JOHNS HOPKINS CHILDREN'S CENTER

Pediatrician

SPRING 2010

Managing Liver Disease

Early detection and constant coordination with the specialists and family are key.

Though Sydney Moss came into the world four weeks early, she appeared pink and healthy. But when pediatrician **Eduardo Fox** first met her in Inova Fairfax Hospital in Northern Virginia a few days after her birth, she looked jaundiced enough for Fox to not take any chances and begin monitoring her liver enzymes. Early detection of pediatric liver disease, he knew, is critical for successful outcomes.

Indeed, the infant's bilirubin levels were high and remained high over the next few weeks, which led to her diagnosis of biliary atresia, a blockage or absence of the bile duct, and referral to the Pediatric Liver Center at Johns Hopkins. There she was able to undergo the Kasai procedure—in which the bile duct is reconstructed with a loop of intestine—well within the preferred timeframe of 8 to 12 weeks. Lindsay Moss, Sydney's mother, credits Fox for this early intervention that reduces the risk of liver failure and the need for liver transplant within the first 1 to 2 years of life.

"These kids can go months before anything is detected, and then it's too late and they have to be transplanted immediately," says Moss. "But Dr. Fox and the staff knew this needed to be closely watched and pushed to have the bilirubin tests done. Since then, he's been our go-to guy."

But aggressively following up on the early signs of liver disease is just one aspect of managing it, notes Fox of Sleepy Hollow Pediatrics, a mid-size suburban practice in Falls Church, Va. Quick referral to a pediatric liver center, as well as constant communication and coordination with the liver specialists and the family, are all key to optimum care.

"I tell the parents that they want to be in a place where their child's problem is something they often see," Fox says. "Then all that needs to happen happens automatically because the systems are already in place. If you're one of a few patients, things get dropped."

Much of the communication surrounding Sydney's care happens virtually among Hopkins Children's hepatologist **Kathy Schwarz** and her staff, pediatrician Fox and the parents. Whether the subject is Sydney's feeding and growth, her latest liver labs, or her risk of infection, all of the participants are in the loop in real time via cell or e-mail. That loop proved vital, notes Moss, when Sydney developed a bacterial colangitis infection while the family was vacationing in Atlanta, some 700 miles from Hopkins Children's. Schwarz and Fox and their respective staffs, whom Moss refers to as "Sydney's Pit Crew," came to the rescue.

"What comforts me most is the access," she says. "We paged Dr. Schwarz on New Year's Day in Atlanta and she responded immediately and arranged for Sydney to be admitted down there.

Falls Church, Va., pediatrician Eduardo Fox with his young biliary atresia patient Sydney Moss and her mom, Lindsay.

With Dr. Fox, it's been constant open communication."

"When she has those acute flare-ups, we have labs done in the hospital and subsequent to her hospitalization to make sure everything is in sync," Fox adds. "Even when she's well, we check her labs every two to three months because of her risk of infection."

In the care plan for a patient with a complex chronic condition like liver disease, all may not be in agreement all the time. For instance, when different opinions surfaced from Fox, Schwarz and an outpatient

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For the Latest on Liver Disease

Pediatric Liver News, a new Hopkins Children's newsletter primarily for pediatricians, reports the latest research findings and developments in diagnosing and treating childhood liver diseases. For a free subscription, visit www.hopkinschildrens.org/ and click "Sign Up for News" in the Quick Links box.

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A Pediatrician's Pediatrician

On March 24 we lost **Henry Seidel**, an inspiring and indispensable force in our institution and pediatric medicine. For many of us who worked with him or trained under him, his death brought back memories of life-changing interactions with a compassionate colleague, a humble mentor, a wise teacher. The reflections from those he touched remind us profoundly of all aspects of our roles as pediatricians, whether we walk each day in academic medicine or private practice. "He was," wrote pediatrician Kai Yang, "our example and our advocate."

As a clinician, Henry Seidel showed us that taking a history and doing a physical exam should always involve the story of a person, not just his chief complaint. Medicine is not an exact science, he noted, because the human condition is marked by variability. In Henry's world, pediatricians were permitted to acknowledge uncertainty. "He made you feel it's okay not to be perfect," said pediatrician Donna Magid.

As a teacher he was "a shaper of wisdom and humanity," contemplated Assistant Dean for Student Affairs Michael Barone. "When I am working with a struggling student, I think about his mastery in such situations."

"He had this way of listening so intently that you knew he had your best interest at heart," echoed Professor of Pediatrics Janet Serwint. "Yet, he also believed that people have their own answer in themselves, and just need a sounding board and some probing questions to help them reach their own solution."

As a mentor, pediatrician Kevin Johnson reflected, his advice transcended medicine. "I can remember discussing the challenges of being a pediatrician who didn't want to spoil his child, while being a father who loved doing so. He assured me that my daughter could only benefit from the attention, and not to let being a pediatrician get in the way of the joy of being a father."

Or, as pediatrician Jane Oski summed it up: "Dr. Seidel taught me more about the human spirit than any single individual in my life. He taught me about grief and acceptance…about strength, hope and perseverance… about entertaining all aspects of a child's illness without losing sight of the things that we pediatricians could impact most directly. He taught me and continues to teach me that the best way to respect life is to embrace it fully."

Amen. ■

Pediatricians pick from a hodgepodge of approaches in managing vesicoureteral reflux, notes pediatric urologist Ranjiv Mathews, with a patient.

Managing VUR to Prevent UTI

How do community pediatricians manage patients with vesicoureteral reflux (VUR)—the flow of urine from the bladder to the kidneys—which may lead to urinary tract infections (UTIs) that in turn can lead to kidney scars and loss of renal function? It's a hodgepodge of approaches, says Hopkins Children's urologist **Ranjiv Mathews**. Some pediatricians immediately send the patient to a pediatric urologist, while others go the entire route in evaluating and managing the patient themselves.

"And then there are the pediatricians in-between who say they'll treat the patient for a year with antibiotics or wait for the patient to have three infections before they even check for reflux," Mathews says. "So it varies in our community."

That variability, Mathews explains, comes from confusion among pediatricians about the most appropriate treatment course for these patients, in part because the research findings vary, too. While current guidelines call for putting all children diagnosed with VUR on a prophylactic dose of antibiotics and then following them with the expectation that the reflux will resolve itself spontaneously, which it does in many children, recent studies suggest that antibiotics may not be very beneficial in preventing urinary tract infections. Also, many children undergo the option of surgery because they develop infections despite antibiotic prophylaxis – raising the issue of antibiotic resistance – or they do not comply with their antibiotic prescription. But surgery as a solution for preventing infection and kidney damage is questionable, too.

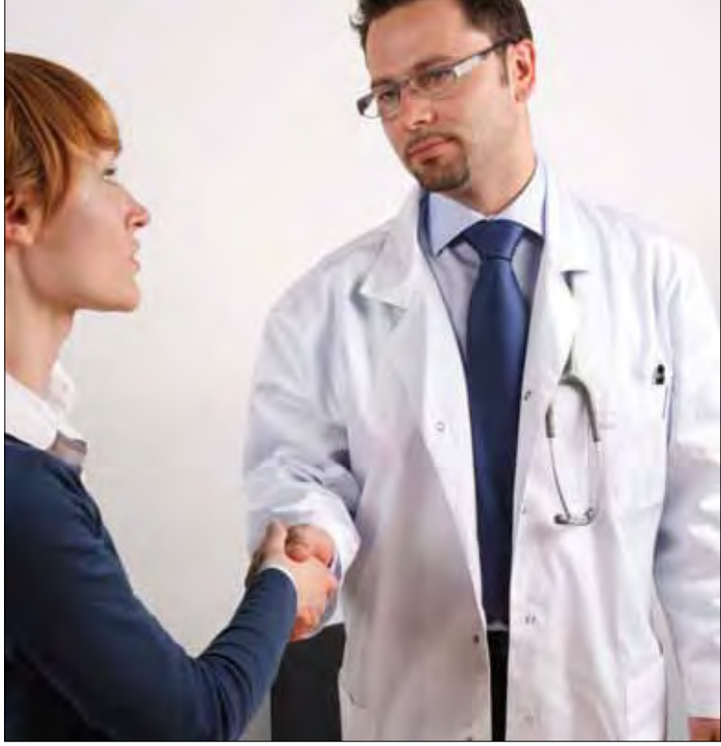
"There's no real study demonstrating that surgery prevents urinary tract infections or changes the eventual course of management," Mathews says point blank.

Confronting this controversy, Mathews says, the National Institutes of Health is launching a two-year trial of 600 patients at 22 medical centers, including Hopkins Children's. Half of the patients will be given antibiotic therapy, the other half placebo, and both groups will be followed up with questionnaires and imaging. Mathews, the principal investigator of the Randomized Intervention for Children with Vesicoureteral Reflux (RIVUR) trial, says its aim is to finally clarify whether antibiotic prophylaxis therapy has any benefit. Also, do these patients on antibiotic therapy tend to develop resistance to other organisms?

"The results of this study," Mathews concludes, "will have a marked impact on the management of all children with vesicoureteral reflux extending far into the future." ■

For more information:
410-955-3693

When Should Pediatricians **Apologize?**



Pediatric resident **Laura Sigman** recalled the case of an adolescent with kidney stones who had been administered twice the amount of pain medicine, which posed a high risk of gastrointestinal bleeding. The resident who ordered the dose rushed to the patient's bedside, relieved that the patient had no abdominal discomfort or signs of blood in his stool. But he was still tormented over his mistake and what to do about it.

"What do patients expect regarding communication about such medical errors," Sigman asked at a recent Hopkins Children's Grand Rounds. "What are common physician practices and beliefs about disclosing medical errors, and how can pediatricians improve communication and outcomes when a medical error has occurred?"

In reviewing the literature on the subject, Sigman found that patients expect the mistake to be acknowledged and explained, and that steps should be taken to prevent similar errors in the future. "They also expect a sincere apology," Sigman said.

Citing a study in the journal *Pediatrics* (December 2005), Sigman noted that 99 percent of parents wanted error disclosure regardless of the severity of the error. Also, the likelihood of legal action decreased with disclosure, with 36 percent of parents saying they were less likely to seek legal action if the error was revealed.

Despite these parental preferences, Sigman noted, physicians face dilemmas. Withholding important information from the patient or others defies a physician's oath, yet admission of an error may be perceived as an acknowledgement of failure. Consequently, while in surveys an overwhelming majority of physicians say they would disclose an error, in real circumstances most do not. In one study, while 95 percent of physicians said they would disclose a hypothetical error, only 41 percent of physicians acknowledged a minor error, and only 5 percent a major mistake (*Journal of General Internal Medicine*, July 2007). Why the discrepancy?

"Physicians want to be open with patients, but they fear embarrassment, damage to their reputation and potential litigation," Sigman said. "There is this notion that physicians must appear infallible to gain patients' trust, and many are not comfortable or lack the skills in presenting bad news."

> "There is this notion that physicians must appear infallible to gain patients' trust." —LAURA SIGMAN, M.D.

While lawyers have advised doctors for decades to make no admissions, Sigman said, legal advice is shifting. Since 2001, disclosure of unanticipated outcomes has been required by the Joint Commission of Healthcare Organizations. Also, there's no evidence that admitting responsibility and apologizing will increase malpractice suits, and some insurance companies are actually encouraging error disclosure. Why? After the University of Michigan Health System adopted a disclosure and compensation policy, Sigman noted, its litigation costs were cut by $2 million a year and new claims by more than 40 percent (*Physician Executive*, March-April 2006).

"Patients are more likely to sue when they feel doctors have not been honest with them," Sigman said. "Apologies can defuse potential medical malpractice cases."

Following up with the patient and family after the disclosure and apology, and explaining what will be done differently in the future, Sigman said, is important, too. Institutions like Hopkins Children's, she added, train doctors how to communicate with patients after an adverse event, But such education has to happen earlier, notes pediatrician **Marlene Miller**, Vice Chair for Quality and Safety at Hopkins Children's, who works with residents on disclosure.

"I hear hesitation on whether it's okay to say 'sorry,' and even hesitation on whether physicians should disclose at all," says Miller. "We need more open education beginning in medical school to create future physicians who are as comfortable disclosing a medical error as they are in writing orders." ■

Managing Liver Disease

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feeding clinic regarding the most appropriate infant formula for Sydney, Moss stresses, Fox brought the group together.

"He e-mailed everybody, asked if we could get on the same page, which I really appreciated," she says. "I don't know if there're many doctors who would try to wrangle all those experts, but we all talked about it and came to the same conclusion about the best path for Sydney."

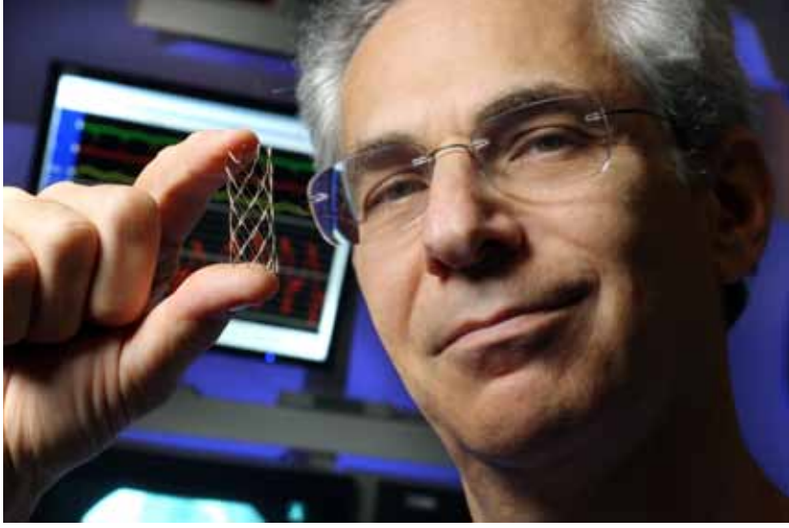
Managing a chronically ill child like Sydney also means being aware of the parents' needs, Fox notes. Even confident, fully engaged parents like Lindsay and her husband, Stephen, may struggle with caring for their child.

"Sometimes you have to remind yourself that it's still scary for parents, even those parents who have the ability to be proactive," Fox says. "I tell them we're going to work on this together, and you don't have to figure it all out."

"Dr. Fox keeps it real for us and looks at the big picture," concludes Moss. "It's reassuring to know someone is always there to help you with your child." ■

Attacking Aortic Coarctation

Pediatric cardiologist Rich Ringel is in search of a safer stent for patients with recurrent narrowing of the aorta.



Always keeping an eye on stents, Richard Ringel, M.D.

As far as pediatric cardiologist **Rich Ringel** is concerned, aortic coarctation, a narrowing of the major artery that leads out of the heart, is an enemy not easy to defeat. Half the newborns with the problem show symptoms in the first few days of life, spurring surgical repair to restore normal blood flow to the aorta and vessels throughout the body. Many of these patients quickly get better, but in about 20 percent of cases the narrowing returns, typically during growth spurts in the middle-school years. Many patients who were missed in early childhood, or with slowly developing coarctation, are also detected around this time, necessitating the need for another complex repair. But in such cases, Ringel notes, many surgeons are anxious about operating a second time.

"There's a lot of scar tissue that's hard to mobilize, and an increased risk of causing injury to the surrounding area," Ringel says. "Surgeons often have to place a tube graft bypassing the narrowed area of the aorta."

As a less-invasive alternative for these patients, pediatric interventional cardiologists like Ringel offer a catheter approach in which they dilate the narrowed portion of the aorta with a balloon and then place a stent to keep it open while the aorta heals. But the stent itself, along with the anatomy of the patient's aorta, poses problems, too. Stents are at risk of fracturing and leading to the formation of aneurysms and, in worst cases, life-threatening aortic dissection—bleeding along the wall of the artery—and rupture of the aorta.

"We're talking about an area of the aorta that is narrow, and we're expanding it under the assumption that all will go well," Ringel says. "Unfortunately, once in a rare while the aorta will just tear instead of stretch and expand. Now you're sitting there with a torn aorta and blood leaking into the chest."

The answer, Ringel notes, lies in the development of safer, more effective stents. Currently there is no FDA-approved stent for aortic coarctation, he explains, and those most often used are biliary stents designed to treat obstructions in the bile ducts. While "not completely unhappy" with these off-label stents, Ringel believes the risks of stent-related complications can be reduced and patient outcomes improved with new devices. That's why he's leading a clinical trial of a new bare metal stent involving 105 patients at 19 medical centers. Outcomes of the Coarctation Of the Aorta Stent Trial (COAST) will be determined by the difference in arm-leg systolic blood pressure over 12 months after the intervention, as well as stent-related complications like fractures.

"This device is stronger and shortens less when you expand it," Ringel says. "We're hoping it will have a lower fracture rate."

Ringel is also enrolling patients in a multi-center trial of so-called "covered" stents, those lined with fabric. These stents in particular, Ringel stresses, may offer the most promise for patients at risk of aortic aneurysms, dissection or rupture.

"We've had multiple patients already whose lives were saved or who avoided major open-heart surgery after rupture of their aorta," Ringel says. "These really are potentially life-saving devices." ■

Research Briefs

Parent's Suicide Raises Child's Risk

Losing a parent to suicide makes children more likely to die by suicide themselves and increases their risk of developing a range of major psychiatric disorders, according to a study led by Hopkins Children's (*Journal of the American Academy of Child & Adolescent Psychiatry*, May 2010).

And because the findings show that parental suicide affects children and teens more profoundly than young adults, it is likely that environmental and developmental factors, as well as genetic ones, are at work in next-generation risk. "Losing a parent to suicide at an early age emerges as a catalyst for suicide and psychiatric disorders," says psychiatric epidemiologist and lead investigator **Holly C. Wilcox**. "However, it's likely that developmental, environmental and genetic factors all come together, most likely simultaneously, to increase risk." The good news, the researchers say, is that though children in this group are at increased risk, most do not die by suicide, and non-genetic risk factors can be modified. Also, there may be a critical window for intervention in the aftermath of a parent's suicide during which pediatricians could carefully monitor and refer children for psychiatric evaluation. Family support is also critical. "Children are surprisingly resilient," Wilcox says. "A loving, supporting environment and careful attention to any emerging psychiatric symptoms can offset even such major stressors as a parent's suicide."

Earlier Better for Cochlear Implants

Receiving a cochlear implant before 18 months of age dramatically improves a deaf child's ability to hear, understand and, eventually, speak, according to a multicenter study led by Johns Hopkins (*JAMA*, April 21, 2010). Believed to be the first nationwide look at the impact of surgical timing on the success of the surgery, which consists of placing into the ear a small electronic device that bypasses the inner ear's damaged nerve cells, the finding points to a critical window for diagnosis and treatment, one that does not stay open long. Delaying implantation, the researchers say, deprives children of essential exposure to sounds and speech during developmental phases when the brain starts to interpret their meaning. "We identified a clear pattern where implantation before 18 months of age conferred a much greater benefit than later implantation, allowing children to catch up fast, sometimes to nearly normal levels," says otolaryngologist and lead investigator **John Niparko**.

A GI Infection or **JRA**?

"It's easy and convenient to put a child like this in this diagnostic box, prescribe the medicines and not think about what else it could be."

– EDWARD SILLS, M.D.

This young patient had some of the classic symptoms of systemic onset juvenile rheumatoid arthritis, or JRA, including unresolved fever over two weeks, and arthralgias and rash that seemed to worsen with the fever spikes. Extensive workups at both an outside hospital and at Hopkins Children's showed pleural and pericardial effusion, elevated inflammatory markers, and a family history notable for a maternal aunt with rheumatoid arthritis—features all consistent with systemic JRA. Also, abdominal ultrasounds and lab tests had already ruled out appendicitis or flu in this young patient.

"There didn't appear to be an infectious process," noted pediatric resident **Alisa Khan** in presenting the case.

But, added pediatric rheumatologist **Edward Sills**, "If you walk out of the conference thinking of this as a typical case of juvenile rheumatoid arthritis, you'll walk out with the wrong idea. It may well be JRA, but there are several things in this case that really are atypical."

One was what the child's parents described as "excruciating" abdominal pain, which Sills noted is seldom seen in JRA. Also, the child's persistent but sporadic fevers didn't fit the predictable pattern of fevers associated with JRA, which typically occur in a precise diurnal pattern.

"When the child with JRA has fever both his rash and irritable personality are flagrantly obvious, but within 3 to 4 hours, regardless of whether or not you intervene, the fever is gone and the child is back to baseline and happy," Sills noted. "Then, at the same time as the previous day, the fever spikes. You can set your clock to it."

Also, while the pericardial effusion was the most alarming concern because of its association with life-threatening cardiac tamponade—in which a large amount of fluid inside the sac around the heart disrupts cardiac function—ultrasound imaging showed that the effusion was very small and not in an area commonly associated with tamponade. Another atypical feature in this case was the child's immediate response to the initial dose of non-steroid anti-inflammatories, which doesn't happen in JRA. Even steroids, Sills stressed, take hours to days to suppress the fevers. The fact that the child's abdominal pain, fever and rash improved may be related to antibiotics he had received over the weeks leading up to and during his admission, pointing to a possible infection after all.

"Indeed, this might have been a GI infection with bacterial origin that may or may not have responded to the antibiotics," said Sills. "But we don't know, and we'll have to follow him for a relatively long period of time."

The take-away for pediatricians? Be aware that all forms of JRA are not the same – they are distinguished by the number of joints involved initially, age of onset, gender predominance, and the severity of presentation. Some forms of JRA, for example, are also associated with anterior uveitis, an inflammatory disease of the eye that may result in vision loss if not properly and promptly treated. Pediatricians, Sills added, might want to think outside the box about other possibilities besides JRA, which tends to be generalized and over-diagnosed.

"Importantly, probably half or more of the children who are so-labeled with JRA don't have it," Sills noted. "It's easy and convenient to put a child like this in this diagnostic box, prescribe the medicines and not think about what else it could be. If an illness is diagnosed as 'atypical X,' it is very likely not 'X.'" ■

Research Briefs

Anemia, Race and Kidney Disease

Black children with chronic kidney disease have more severe anemia than white children even when they receive the same treatment, according to a multicenter study led by Hopkins Children's (*American Journal of Kidney Disease*, May 2010). The findings suggest that inherent biological differences, rather than access to care and treatment, may be at play, raising the question whether current guidelines for anemia treatment should be tailored to reflect race. Anemia is diagnosed by measuring levels of the protein hemoglobin, which carries oxygen in and out of red blood cells. Hemoglobin levels below 11 grams per deciliter of blood generally indicate anemia, but the number is adjusted for a child's age and gender. In the new study, black children with kidney disease had lower hemoglobin than white children, 0.6 grams per deciliter on average, and a greater proportion of black children were anemic when compared with white children. "As we move from one-size-fits-all medicine toward individualized medicine, we should study further racial disparities and, perhaps, adjust hemoglobin targets to reflect what appear to be genetic variations," said pediatric nephrologist **Meredith Atkinson**, lead investigator of the study.

For more information about this and other studies, visit www.hopkinschildrens.org and see "News & Events."

Surgeon Jeffrey Lukish Brings Innovation to the Table

It's no surprise that pediatric surgeon Jeffrey Lukish's favorite operation is video-assisted thoracoscopic ligation of a patent ductus arteriosus in low-birth-weight infants using a novel retractor, so much so that he wrote a paper on the approach (*Journal of Pediatric Surgery*, May 2009). After all, the surgery is innovative, minimally invasive and technically challenging, and the patient an extremely small infant. Using innovation to make the impossible possible in pediatric cases—and with the least amount of pain, discomfort and recovery time as possible—is what Lukish is all about.

"The one common denominator in the procedures I do is innovation," says Lukish, "to reduce pain, hospitalization, drug usage, operative time and post-operative time. It's a triple win—the child is happy, the family is happy, and I'm happy."

Now a surgeon at Hopkins Children's, the former chief of pediatric surgery at the National Naval Medical Center and Walter Reed Army Medical Center has developed and refined a number of minimally invasive surgical techniques in addition to thoracoscopic PDA ligation. Colorectal and thoracic anomalies, recurrent inguinal hernias, gastrocutaneous fistulas—you name the condition and Jeffrey Lukish will find a patient-friendlier approach.

"General pediatric surgeons typically have not been the ones to do these operations," Lukish says, "but I love the breadth of cases."

The son of an engineer, Lukish says innovation is in his DNA. But much of his motivation, he adds, dates back to his childhood years in Virginia watching Army surgeon Hawkeye Pierce operate on young wounded soldiers in the TV series "MASH." So it was only natural that he'd later attend the Naval Academy, major in medicine, and find himself deployed as a family practitioner/general surgeon to places like Guantanamo Bay and Okinawa—austere environments where he had more autonomy than resources, where the concept of finding better ways to provide care kicked in.

"I very much enjoyed being a physician in the military, it broadened my experience," Lukish says. "It allowed me to discuss problems and come up with solutions with a whole different set of people. Communication and teamwork, you realize, are more important than the environment."

During his years in the Navy, Lukish envisioned himself as a transplant surgeon, until he rotated through pediatric surgery at the National Naval Medical Center. "Working with the pediatric surgeons, seeing the children they cared for and their problems, I realized I wanted to be a pediatric surgeon," Lukish says. "Interfacing with the parents and the child really sealed the deal." ■

"The one common denominator in the procedures I do is innovation to reduce pain, hospitalization, drug usage, operative time and post-operative time. It's a triple win—the child is happy, the family is happy, and I'm happy."

– JEFFREY LUKISH, M.D.

Dr. Lukish sees patients at Greater Baltimore Medical Center in Towson and at Hopkins Children's. For referrals and appointments at GBMC, call 443-849-6275; for appointments at Johns Hopkins, call 410-502-6649.

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