genitals. But that loss doesn't necessarily have to be forever, says **Justin Sacks**, director of oncological reconstruction within the Department of Plastic and Reconstructive Surgery at Johns Hopkins. He and his team work in conjunction with surgical oncologists to replace the form and function that can be lost after cancer surgery.

Their work begins long before patients are wheeled into the operating room, Sacks explains. He and his colleagues attend tumor boards and meet regularly with the host of other specialists they frequently work with, including breast surgical oncologists, thoracic surgeons, general surgeons, orthopaedic oncologists and urologists.

"Anyone taking skin, fat, muscle or bone out of the body calls us," he says.

After formulating a preliminary reconstruction strategy with surgical oncology colleagues to make sure resection and reconstruction can occur seamlessly in conjunction, Sacks and his team then meet with patients in preoperative consultations to relay the plan. Sometimes, operations are textbook—for example, most of the hundreds of breast reconstructions that Sacks performs each year. But depending on the peculiarities of patients' tumors, Sacks and his colleagues might be performing a procedure unlike any a reconstructive surgeon has done before.

"Sometimes when I go to work, I know exactly how I'll do a case. But sometimes, I'm not sure how it will work until I get there," Sacks says. "We need to innovate on the fly, develop new ways to put people back together." Such innovation has involved using a section of small bowel to replace an esophagus, or moving skin, fat and blood vessels from a patient's thigh to reconstruct an abdominal wall.

On the day of surgery, Sacks and members of the oncological reconstruction team scrub in and work alongside their surgical oncology colleagues, resecting tumors and reconstructing tissue in the same procedure. Oftentimes, says colon and rectal surgeon **Jonathan Efron**, Sacks' skill makes it possible to remove tumors that other surgeons might consider not resectable.

"I create some very large defects in patients' abdominal wall or in their perineum, and Dr. Sacks always finds a way to close those holes," Efron says. "He makes it possible for me to take care of people with very complicated cancers. His skill is a key component of the complex cancer surgery performed here."

In the weeks and months after procedures are completed, Sacks follows up with patients, making sure his reconstructions adequately restored not only patients' aesthetic appearance but also their functional needs.

"The most rewarding part of my job is when patients walk into my clinic and tell me they thought they were going to lose who they were, and now they feel natural," Sacks says. "Even though they know they've been operated on, they feel whole again." ■

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— JUSTIN SACKS

CEREBROSPINAL FLUID DISORDERS

Holistic Treatment for **CSF Disorders**

Hydrocephalus and cerebrospinal fluid (CSF) leaks stem from very different anatomical causes. But both fall under the umbrella of CSF disorders, conditions that change CSF pressure and lead to numerous related consequences, including a host of neurological symptoms. Though such disorders—which also include pediatric and adult hydrocephalus, Chiari malformations, pseudotumor cerebri, cerebral and spinal cord cysts, and periventricular tumors—are often treated in a fragmented fashion by different types of providers, concentrating expertise within a single group significantly benefits patients, says neurosurgeon **Mark Luciano**.

"These conditions as a group are difficult to treat effectively," he says. "Patients often lack specialists in CSF disorders with the necessary knowledge to fully address their condition."

That's why Luciano and neurology colleague **Abhay Moghekar** co-direct the Cerebral Fluid Center at Johns Hopkins, where other neurosurgeons, neurologists, pain experts and therapists treat patients together.

One of the most common disorders seen through the program is adult hydrocephalus, explains Moghekar. Though hydrocephalus is often thought of as a pediatric disorder, it's typically treated with shunts, which require lifelong care. Patients who age out of pediatric care can have difficulty finding a specialist willing to follow up with them in adulthood. "We can help patients make a comfortable transition from pediatric to adult care," Moghekar says.

Adults also suffer from a unique type of hydrocephalus known as normal pressure hydrocephalus (NPH). This problem can be difficult to diagnose because its constellation of symptoms—including gait disturbance, urinary incontinence and cognitive problems—are common to many other diseases.

When patients come in for an NPH evaluation, Moghekar says, a physical therapist tests their gait and balance, and a neurologist provides a comprehensive neurologic workup to assess for all potential diagnoses. If NPH is suspected, a spinal tap is performed with quantitative testing of gait and balance to determine if the patient would benefit from a shunt. If the patient's gait improves after the spinal tap, then it's a positive sign that a shunt might provide long-lasting relief, and the patient is referred to Luciano for shunt surgery.

"We truly provide a comprehensive combination of medical and surgical services to treat patients as effectively as possible," says Luciano. ■

Abhay Moghekar, left, and Mark Luciano of the Cerebral Fluid Center at Johns Hopkins, where neurosurgeons, neurologists, pain experts and therapists treat patients together.

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PEDIATRIC GASTROENTEROLOGY

Saving the Child with Short Bowel

Pediatric surgeon **Sam Alaish** recalls this decade-old case like it happened yesterday. The 2-year-old was in shock and being rushed into the OR for an emergency laparotomy. Opening the abdominal cavity, Alaish found one of the worst cases of intestinal malrotation with midgut volvulus that he would ever see. In such cases, he knew, patients have only six to eight hours before the twisted intestine chokes off its own blood supply. By the sight of it, necrosis had already started spreading. He saved what he could, but it was too little—the child would require complex, multispecialty care for most of his life.

"You take out what's dead, leave in what's borderline and come back at a later time to see if it survived to give him his best chance to have the most intestine possible, but we knew he would need TPN [total parenteral nutrition] for a long, long time," says Alaish. "These are kids with severe disease, who need a lot of follow-up and multidisciplinary care to optimize their outcomes."

Sam Alaish, left, and Darla Shores, far right, team up to treat a young patient.

Unfortunately, Alaish adds, there are too few multidisciplinary centers to treat children with short bowel and intestinal failure secondary to congenital conditions, like Hirschsprung's disease, gastroschisis, intestinal atresia and necrotizing enterocolitis (NEC), all of which can lead to disabling and even life-threatening complications. The four-year survival rate for newborn infants on TPN was 70 percent, according to a 2005 report in *Annals of Surgery*, which also noted that in newborns with less than 10 percent of expected intestinal length, the five-year survival was 20 percent. Those numbers have improved with the advent of multidisciplinary coordinated care, but the mortality still remains high, at 7 to 28 percent. Children fortunate enough to survive continue to suffer greater morbidity from both infections and liver failure.

To further improve outcomes, Alaish and pediatric gastroenterologist **Darla Shores** have led development of a new clinic at Johns Hopkins that brings multiple specialists together under one roof to treat such complex patients.

"We know that if you put these very challenging patients in a multidisciplinary clinic, they do better, but not every state has such a clinic for kids," says Alaish.

Adds Shores, "All of the specialists come together, agree on one course of action and present a uniform plan to the family, which should improve patient care and patients' quality of life."

In addition to pediatric surgeons, gastroenterologists and gastrointestinal nurses, staff members at the monthly clinic include a nutritionist, occupational therapist, pharmacist, psychologist and speech pathologist. The strength of Johns Hopkins' long-standing pediatric nutrition program is a value-added feature of the clinic, says Shores, citing the myriad nutritional complications and needs of patients with short bowel.

"After surgery, nutrition is the biggest piece in caring for these children," says Shores. "So the idea of liking food and eating normal meals is not something they grow up with. Getting them transitioned to eating by mouth is definitely challenging."

Another value is having pediatric surgeons like Alaish and **David Hackam**, Johns Hopkins' pediatric surgeon-in-chief who specializes in treating patients with NEC. Also, Alaish and Hackam, along with Shores, are aggressively pursuing research to improve existing treatment protocols and to develop new, more effective therapies for short bowel. They've created the Center for Intestinal Rehabilitation and Cure Using Science, or CIRCUS.

"The reason I came here was to put some science behind our treatments, to measure our outcomes, to see how we're doing and what can we do better," says Alaish. ■

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View a video Q&A with Alaish at: bit.ly/AlaishShortBowel

GYNECOLOGIC ONCOLOGY

When Cancer and Pregnancy Intersect

For Ashley Kulp, a 29-year-old new patient of **Amanda Nickles Fader**, the diagnosis alone was harrowing. Kulp had presented with abdominal pain and bleeding, and examination revealed a 3-centimeter fungating mass on her cervix. A biopsy showed that it was neuroendocrine carcinoma, a rare cervical cancer subtype with a typically poor prognosis. An MRI provided further bad news: There was a second mass growing on Kulp's right ovary, a possible sign of metastatic disease. But complicating the case even further was that Kulp was 29 weeks pregnant with her first child.

In the rare instance when cancer and pregnancy intersect, it creates a very difficult dilemma for patients and those of us who treat them," says Fader, director of the Johns Hopkins Kelly Gynecologic Oncology Service. "We and they have to make some tough decisions about the best treatment options and how to optimize outcomes for both mother and fetus."

Options for Kulp included allowing the pregnancy to proceed without intervention, delivering a course of chemotherapy during pregnancy, or delivering the baby through cesarean section to avoid the fungating mass and performing a concurrent radical hysterectomy. The last option provided the most positive prognosis for Kulp but would increase risk to the baby.

Fader worked with maternal-fetal medicine colleague **Linda Szymanski**, medical director of labor and delivery and inpatient obstetric services at The Johns Hopkins Hospital, as well as others in neonatology and anesthesiology to ensure that Kulp received extensive counseling to help her make the most informed decision.

Kulp elected early delivery and radical hysterectomy. After she received steroids to speed fetal lung maturity, her baby boy, Kayden, was delivered without complication at 30 weeks gestation, and Fader performed a radical hysterectomy and surgical debulking procedure that included staging a biopsy of the lymph nodes and the removal of large tumor masses in the ovary and bowel mesentery.

Ashley Kulp today with her healthy, active 2-year-old, Kayden.

Although each member of the care team agreed that early delivery wasn't ideal, Kayden had an uncomplicated hospital stay, leaving after just four weeks. After surgery, Kulp received an innovative combination of chemotherapy and radiation and also remains healthy, with no evidence of disease recurrence.

"Having the privilege of collaborating with an exceptional treatment team to help women like Ashley Kulp live the best, most meaningful life possible—it doesn't get any better than that," says Fader. ■

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PEDIATRIC PSYCHIATRY

Impulsivity and Binge Eating in Children

In recent years, pediatric psychiatrist **Shauna Reinblatt** began seeing a trend in her clinics: More children with attention deficit hyperactivity disorder (ADHD) are also obese, even though their constant activity and the stimulants used to treat ADHD typically cause these children to lose weight. So, Reinblatt wondered, could impulsive behaviors be triggering binge eating?

Her search for studies on the topic turned up few. There are data linking ADHD and adult binge eating—the most common eating disorder in adults—but there is scant information on children with that problem, which Reinblatt prefers to describe as loss of control eating syndrome (LOC-ES).

Binge eating, she explains, is harder to define and standardize in children of different ages because of growth spurts, which can naturally boost appetites. LOC-ES "means these kids are eating considerably more than their peers and are unable to control what or how much is being consumed," says Reinblatt, who founded the Johns Hopkins Child Psychiatry Overeating Clinic for children and teens.

Reinblatt speculated about possible shared mechanisms in these children, such as impulse control deficits, that are at play in both ADHD and LOC-ES. A recent study she led validates that hunch.

In the study of 79 children ages 8 to 14 whose body mass index was over the fifth percentile, the odds of LOC-ES were increased 12 times for children with ADHD. In addition, children with LOC-ES had much greater impulse control deficits during performance-based neuropsychological tests and on parent reports than children without LOC-ES.

Though the study's findings suggest a link between ADHD and disinhibited eating, Reinblatt cautions that the roots of any underlying connection remain obscure, and longitudinal studies are needed. Children with ADHD who also have LOC-ES might have a more severe form of ADHD marked by more episodes of impulsive behavior. Alternatively, she says, children with both ADHD and LOC-ES could share an underlying risk factor, such as genetic predisposition to impulsivity.

Associated symptoms, such as negative feelings and secrecy, also may play a role in this wider definition of binge eating, says Reinblatt. During her treatment sessions with parents and children, for example, a parent might report that the child regularly sneaks food. "We need to find out if these kids feel guilty or embarrassed about eating," she says.

Ultimately, Reinblatt hopes to identify any underlying mechanism connecting ADHD and LOC-ES "to figure out what's going on in these cases and better understand when to use stimulants, cognitive behavioral therapy or other treatments." In the meantime, she adds, clinicians who see children with ADHD should be mindful that they may be bingeing. ■

"Some studies look at the behavior of bingeing," says Shauna P. Reinblatt. "We look at the syndrome. This is a disorder that has an impact on how the person functions."

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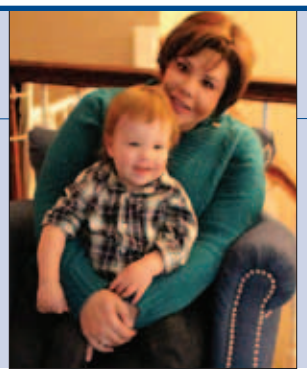

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