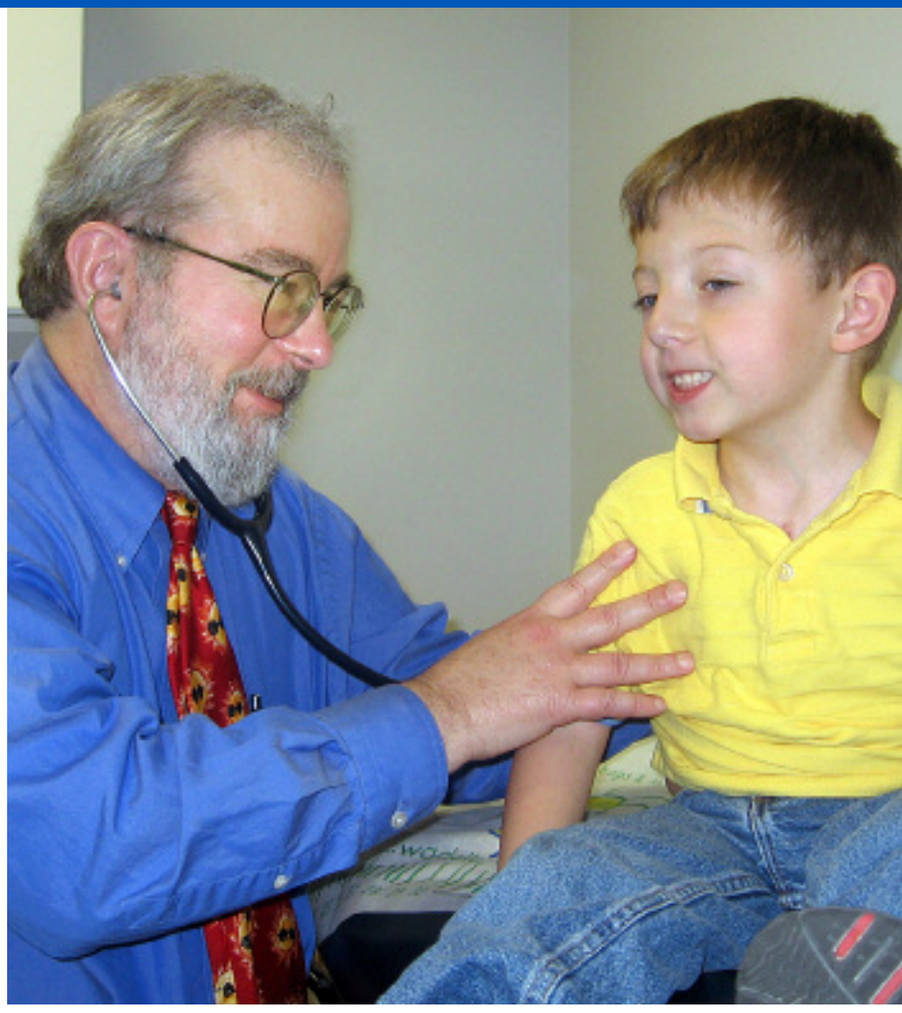
JOHNS HOPKINS
CHILDREN'S CENTER

Pediatrician

SUMMER 2006

Saving a Blue Baby

AN EASTON PEDIATRICIAN AND CHILDREN'S CENTER CARDIOLOGIST GET TO THE HEART OF A COMPLEX, EMERGENCY CASE.



Pediatrician Brian Corden with patient Chase Marine, now 5.

Within hours of Chase Marine's birth in December 2000, he had begun to turn blue. His saturation levels—the percentage of oxygen in his lungs—were in the low 70s, prompting Easton, Md., pediatrician **Brian Corden** to order oxygen. But when the infant didn't improve, and X-rays showed no signs of pneumonia, Corden was mystified: "There was no apparent reason why he wasn't doing well. We didn't see any lung disease."

But the bluish color in the newborn's abdomen and chest made Corden suspect a serious problem with the heart. He immediately had the infant rushed by ambulance to Johns Hopkins Children's Center, which turned out to be a life-saving decision. Indeed, an echocardiogram revealed that the left side of Chase's heart had not fully developed, an extremely rare condition known as hypoplastic left heart syndrome. His left ventricle was so tiny that it was unable to pump blood returning from the lungs to the body. Without immediate surgery, he would die.

In a series of complex operations called the Norwood procedure, Hopkins surgeons re-rigged Chase's cardiovascular network to restore blood flow throughout his body, using the healthy right ventricle to pump what the left ventricle could not. The operations went well but not entirely without complications. In April 2005, Chase began turning blue again as oxygen levels in his blood declined due to venous malformations—abnormal and enlarged veins—in his lungs. Surgery fixed the problem, but the setback showed how patients like Chase are more at risk of pulmonary infections as well as other serious problems like ventricular dysfunction because one ventricle is doing the work of two. On top of that, Chase faced a clotting disorder that needed treatment, a twisted colon that required surgery, and club feet, which meant another operation. How has pediatrician Corden managed this complex patient?

"They take care of the big stuff, we follow him in between," Cordon quips, acknowledging the close collaboration with Children's Center cardiologist **Philip Spevak.** (See "Managing the Child with Heart Disease," p. 3.) He adds that he monitors Chase more often than most of his other patients because normal pediatric illnesses pose a greater threat to him. The Easton pediatrician's biggest concern is a serious respiratory virus like influenza, which could be fatal for a patient like Chase. In Spevak's mind, however, Corden has already saved Chase's life. He stresses that the signs of this disorder could have been easily missed—they typically don't show up until a day or two after birth, when a fetal heart valve called the patent ductus naturally closes. In patients with hypoplastic left heart syndrome, the valve actually keeps them alive.

"If the child goes home and the patent ductus closes, the blood flow will be compromised even more, and the baby will get sicker from end-stage organ damage," says Spevak. "Dr. Corden recognized the signs and prevented the baby from dying."

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

Collaboration Is Key

Our cover story this issue is about Easton, Md., pediatrician **Brian Corden's** speedy transfer of a newborn with a life-threatening heart disorder to pediatric cardiologists and surgeons here at the Children's Center, and the effective co-management of that child by Dr. Corden and Hopkins physicians. This story, the experience they shared, underscores the collaborative role community and academic pediatricians play in diagnosing and treating children today to reach the best possible outcome for the patient. As technology changes—producing paperless medical records, Web-based lab records and more efficient scheduling—these collaborations should improve even more. But I worry about the lack of sufficient beds and physicians to meet the demands of our pediatric partners and their patients for our services.

Interestingly, as we publish this issue of *Pediatrician,* we're celebrating the groundbreaking of a new children's hospital and the opening of a new pediatric outpatient facility, the Harriet Lane Child Health Building, on our East Baltimore medical campus. Our pediatric specialists plan to open their new clinics to your patients in the fall; our new hospital's ribbon cutting is planned for 2009. With these new facilities will come even newer technologies and advances in diagnostics and therapeutics borne from the research bench and our clinical experience. The shunt that was used in our young heart patient's surgery is named after Hopkins' pioneering surgeon **Alfred Blalock** and pediatric cardiologist **Helen Taussig.** Similarly, the research and clinical work we're doing today may result in a new operative procedure or treatment that saves the life of a patient tomorrow. State-of-the-art facilities and all that they bring help that happen, but not without our shared knowledge and close collaboration with you, the community pediatrician. Thank you and enjoy this issue!

Identifying Infections Before Hospitalization

Pediatrician Chris Lehmann linked hospital databases to identify infected patients.

It was a nurse from another unit, who happened to catch a glimpse of the patient, who sounded the alarm on the fourth floor of the Children's Center. She recognized the child and his troubling diagnosis from a prior hospitalization. Unbeknown to the charge nurse or shift coordinator, the newly re-admitted patient, now sharing a double room, had vancomycin-resistant *enterococci,* or VRE. An infectious pathogen, *enterococcus* is resistant to most antibiotics. Resistance to vancomycin—the antibiotic of last resort—meant the child's infection was untreatable. VRE does not pose a risk to healthy individuals but presents a serious threat to hospitalized, immune-compromised children. In this patient's case, thanks to the nurse from another unit, appropriate infection control and isolation measures were initiated and the infection was contained.

"The outcome could have been far different," stresses pediatrician **Christoph Lehmann,** who was notified of the safety lapse.

To prevent another such occurrence that might not be caught, Lehmann, director of medical technology at the Children's Center, designed within 24 hours a computer program linking databases from the Children's Center admitting office and the division of pediatric infectious diseases. Cross-referencing patients' medical record numbers, the system identifies children who have a known chronic infection. The system then automatically pages the shift coordinator and e-mails the charge nurse, alerting them that such patients are en route to the hospital.

"We're now considering screening new and returning patients for respiratory cases and instituting universal precautions for all patients," says Lehmann, a noted developer of patient-safety technologies. A next step may include notifying caretakers of children who have a new onset of infection, alerting them quickly to infection trends. ∴

A Painless Solution for Spondylolysis

Using bone grafts to seal fractures, orthopedic surgeon Paul Sponseller helps patients to be active without pain.

The pain in Carrington Croft's lower back began in the fall of 2004. By that winter, it was constant. Croft, an avid hockey, basketball and lacrosse player, had a stress fracture. Seems the Virginia 16-year-old was born with a lower vertabra that couldn't take the athletic workouts. Worse, none of the treatments—from heat packs to physical therapy—worked.

"I was so miserable I had to stop playing basketball that winter, just two weeks into the season," says Croft.

But then she found Children's Center orthopedic surgeon **Paul Sponseller,** who diagnosed Croft with spondylolysis, one of the most common causes of chronic back pain and spasms in young athletes. Spinal fusion between the lumbar vertebra and the sacrum is the most typical surgical repair, but it reduces flexibility and requires lengthy immobilization and hospitalization. An internal rod and screws are sometimes used to hold the vertebrae together as the fusion heals. None of this sounded too appealing to Croft.

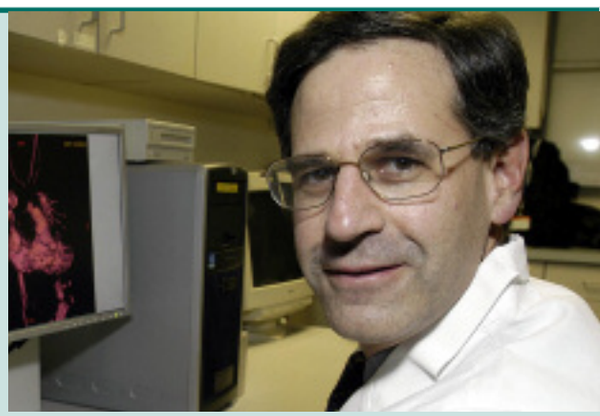
Instead, Sponseller proposed repairing the fracture itself. He packed and bridged the fracture with healthy bone from the iliac crest, the bony ridge at the top of the pelvis, using the bone graft as a natural glue to seal the fracture. Two stainless steel screws held everything in place.

"This approach allows active teenagers like Carrington to go back to competition without any stiffening of their backs," says Sponseller. "Movement causes pain, so eliminating that movement by getting that little crack to seal—and the vertebra to reestablish itself—eliminates the pain. Because spinal fusion wasn't needed, she didn't lose any mobility."

Croft agrees: "Three or four weeks after the surgery I was swimming. Moving again was heaven. I went on to play varsity basketball and then lacrosse, pain-free."

For more information, call 410-955-3136.

Philip Spevak, M.D.
Managing the Child with Heart Disease



What's the current trend in management of heart disease by pediatricians?
Because of an increased supply of specialists, pediatricians are increasingly being separated from the co-management of specialty problems, including heart disease.

And the impact of that?
Primary care docs are losing their skills in caring for these patients, which translates into more expensive—and not necessarily better—health care.

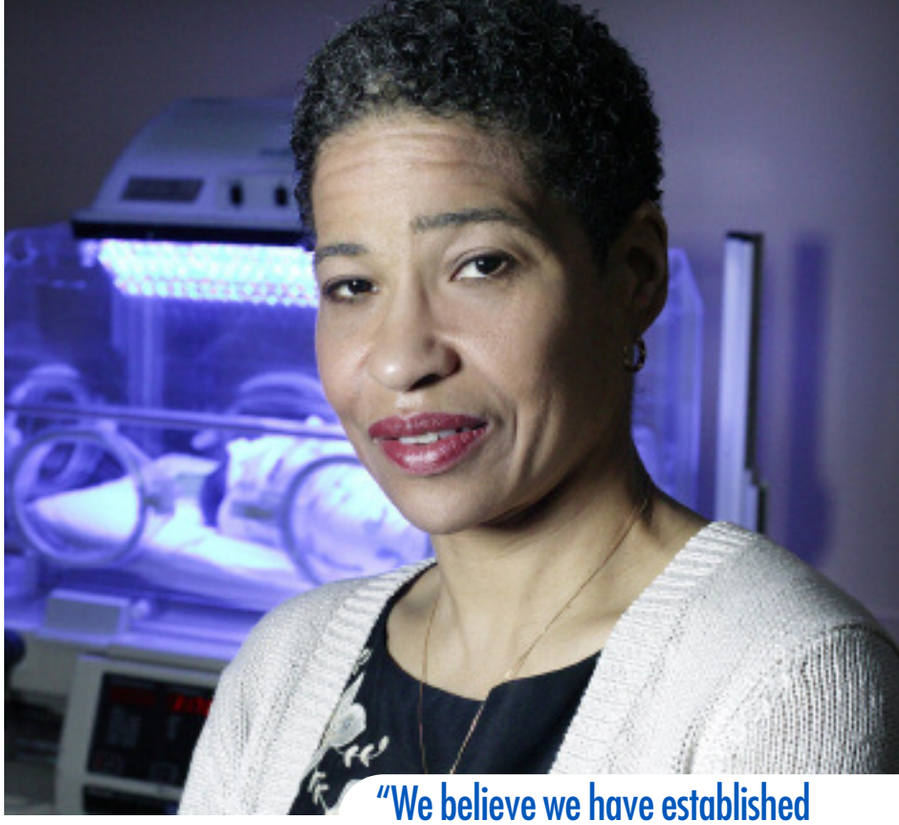
What can pediatric cardiologists do?
Educate pediatricians beyond the recognition of the disease, incorporate the pediatrician into co-management, and emphasize the advantage of an increased primary care role.

And what is that role?
In heart disease, it can be many things—monitoring the patient, adjusting feedings and medicines, assuring that noncardiac care is still happening, and recognizing when medical management of heart disease is failing. Pediatricians are also important in preventing heart disease by screening for hypertension and other risk factors of premature atherosclerotic disease. In some cases, it is appropriate for the pediatrician to order diagnostic tests, including an echocardiogram, when there's a family history of cardiomyopathy.

When is it inappropriate for a pediatrician to order such imaging?
When the patient requires sedation, when there's a high likelihood of an abnormality, and when there is uncertainty as to the experience of an imaging lab in examining children with heart disease (for example, imaging a disorder like Kawasaki's disease when the echo lab is inexperienced in imaging coronary arteries).

Any down sides to a greater primary care role?
Sure. Pediatricians can overuse resources or get in over their head. That's why it's important for the pediatrician and pediatric cardiologist to collaborate in managing the patient with heart disease.

Easing Withdrawal for Addicted Infants



"We believe we have established a new and better protocol for treating these infants."
—Estelle Gauda, M.D.

Approximately 200 babies a year at Johns Hopkins spend their first weeks or months of life in the hospital withdrawing from drugs they were exposed to in utero—heroin and methadone—and often need opiates to calm down. After years of treating these little patients, primarily with tapering dosages of oral opiates to quell their hyperactive symptoms, neonatologists **Estelle Gauda** and **Alex Agthe** decided to explore therapies that would shorten these infants' exposure to opiates and get them home sooner. Now they think they may have one.

With NIH and FDA approval, Gauda and Agthe launched a large study of the drug clonidine in combination with tapering doses of diluted tincture of opium (DTO) to determine whether the treatment can significantly reduce patients' length of hospital therapy and opiate exposure. Clonidine has been used in conjunction with opiates for several years to effectively treat symptoms of opiate addiction in adults. How does it work?

An alpha 2-adrenergic receptor agonist, clonidine reduces the excessive autonomic symptoms created by the withdrawal itself by blocking the release of large amounts of noradrenaline from cells in the brain during withdrawal. In the treatment study, of 80 infants exposed to heroine or methadone either in utero or after birth, the 40 infants who received the combination showed more rapid improvement and required lower dosages of DTO than those who received only DTO. Their median length of therapy was 23 percent shorter, and they were able to go home, on average, five days sooner.

"We believe we have established a new and better protocol for treating these infants," says Gauda of the combination therapy, the first NIH-approved use of clonidine to treat moderate and severe neonatal abstinence syndrome.

Research Briefs

SCRATCH Insect Bites

Insect-bite rashes can mimic a variety of symptoms and are often misdiagnosed, resulting in extensive and expensive testing. But a new set of guidelines developed by Children's Center researchers should help pediatricians recognize the source of the rash early on (*Pediatrics* online, July 2006). Called SCRATCH, the guidelines follow criteria like the **S**ymmetry of the rash, **C**luster pattern of the bites, whether **R**over or another pet was a culprit, the **A**ge of the patient, whether the lesion has a **T**arget or bulls-eye shape, parental **C**onfusion over what's going on, and the number of family members in the **H**ouse affected. "Common sense might tell us that fleas and mosquitoes would affect other members of the family, but we must keep in mind that these rashes develop in children who have hypersensitivity that others do not have," says pediatrics resident **Raquel Hernandez,** who helped develop the tool. "These guidelines are really intended to make pediatricians consider insect-bite hypersensitivity as a diagnosis and think twice before referring a child for a skin biopsy or another invasive procedure," adds pediatric dermatologist **Bernard Cohen.** "If the rash fits the SCRATCH criteria, it's likely bug-borne."

Screen for Autism

Few Maryland and Delaware primary care pediatricians regularly screen their patients for autism and autism-spectrum disorders (ASD), according to a Children's Center study that found while 82 percent of pediatricians regularly screen their patients for general developmental disorders, only 8 percent test for ASD. Physicians said they failed to screen for ASD because of unfamiliarity with the screening tools or lack of time. Screening is essential for early detection and treatment of autism, which has grown over the past decade to an estimated 12 to 40 cases per 10,000 children. "This study suggests that current national efforts may not be sufficient to actively promote the use of ASD screening tools in the general pediatric practice," says **Susan dosReis** of Child and Adolescent Psychiatry. "So it is important to learn what some obstacles might be and what needs to be done to overcome them."

Handling Adolescent HIV, **Ethically**

Pediatrician **Maggie Moon** had seen cases like this before. A young teen comes in through the ED, presents with all the signs of PCP—pneumonia related to HIV—but there's no record of the virus in the patient's history. That's because the child's pediatrician, at the urging of the mother, who has HIV, had not ordered testing after the child was born. If and when the child got sick, like now, they would have her tested.

After this scenario was presented at a recent case conference, some residents in the room angrily asked, "But shouldn't the community pediatrician have forced the issue, pushed for testing over the mother's objections? Couldn't the child have been treated earlier to prevent the complication she was now facing? Weren't the child's best interests compromised?"

Such questions are the stock of Moon's trade. An attending pediatrician and specialist in bioethics, she was on service the day this Washington, D.C., teen was admitted, and she saw in the case illuminating teaching points for the team caring for the child. While she understood the residents' irritation over the pediatrician's inaction, she realized the community doc's perspective, too.

"I can see a wise and thoughtful pediatrician saying, If I push this mother too hard she will take this child and never come back—then I won't even have access to this child when she does get sick," Moon says. "So the smart thing is to keep encouraging testing but maintain contact with the family to foster the child's health."

Learning to manage a relationship with parents in a child's best interest is one ethical issue; how to inform the child of his or her HIV diagnosis is another. What are your goals in this disclosure, Moon asks. Who should be present? How do you bring a parent on board who is trying to protect her child from the stigma of HIV? In such cases, Moon stresses, it's important for the child, who is typically terrified of the unknowns, to get a good sense of what's happening.

"The whole idea is to promote health in the long run so the patient doesn't feel overwhelmed, so she doesn't fall into a depression but sees it as something she can manage," Moon says.

> "The smart thing is to keep encouraging testing but maintain contact with the family to foster the child's health."
>
> —Maggie Moon, M.D.

In such cases, Moon also consults with pediatrician **Nancy Hutton,** director of the Intensive Primary Care Clinic at the Children's Center, where children with HIV receive care. Hutton sees an increasing need for community pediatricians to address these issues, too. While new HIV treatments have resulted in a dramatic decrease in newly infected newborns, HIV in women hasn't declined—so the potential for passing on HIV infections is still high. Just as adult practitioners ask about HIV, pediatricians need to ask the same questions, too.

"Pediatricians should know whether the mother was tested or not, and if she wasn't tested they should write that down," Hutton says. "They should ask about HIV as part of the routine family history, just as you would ask about cancer and diabetes. If it's on the list, it's not a stigmatizing question."

At CME

Pediatrics for the Practitioner 2006

Bipolar disorders, childhood obesity, food allergies, headaches and newborn screening—these are some of the commonly encountered problems that will be covered at the Children's Center's annual **Pediatrics for the Practitioner Update,** September 28–29, at the Thomas B. Turner Building on the Johns Hopkins Medicine campus. Through other sessions, participants will learn how to carry out a comprehensive pre-college evaluation, screen and manage lipid disorders, use antibiotics rationally in managing otitis media, and diagnose and manage common sports injuries. *For more information or to register by phone, call 410-955-2959.* Register online at *www.hopkinscme.net.*

The Times They Are A'Changing

After 26 years as a general pediatrician, **Ken Schuberth** told his patient families last spring that he was ending his general practice this summer; he'd decided to devote his attention to his specialty, allergy and asthma. Their response, he says, was professionally gratifying, but personally wrenching.

"Parents expressed appreciation for all of our years together," says Schuberth, an associate with the Maryland Pediatric Group (MPG) in Lutherville, Md. "We'd become family, part of one another's lives. 'So surely,' they'd tell me, 'when you said you'd no longer be seeing general patients, you couldn't mean *us*?' I had not expected they would feel about me the same way I felt about them."

In the end, the increasing demands of a dual practice and the changing complexities of each specialty forced a choice. Spending three-quarters of his time as a general pediatrician and three-quarters as a pediatric allergist "just didn't add up," he says, to a balanced life. As the number of kids with allergies and asthma was increasing, he was running out of hours in the week to accommodate them.

"I came to understand that I had a certain amount of energy I wanted to devote to work, and a certain amount for the rest of my life, a hard realization for us physicians. I decided to alter the pace."

Still seeing patients, but strictly as an allergist, says Ken Schuberth.

Some observers say immunology is more straightforward and less emotionally draining than general pediatrics, with its multitude of psychological, societal and developmental issues. Nevertheless, immunology demands all of Schuberth's experience, as well as his appreciation for the role families play in helping allergic kids stay healthy. "The number-one reason people with chronic illnesses are not successful in the care of their illness is the lack of compliance with their medicine," he says. "Kids are no exception."

A graduate of Johns Hopkins School of Medicine, Schuberth's training at Hopkins included a pediatric chief residency and a two-year fellowship in allergy and immunology, tailor-made for him. He saw patients in the specialty clinics at Hopkins, while in his private practice he saw only the occasional allergy case. Food allergies were especially rare, unlike today when allergies account for nearly a third of all cases in a general pediatric practice.

Like Schuberth, who himself has allergies ("a prerequisite for the specialty," he quips), most of the affected children have a strong genetic predisposition. A "hygiene hypothesis" is gathering steam as an explanation for their growing numbers. It theorizes that when our immune systems are drawn toward fighting infection, they don't have time to make allergies. Plunk young kids in a modern environment that is relatively infection-free and their developing immune systems will drift to the other side. Studies show, Schuberth notes, that kids who live on farms or in families with numerous children rarely develop allergies.

If this hypothesis is correct, a new generation of allergy shots, now being tested in animals, may hold promise. The shots include not only specific allergens, but small pieces of proteins from bacterial cell walls. The idea is to turn on infection fighting and turn off the allergy. "It has never been done, but seems to be working," says Schuberth.

A better means of controlling allergic symptoms could not come too soon.

"Before long," Schuberth observes, "there's going to be more allergy business for docs than we're equipped to handle." ∴

JOHNS HOPKINS CHILDREN'S CENTER Pediatrician

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