JOHNS HOPKINS CHILDREN'S CENTER

Pediatrician

FALL 2013

Reflections on a Half-Century Practice

When Columbia, Md., pediatrician **Will Standiford** first hung out his shingle he worked long hours, diagnosed patients with his hands, performed his own spinal taps, and, because health insurers didn't cover office visits, sometimes got paid in vegetables and whiskey.

With a young patient, community pediatrician Will Standiford.

What was pediatrics like when you opened your practice?
When I started in 1966 we charged $4 for office visits and $7 for house calls. We'd see patients in the morning, schedule a block of time in the afternoon for house calls, then come back and see patients into the early evening. We saw patients on Saturday mornings, too, and on Sunday mornings for urgent cases.

Why so many hours?
At that time there were no answering services, no pediatric EDs, no hospitalists. If you got a call in the middle of the night you'd see the patient. If they had a condition like croup or epiglottitis, which could completely close off the windpipe, you had to see them because there was no alternative. In those days these kids would have to undergo tracheotomy so they could breathe. We didn't have the vaccines we have today.

Did you see patients at the local hospital as well?
Yes, we admitted our patients and took care of them at St. Agnes, our primary hospital. We'd communicate with the residents, see patients on rounds in the morning and sometimes later in the day, depending on how sick the patient was. One of us in our four-physician practice would be on-call overnight for our patients in the hospital, and then be expected to work the next day and possibly the next night. It was a demanding schedule but we were used to a demanding schedule. Now we have office hours until 6 p.m., send patients to the pediatric ED or an urgent-care center, and admit our patients to hospitalists, which has taken a lot off our load.

Were you doing procedures you don't do today?
We were seeing our newborns in the hospital, but there was no neonatal intensive care unit. One issue we had to deal with was RH incompatibility, which required exchange transfusions to remove and replace the patient's blood. Exchange transfusions were frequent and took a lot of time, and we were the ones who did them. We also performed our spinal taps in the ED, but I haven't done one in the past 20 years.

Any other significant changes?
Radiology has changed our practice a lot, too. If one suspected the child had a tumor, a neurosurgeon had to put the kid to sleep, make a burr hole in the head, put a needle through the brain into the ventricle, aspirate the spinal fluid and inject air. Then, by manipulating the air and fluid levels around the ventricular system, one would attempt to identify where the tumor was. We could order arteriograms back then, too, to detect problems in blood vessels around the brain, heart, kidneys and lungs. When I first saw a CT scan I couldn't believe what I saw. It was all there—everything you always wanted to see.

> "I didn't particularly enjoy taking care of geriatric patients and I didn't have the personality to be a good surgeon. I enjoyed working with kids—I still do."
>
> – WILL STANDIFORD, M.D.

Do you see any downside to such technological advances?
With more information gained in electronics and imaging, there's less physical diagnosis today. When I was a resident, a senior doctor, who was an excellent diagnostician, said you don't need an EKG, just look at the AVC (atrial ventricular contraction) wave in the neck. We said, 'What?' but we learned you can see the venous wave in the neck, which indicates how the heart is functioning. So there were

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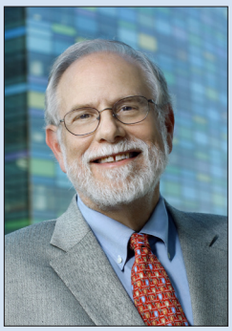

George Dover, M.D.
Director, Johns Hopkins Children's Center Given Professor of Pediatrics

A Doctor's Touch

A recent TED talk by physician writer **Abraham Verghese** reminds us in this era of modern medicine that much of the art of medicine is still in the hands. He makes the case that when we shortcut the physical exam and lean toward ordering tests, we not only overlook simple diagnoses that can be made at an early, treatable stage but also risk losing a ritual at the heart of the patient-physician relationship—in his words "the power of the human hands, to touch, to comfort, to diagnose."

Our cover interview with long-time community pediatrician **Will Standiford** also reminds me of the power of palpation and percussion in diagnosis—and how so much of what pediatricians used to do regularly has been succeeded by developments in imaging, among other advances. We're grateful for innovations, some of which come to us daily and serendipitously—like pediatric dermatologists **Bernard Cohen** and **Kate Puttgen**'s research of the hypertension drug propranolol as a swift remedy for severe infantile hemangiomas. Another surprise is pediatric gastroenterologists **Maria Oliva-Hemker** and **Suchitra Hourigan**'s fecal transplant clinic for patients infected with the drug-resistant bacterium *C. difficile*. Who would have thought?

We do have to wonder what else will come our way. Interestingly, we are moving from a field focused on detection of existing disease to one focused on diagnosis of disease before it becomes apparent. In the process we may prevent the onset of complex and common adult disorders. The genetic code is only part of the puzzle—how the genome reacts to the environment is another. What about proteomics? Will we be increasingly identifying biomarkers as sources for early detection of disease? We can't help but wonder at such possibilities—we also can't help but feel grounded by the Will Standifords who personify the simple transcendent truth of pediatric practice. As he concluded, "I enjoyed working with kids—I still do." Now, that's a nice touch.

Call Center Centralizes Scheduling

The Children's Center's now offers a one-stop shop for scheduling all pediatric visits.

In the past, a community pediatrician or parent would have to make three different telephone calls to schedule three individual appointments for one patient with three separate specialists. No more!

Now referring physicians and parents have one central source, the Call Center, and a single telephone number—443-997-KIDS (5437)—for scheduling appointments with Children's Center pediatric primary care physicians and specialists. Several area physicians interviewed said they had encountered substandard service in the past when scheduling appointments for their patients at Johns Hopkins. The Call Center, under the data-driven leadership of Patient Access Manager **Latisha Smith**, is moving to change that perception.

"The goal is that callers are not kept waiting, not going to voicemail, and not holding for long periods of time. We want to get scheduling done in a timely fashion," says Smith. "We've set up phone metrics where 80 percent of our calls have to be answered within 30 seconds, and less than 3 percent of our calls end up being abandoned (where the caller hangs up). We're also saying 'Thank you,' getting callers' names and telephone numbers, getting them to the right physician at the right location."

Callers, Smith explains, receive a standard greeting that triages their call to the pediatric primary, specialty or surgery service. Once they've made their choice, a Call Center agent takes the call from there, following the protocol for the particular service selected. Quality assurance monitoring assures that agents follow protocols in a professional manner, are asking the right questions and closing the call with a re-cap or summary of the scheduled appointment.

> "Patients and referring physicians now have a central line to call to get the patient seen."
>
> – LATISHA SMITH, PATIENT ACCESS MANAGER

In the past, Smith says, referring physicians and patients trying to schedule appointments would call individual divisions at the Children's Center, which had their own unique way of managing appointments. Under the Call Center that process is now uniform for most—and under the rollout eventually all—Children's Center divisions.

"There were so many different mechanisms in trying to reach a physician," Smith says. "Patients and referring physicians now have a central line to call to get the patient seen."

For non-appointment related calls, Call Center agents will facilitate a "warm transfer" in which they stay on the line and relay information to the appropriate division before handing the call back to the patient. "We communicate the patient's needs so he or she doesn't have to keep saying the information over and over five times to five different people," says Smith.

The Call Center is accessible from 8 a.m. to 5 p.m. Monday thru Friday. For referrals in which the pediatrician needs to consult with a Children's Center doctor, Smith notes, physicians should call the Hopkins Access Line, or HAL, at 410-955-9444, or 1-800-765-5447. ■

AT CME

Congenital Heart Disease Symposium

Focusing on "Diseases of the Cardiac Valves from the Neonate to the Adult," the 14th Annual International Symposium on Congenital Heart Disease is being held Feb. 14-18, 2014 at All Children's Hospital Johns Hopkins Medicine in St. Petersburg, Fla. For more information or to register, email cme@allkids.org.

Treating the Tricky Spinal Vascular Malformation

Following her treatment at Johns Hopkins Children's Center for a large spinal vascular malformation, "Madalyn is moving perfectly," says her mom, Meredith Carter.

Born five and a half weeks early, Madalyn Carter was admitted to the NICU at a Washington, D.C. hospital, where neonatologists quickly noticed she wasn't moving her arms. An MRI revealed the cause—a spinal vascular malformation (SVM) compressing the newborn's spinal cord and cutting off circulation to her brain. Madalyn was at risk of suffering a severe stroke, doctors told her mother. Worse, they had no treatment to offer.

"We were devastated," says Meredith Carter. "It was like a ticking time bomb from which she might end up paralyzed."

Seemingly with nowhere to go, the Fulton, Md., mother remembered that Johns Hopkins pediatric neurosurgeon **Ben Carson** was a member of her church. She contacted Carson, and he referred Madalyn to pediatric neurosurgeon **Rafael Tamargo** and interventional neuro-radiologist **Philippe Gailloud**, who have deep experience in treating these rare but lethal lesions.

"Experience is important," says Gailloud. "With it, you have seen these lesions before and you know what you have to deal with."

"Most doctors, even neurologists and neurosurgeons," says Tamargo, "will only see a handful of spinal vascular malformations in their entire careers."

Also critical to a good outcome, say Tamargo and Gailloud, is an early and accurate diagnosis. Undetected and untreated, these lesions can grow, bleed and even kill, says Gailloud, noting that dramatically increased pressure in the venous system can cause hypertensive cardiomyopathy.

"When the spinal cord circulation fights against such high venous pressure, everything starts to dilate, and the child gets weak and loses bowel and bladder function," explains Gailloud. "These very big vascular malformations compress the spinal cord like a tumor, resulting in a bleed or stroke."

Tamargo and Gailloud strive to see patients the day after every patient inquiry, but getting the right diagnosis can also be delayed by the challenge of imaging these conditions. Some spinal fistulas often aren't visible on MRI, and the ones that show up on CT scan require a shrewd radiologist to spot them. Definitive diagnosis requires a spinal angiogram—a specialized imaging technique often eschewed by doctors. When this technique was invented in the late 1960s, Gailloud explains, it was blamed for spinal infarctions and other serious complications. Spinal angiograms have become much safer with decades of experience, and Gailloud and his colleagues perform two or three of these procedures each week. In a recent paper they described spinal angiogram outcomes on 350 patients.

"There were no significant complications," Gailloud says. "It turns out to be a very safe procedure."

Once they diagnose an SVM, Tamargo and Gailloud formulate an individualized treatment plan. Most patients can be treated endovascularly by embolizing their problematic blood vessels with coils or glue. For those whose anatomy or conditions don't allow such treatment, surgery is the next best option. Madalyn's case was even trickier. Her SVM—a large perimedullary venous fistula—was on the interior side of her spinal cord rather than on the back of the neck, making access through the femoral artery the only option. In such cases, says Gailloud, it's important to stage the procedures.

"If you try to treat the whole thing in one session, the lesion may get so angry that it begins to swell and damage the spinal cord," says Gailloud.

Madalyn underwent her first embolization at 9 months of age, her second at 15 months, both times with no complications. "Within a day of her first procedure she started to move her arm—it was incredible," says Carter. "Now at 4 and a half years old, Madalyn is moving perfectly. You would never know anything was wrong." ■

For more information, call 410-614-1533.

Pediatric gastroenterologists Maria Oliva-Hemker, left, and Suchitra Hourigan with a fecal transplant patient.

A Fecal Transplant Clinic?

Call it therapeutic poop, if you will, but the best hope yet for an effective treatment of childhood infections with the drug-resistant bacterium *C. difficile* may come straight from the gut, according to recent research. Parlaying that research into practice, the Johns Hopkins Children's Center has launched a fecal transplantation program for patients with recurrent diarrhea caused by what they say is a wily pathogen that is increasingly impervious to drugs and a rapidly growing problem among children and adults.

"Fecal transplantation—or the transfer of 'good' bacteria from the colon of one person into the colon of another—should be considered for all children with *C. diff* infections who don't respond to two standard courses of antibiotics," says pediatric gastroenterologist **Maria Oliva-Hemker**.

Physical Therapy for Cancer Therapy

Faith Tittle was a "completely normal child" until one month after she turned 4, notes her mother, Deborah Howe of Odenton, Md. The usually active child started suffering fevers, fatigue and frequent vomiting. Her pediatrician suspected a virus but after a few more days with no improvement in her symptoms, Howe brought her to the emergency department at a hospital in southern Pennsylvania, where she was visiting relatives. There she discovered her daughter had leukemia. If that diagnosis wasn't bad enough, Howe then learned firsthand that the very treatments designed to curb the cancer's growth—chemotherapy, radiation therapy and two bone marrow transplants—would take a toll as well.

"She was feeling very sick and extremely weak, she didn't want to get out of bed," says Howe. "It was hard to get her up and moving."

This is where pediatric physical and occupational therapists step in, with the goal of improving patients' strength, balance and endurance, as well as fine motor and cognitive skills. To facilitate independence and a smooth transition back to the community and school, occupational therapists help patients focus on daily living skill. But these goals are challenging as patients like Faith may suffer severe deficits from weeks and even months of inpatient and outpatient cancer treatments, explains **Hallie Lenker**, a physical therapist at Johns Hopkins.

"Facing balance problems and body weakness that arise during cancer treatment, they experience a lack of energy and ability to participate in the things they would be doing in the community," says Lenker.

Deborah Howe notes that her daughter had to relearn how to walk after her second bone marrow transplant. Three-times a week physical therapy got her back on track.

"It took a while but she was really tough, and in a couple of months she was walking again on her own," says Howe. "The physical therapists were extremely helpful in figuring out ways to motivate her."

Part of the therapists' work involves tracking deficits caused by cancers and cancer treatments to better predict the type of deficits patients will experience. That knowledge helps them tailor physical and occupational therapy to the patient's individual needs.

"Of course, we're thankful when these children are doing well medically, but we haven't thought enough about the deficits these treatments cause in strength and endurance," says physical therapist **Julie Quinn**. "Now we're looking specifically at measures for balance and endurance, and collecting data to determine specific progress for patients who are in and out of the hospital, which will help us develop a more cohesive program for both inpatients and outpatients."

Having a comprehensive physical therapy (PT) and occupational therapy (OT) program for both pediatric inpatients and outpatients in the new Charlotte R. Bloomberg Children's Center building helps, notes Quinn. Physical and occupational therapists are familiar with the patient's in-hospital experience and their specific PT/OT needs, which helps ensure a seamless transition to outpatient care. Also, PT and OT appointments can be scheduled easily to coincide with follow-up medical appointments.

Howe agrees: "It's nice for your child to have the same place for inpatient and outpatient care, to have that familiarity and not have a lot of change going on."

And where does she see Faith today?

"She is very resilient, she is a miracle—she's pulled through many times and has surpassed doctors' expectations," Howe says. "She's surprised us all." ■

> "Of course, we're thankful when these children are doing well medically, but we haven't thought enough about the deficits these treatments cause in strength and endurance."
>
> **JULIE QUINN**
> PHYSICAL THERAPIST

Faith Tittle, with a therapist, works on her balance.

Such beneficial bacteria work by keeping rogue players in check, Oliva-Hemker explains, so any shifts in gut environment—such as ones caused by antibiotics—can have dire consequences. When good bacteria are killed off by antibiotics, the bad guys multiply causing an imbalance or "dysbiosis," Oliva-Hemker says. Typically, gut infections caused by one antibiotic are treated with another one to eradicate the overgrowth of harmful pathogens, but drugs often fail to do so fully or permanently because they only treat part of the problem.

"When we administer an antibiotic to treat the *C. diff* infection, we destroy some of the bad bacteria, but that does not address the other half of the problem—the loss of good bacteria that might have led to the infection to begin with, so we never truly restore the balance in the gut and often the diarrhea returns with a vengeance in a matter of weeks," says pediatric gastroenterologist **Suchitra Hourigan**.

The concept of treating poop woes with poop is hardly new. The method originated with ancient Chinese healers who gave their diarrhea-ravaged patients "yellow soup," a concoction of fecal matter and water. Nowadays, fecal transplants are often performed during a colonoscopy, and improvement can be seen in as short as two weeks, as beneficial bacteria start to repopulate the patient's gut, Hourigan says. Studies in adults show that more than 90 percent of patients are cured following such therapy and, experts say, they have every reason to believe the numbers would be equally impressive in children.

Johns Hopkins Children's Center is one of only a handful of pediatric hospitals in the country to offer this therapy for a condition that can cause dehydration, anemia and pain, and can seriously affect a child's quality of life, leading to absence from school. ■

A Standard of Care for Hemangiomas

In clinic, pediatric dermatologists Cohen and Puttgen.

First, in 2008, pediatric dermatologists **Bernard Cohen** and **Kate Puttgen** experienced firsthand what some French doctors had discovered serendipitously in treating infants with heart problems who also happened to have infantile hemangiomas—the hemangiomas swiftly faded after the infants received the hypertension drug propranolol. After administering the drug to their first patient, who had large hemangiomas threatening her airway and vision, Cohen and Puttgen reported a similar effect.

"Within 48 hours, the hemangiomas became softer," Puttgen says.

"The hemangiomas were just a fraction of their initial size, and the eye complication completely resolved," adds Cohen.

Parlaying their findings, Cohen and Puttgen collected more evidence showing the efficacy of propranolol for infants with function-threatening or severely disfiguring hemangiomas. In their retrospective analysis of 70 infants treated with propranolol, 51 patients had fair to marked improvement in their hemangiomas, whereas 17 percent had minimal improvement. The single reported complication, hypoglycemia, occurred in a patient with a viral illness who recovered. It seemed the two pediatric dermatologists had found a solid alternative for the standard therapy at the time, steroids, known for their serious side-effects.

But there was another issue the two pediatric dermatologists had to deal with—the requirement that infants under 1 year of age be admitted to the hospital for administration of the drug and monitored because propranolol poses the risk of lowering their blood pressure. But after finding only two cases with complications among some 450 infants treated with propranolol in the past five years, Cohen and Puttgen moved ahead with an outpatient protocol for infants 2 months and older. Newborns under 2 months of age, as well as those with comorbidities, they note, still need to be admitted.

Under the outpatient protocol, Cohen explains, the child receives the first dose of propranolol in clinic and vital signs are checked before, during and after administration of the drug. Parents, meanwhile, get instructions on how to give the drug and monitor the child for side effects as the dosage is gradually increased over 10 days. The protocol was launched eight months ago. The results?

"We've had no problems. Zero. None of the kids had symptoms and there was no further drop in blood pressure after the first dose," says Cohen. "The response to treatment is good or better than systemic corticosteroids, which was the standard of care before 2008."

So, does all this mean propranolol is the new standard of care for severe hemangiomas?

Noting that there is no official standard of care—no U.S. Food and Drug Administration approved agents—for treating infantile hemangiomas, Cohen suggests the door is now open for propranolol, an already FDA-approved drug.

"We're not quick to adopt new treatments but we were one of the first centers in the United States to initiate this treatment and now we're very comfortable doing this," says Cohen. "This is a big change in how we approach complex and severely disfiguring hemangiomas, and a big deal for pediatricians, patients and their families. In my mind, this has become the standard of care." ■

Research Briefs

Low Vitamin D Levels Raise Anemia Risk

Low levels of the "sunshine" vitamin D appear to increase a child's risk of anemia, according to new research led by investigators at the Johns Hopkins Children's Center. The study, published online Oct. 10 in the *Journal of Pediatrics*, is believed to be the first one to extensively explore the link between the two conditions in children. The researchers caution that their results are not proof of cause and effect, but rather evidence of a complex interplay between low vitamin D levels and hemoglobin, the oxygen-binding protein in red blood cells. The investigators say several mechanisms could account for the link between vitamin D and anemia, including vitamin D's effects on red blood cell production in the bone marrow, as well as its ability to regulate immune inflammation, a known catalyst of anemia. "If our findings are confirmed through further research, low vitamin D levels may turn out to be a readily modifiable risk factor for anemia that we can easily tackle with supplements," says pediatric nephrologist **Jeffrey Fadrowski.** ■

Child Born with HIV Still in Remission

A 3-year-old Mississippi child born with HIV and treated with a combination of antiviral drugs unusually early continues to do well and remains free of active infection 18 months after all treatment ceased, according to an updated case report (*The New England Journal of Medicine,* Oct. 23, 2013). Early findings of the case were presented in March 2013, but the newly published report adds detail and confirms what researchers say is the first documented case of HIV remission in a child. "Our findings suggest that this child's remission is not a mere fluke but the likely result of aggressive and very early therapy that may have prevented the virus from taking a hold in the child's immune cells," says lead author and pediatric HIV expert **Deborah Persaud.** ■

Reflections on a Half-Century Practice

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things he saw on physical exam that we didn't see because we would order an EKG. Today, many physicians are no longer putting their hands on the patient.

Has that influenced your practice?

We don't have an X-ray machine in this practice, so in our clinical clerkship students make a diagnosis of pneumonia by listening and looking. Here, everything is hands-on.

Do you enjoy teaching?

Teaching has become more important for me. Also, I appreciate the insights of the younger students and residents while sharing my experiences with them. I don't want to be the pediatrician who's forgotten everything.

Do you see any changes in the way parents interact with you?

Parents are more savvy today because they can go online and have a whole differential diagnosis ahead of time. That's fine. I kind of enjoy that because you have a basis for discussion rather than a dictation of what you think. That may threaten some parents but it shouldn't because it's helpful, though sometimes it creates anxiety for them before they get a chance to talk about it.

Any observations about health insurers over the years?

When I first started practice insurers only covered hospital stays and not office visits, so many times I got paid in vegetables and whiskey. In recent years, insurance companies have presented a complicating factor in the practice of medicine for patients as well as physicians. Many times they try to dictate patient care. Today, I leave a lot of this hassle to the younger people on our staff.

Anything you would change?

No. I enjoy the practice of pediatrics today as much as ever, maybe more. I get to teach the residents, and hear case presentations and didactic talks with community experts. I always thought pediatrics was a good fit for me—I didn't particularly enjoy taking care of geriatric patients and I didn't have the personality to be a good surgeon. I enjoyed working with kids—I still do. ■

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