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CHILDREN'S CENTER

Pediatrician

FALL 2011

Our Medical Home

Annapolis Pediatrics didn't set out to develop a medical home practice model—it just happened.

Annapolis, Md., pediatrician Jim Rice with his young patient, Kaira Quesada, and her mom, Ave. Born with chronic lung disease of prematurity, Kaira benefits from Rice's medical home approach for patients with special healthcare needs.

Pediatrician **Jim Rice** notes that the members of Annapolis Pediatrics didn't decide to sit down one day and draw out a plan to develop a medical home practice for their most medically complex patients. But changing dynamics in pediatric medicine like decreased hospital stays and increased community services made the need for a medical home model even more essential. Consequently, in a step-by-step and somewhat serendipitous approach, they've created the components of a medical home.

"The hospital was really the center of the medical home 30 years ago for kids with special healthcare needs," Rice says. "Today there's much more being done in the outpatient realm, which had certainly posed a challenge for us community pediatricians."

Those challenges also include today's typically busy, high-volume practice. While the majority of Rice's patients come with bread-and-butter pediatric issues like the flu, summer injuries and swimmer's ear, mixed in with that caseload is the recent heart surgery patient dealing with a G-tube, a tracheostomy and a dozen meds daily.

"The challenge for me and most pediatricians," Rice explains, "is how do we handle that at 3 in the afternoon when the waiting room is full?"

Among the steps Annapolis Pediatrics took was creating a new staff position to coordinate follow-up services for the more medically complex cases, like the patient who needs a referral to pediatric pulmonary or scheduling for an upper GI. The practice also purchased a faster, more accurate and more efficient electronic patient record system, which helps track medications and labs.

Citing a recent hypoplastic left heart syndrome patient who needed prescriptions filled for an upcoming trip, Rice notes, "With a few clicks I was able to pull up his doses and quickly fax them at the end of the visit with less chance of errors."

Another challenge for the pediatrician is sustaining the role of central coordinator of care for the patient and family. With so many entry points for healthcare services for the recently discharged hospital patient with special needs—home care and rehab, multiple subspecialists and the local hospital—what can the primary care pediatrician do to act as the hub?

"The availability of home care and other services has allowed a lot of these patients to go home, which is good, but we run the risk for some of these patients not having a home," Rice says. "Are they interacting with their specialist from Hopkins? Are we the point of contact? Sometimes patients can become somewhat triangulated between our office, the subspecialist and home care."

Electronic communication helps, says Rice, though for him and others the practice of emailing personal health information raises flags about patient confidentiality. Rice still relies mostly on the traditional mode of communication with patients—the

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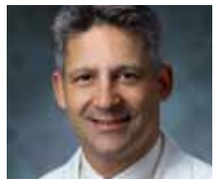

George Dover, M.D.
Director, Johns Hopkins Children's Center Given Professor of Pediatrics

Making a Medical Home

So, "How do you handle the medically complex case at 3 in the afternoon when the waiting room is full?"

That question, raised by Annapolis pediatrician **Jim Rice** in our cover story this issue, has likely been asked by all community pediatricians at one time or another. He gives us some answers via his practice's somewhat spontaneous approach to making a medical home, and Hopkins Children's pediatrician **Barry Solomon** fills in the rest on the subject in his research review of the benefits and challenges of employing a medical home model.

Communication with specialists is key to a medical home, and being informed of their latest diagnostic and therapeutic services helps, too. In this issue, pediatric cardiologist **Priya Sekar** and pediatric urologist **Ming-Hsien Wang** show us that diagnosis of complex medical conditions can't happen soon enough. See their page-3 stories on how they use prenatal imaging to screen and treat as early as possible conditions like congenital heart disease and urethral valve disorders.

At another point along the spectrum of care these patients will begin to make the transition from pediatric to adult care. Read on page 4 how Hopkins Children's cardiologists **Jane Crosson** and **Rich Ringel** guide them on this journey. And on page 6, pediatric specialists **Jeffrey Lukish** (surgery), **Christine Karwowski** (gastroenterology) and **Peter Mogayzel** (pulmonary) talk about their new collaborative clinics with Anne Arundel Medical Center in Annapolis.

Consistent with the original aim of this newsletter, this issue is about sharing our experiences—and insights gleaned from them—in pediatric medicine. Please enjoy this issue.

Advances in neonatal medicine are increasing survival in low birth-weight babies, but they're also increasing the incidence of pulmonary hypertension.

Treating Pulmonary Hypertension Earlier

Pediatric pulmonary fellow **Bridget Stuart** described the complex case of an infant born at 27 weeks, intubated on mechanical ventilation for 5 days, transferred to a chronic-care facility on oxygen and diuretics, discharged home on oxygen, and then, at 3 ½ months of age, transferred to the Johns Hopkins pediatric intensive care unit after suffering symptoms of bronchopulmonary dysplasia (BPD), an upper respiratory infection and respiratory distress. Imaging showed severe pulmonary hypertension, which led to the infant's death.

"Unfortunately, mortality in patients with pulmonary hypertension is not uncommon," Stuart shared at a mid-June conference at the Mt. Washington Pediatric Hospital. "But I hope this case piques your curiosity more about how to care for these complicated patients."

There were many minds to pique—some 40 community cardiologists, intensivists, neonatologists and pulmonologists who had been invited by a multidisciplinary team of Hopkins Children's specialists to strategize and share ways to combat what appears to be an increasing incidence of pulmonary hypertension in premature infants with BPD. Advances in medications and mechanical ventilation in the neonatal intensive care units have increased survival of low-birth weight infants, but they've also introduced a new type of BPD characterized by inflammation, oxidative stress, pulmonary hypertension, and, consequently, greater mortality.

"We're now seeing this comorbidity of pulmonary hypertension where 20 years ago these children didn't live to discharge," said pediatric pulmonologist **Sharon McGrath-Morrow.** "We know that kids who have BPD without pulmonary hypertension have a much higher rate of survival than kids with BPD and pulmonary hypertension."

Noting that there are no clinical guidelines for treating pulmonary hypertension in infants with BPD, McGrath-Morrow called on the group to screen "at-risk" children. Early risk factors associated with pulmonary hypertension may include birth weight below 500 grams and a history consistent with pulmonary hypoplasia.

Echocardiogram, McGrath-Morrow noted, may be considered as a screening tool in preterm infants. Other tests include brain natriuretic protein (BNP), which may be useful in following response to treatment in infants, and high resolution CT and CT angiogram, which can rule out pulmonary vein stenosis. Cardiac catheterization should be considered if pulmonary hypertension is suspected but echo is not diagnostic, or if severe pulmonary hypertension is present but the patient is not responding.

"This was our first attempt in letting the community know we see this as an emerging problem associated with very high morbidity and mortality," McGrath-Morrow said. "The more aggressively and earlier you treat the lung injury, the better your outcomes."

For more information, call 410-955-2795

Getting to the Heart of the Problem

Pediatric cardiologist Priya Sekar with her young patient, Elliana, and mom, Jenni Wilson.

At her 12-week ultrasound, Jenni Wilson was concerned when her Howard County obstetrician referred her to Hopkins for fetal echocardiography. Then she was stunned when physicians there told her they saw signs of congenital heart disease. The mother of two with a husband in the Air Force in Korea knew she had a tough road ahead.

"It was very rough with my husband being overseas at the time," Wilson says, "but we had help from our church and people around us."

Things got even tougher, however, when the pediatric cardiology group detected the specific problem—tricuspid atresia, a disorder in which the tricuspid valve is missing or abnormally developed and blocking blood flow to the right ventricle. But Hopkins pediatric cardiac surgeons offer a three-step surgery for the condition, which they and the family could prepare for thanks to the early diagnosis.

"We can identify at 18 to 20 weeks a major congenital heart lesion or a significant anatomic defect that's going to require surgery, which is a real benefit," says **Priya Sekar** of Hopkins Fetal Cardiology Program. "Prenatal diagnosis allows us to help families wrap their mind around the diagnosis, make pregnancy decisions, and plan for delivery and care."

When should fetal echo be done? Criteria include family history, maternal diseases like lupus known to affect the fetal heart, and signs of fetal mid-line defects like cleft palate, which are associated with congenital heart disease. Following up with fetal echo, pediatric cardiologists use ultrasound machines similar to those used by obstetricians, though their focus is the structure and function of the heart. "What do the valves and the chambers of the heart and the blood flow to and from the heart look like?" Sekar says.

Such findings may not only trigger preparations for surgery but also therapeutic strategies in the neonatal intensive care unit (NICU) following delivery. Medicines given in the NICU can keep blood vessels open, preserving fetal physiology to protect the neonate. Heart rhythms can be controlled by giving the mother anti-arrhythmics that cross over to the fetus. Excessive fluid in the fetus, or Hydrops fetalis, can also be countered in intensive care. All of these interventions suggest that delivery should be in an academic medical institution to avoid transporting a fragile newborn.

A prenatal diagnosis may also be life-saving. Citing a case in which fetal imaging showed triscuspid atresia, transposition of the tricuspid valve and coarctation of the aorta, pediatric cardiologist **Phil Spevak** says, "Without the prenatal diagnosis, a critical fetal blood vessel would have closed, cutting off perfusion to the body and resulting in renal failure, liver failure and brain damage."

But isn't it a shock for parents to learn that their next child may have a serious heart problem?

"It's true if you didn't know you would worry less," says Sekar. "On the other hand, the family has more time to become more comfortable about having a child with heart disease."

Wilson, for one, says she is glad she knew. Her daughter, Elliana, now six months of age, had her first procedure—insertion of an artificial shunt to maintain blood flow to the lungs—a few days after birth. "She has good coloring, and her oxygen saturations are now appropriate," says Wilson. "Elianna is doing amazing. You would think she's a completely normal baby." ■

Catching a urethral valve problem early

Pediatric urologist **Ming-Hsien Wang** recalls the case of a newborn boy with a peeing problem—no urine output—that wasn't picked up the first few days of life. Meanwhile, his serum creatinine levels kept going up and up, signaling possible kidney malfunction or even failure. Being born in a community hospital didn't help, as there was no neonatal intensive care unit (NICU) for close monitoring or nephrology service to consult. He was transferred to Hopkins Children's where Wang pinpointed the problem pretty quickly.

"We saw on ultrasound signs of urine backup, thickening of the bladder wall, dilated urethra, and hydronephrosis, or swelling of the kidney," Wang says. "The usual cause is posterior urethral valve."

The condition, Wang explains, is an obstructive membrane in the urethra that occurs in utero exclusively in males. Posterior urethral valve, or PUV, varies in degree, though severe cases can result in renal failure, respiratory failure from lung underdevelopment as a result of low amniotic fluid volumes, and loss of the fetus in utero. That's why it's important, Wang stresses, to detect the disorder early, ideally through prenatal sonogram. If signs of PUV are present, a pediatric urologist can visualize and resect the valve during the first few days of life by utilizing a scope with a tiny camera.

"You can see these little translucent flaps of tissue blocking the urethra, which in normal fetal development should never have happened," Wang says.

In the past, one treatment option was fetal surgery to divert urine from the bladder directly into the amniotic fluid, but the risks to the fetus proved too high. Today, pediatric urologists like Wang first use tubes to drain the bladder as soon as the baby is born, and then once anesthesia is safe—with the endoscopic camera and a tiny scalpel—cut the membrane.

"We did the resection and his creatinine came down," Wang says about her recent case. "He had mild hypertension due to the kidney problems but now he's doing pretty well."

For more information, call 410-955-6100.

When Congenital Heart Disease Patients **Become Adults**

At the age of 12, Tyler Littleton of Parkville, Md., wasn't about to let his diagnosis of congenital heart disease slow him down. Hand him a baseball glove, lacrosse stick or soccer ball and he was off and running. But when he reached adolescence and his condition bumped up from mild to moderate aortic stenosis—a narrowing of the aorta that reduces blood flow from the heart—pediatric cardiologist **Rich Ringel** steered him away from contact sports. Littleton was devastated.

"I was pretty good in those sports and looking forward to playing in high school," Littleton says. "Not playing lacrosse was a big disappointment."

At the time, however, Littleton was beginning to transition from the pediatric to the adult clinic for patients with congenital heart disease, which helped cushion the blow. There patients like Littleton learn more about their particular condition, the risk factors presented by young adulthood, and how to independently manage their disease.

"We involve them more in their care while they're still in high school, before they face myriad issues like birth control, employment, health insurance, marriage and sexuality," notes pediatric cardiologist **Jane Crosson**. "Their disease may require some modification of those issues."

Crosson and Ringel point to Littleton, now 18, as a patient who has adapted well. After reviewing with Ringel sports appropriate for patients with his condition, and undergoing a stress test, Littleton joined the volleyball team at his high school, Calvert Hall, where he was named to the all-conference team. This past summer he worked as a lifeguard at a local pool, and is now a freshman at High Point University in North Carolina, which poses other issues.

"Dr. Ringel advised me about college life and knowing my limits and not testing my limits, which means avoiding alcohol and activities like binge drinking," Littleton says. "But I think I've matured with my heart disease, knowing it's something I have to deal with. I know what my condition is."

Littleton will also be taking with him a Hopkins health passport—containing all of his medical records and echocardiogram test results—prepared by cardiovascular nurse practitioner **Kathy Byrne** for the patient.

"We make sure the college clinic has a copy and that they have access to us," says Byrne. "We like to have these patients ready for all medical and social issues by the time they go to college."

Unfortunately, notes Crosson, too many patients are not ready for the adult transition, which explains the critical need for an adult clinic.

"The reality is that many patients go off to college and out of their parents' sphere feeling invincible, and they just don't get care," Crosson says. "That's a huge problem not just for cardiology patients but for patients with any chronic disease, from cystic fibrosis and liver disease to renal disease and sickle cell."

For more information or to schedule an appointment, call 410-955-5910.

With the help of Hopkins Children's clinic for adults with congenital heart disease, 18-year-old Tyler Littleton has learned how to independently manage his condition.

ECMO Goes on the Road

Hopkins Children's already had the heart-lung bypass system known as ECMO (extracorporeal membrane oxygenation) when it began to develop a ventricular assist device (VAD) program two years ago. That meant children awaiting heart transplant could be weaned off ECMO, which comes with bleeding and infection risks, to a VAD as a longer and more stable bridge to transplant. Great news for Hopkins patients, but what about those heart failure patients already on ECMO at other hospitals?

"It became apparent that we were the only pediatric heart transplant and VAD program in the area, which raised the question, 'How could we handle other patients already on ECMO?'" says ECMO coordinator **Gary Oldenburg**. "It was an aha moment."

The aha answer was ECMO transport, an option available at only a few other pediatric heart transplant centers nationwide. Oldenburg and critical care specialist **Jamie Schwartz** set the wheels in motion and with their multidisciplinary team began designing ways to convert an existing ground ambulance into a ground ambulance with ECMO—no easy chore when you consider an ECMO machine is about the size of an office desk. Also, the system would have to be extremely stable to reduce bleeding risks.

The solution was a portable ECMO pump about the size of a desktop computer connected to a customized shelf fastened overhead to an ambulance stretcher. The patient stretcher and portable ECMO pump became one unit that could smoothly slide in and out of the ambulance and pediatric intensive care units. Staff training and a simulated dry run with a mannequin led to some troubleshooting, which turned out to be serendipitous when the team got its first call two weeks later. The parents of an infant in heart failure at a Virginia hospital was about to call a mobile ECMO program in Pittsburgh when it learned about ECMO transport to Hopkins. The team successfully transported the patient to Hopkins, where two months later she received a donor heart.

"We gave the patient the opportunity to have a heart transplant, which was not offered at the other hospital, and to be close to home," says Schwartz. ■

For more information, call: 410-955-5260.

Do Your Patients **Have a Medical Home?**

"Employing a medical home model, we're seeing benefits like improved access to specialty care, better coordination of care with community-based organizations, and improved child-health outcomes."
—Barry Solomon, M.D.

Imagine you're a primary care pediatrician meeting a new family who recently moved to your city or town. The family has two children—Dion, a 6-month-old who was born prematurely at 28 weeks with feeding difficulties, chronic lung disease and developmental delays; and Shawna, a 12-year-old with moderate persistent asthma, anxiety and depression. The mother, a 32-year old with a history of depression, is unemployed and lives with friends. So, how do you manage these children and this family?

Such cases, pediatrician **Barry Solomon** explained at a recent Grand Rounds, illustrate the need for a medical home model in pediatric practice, especially for patients with special healthcare needs. Those children, who are at increased risk for chronic conditions—physical, emotional or developmental—require health and related services beyond those required by children in general.

"We can't just think about the patient in the context of the office or the clinic," Solomon said. "We have to think about what's going on in their homes, neighborhoods and schools."

Solomon noted that the medical home concept dates back to 1967 when the American Academy of Pediatrics (AAP) first established practice standards for children with special needs. A decade later the AAP cited the need for a "repository for medical records" for children with multiple-care needs to ensure continuity of care.

The model further evolved through physicians like Hawaiian pediatrician Calvin Sia, who incorporated concepts like family-centered and community-based care, and who developed training programs that tapped into neighborhood resources and focused on the emotional as well as medical aspects of care. Such contributions led to a 2002 AAP definition of medical home care as "accessible, continuous, comprehensive, family-centered, coordinated, compassionate, and culturally effective…. and delivered or directed by well-trained physicians who provide primary care and help to manage and facilitate essentially all aspects of pediatric care" (*Pediatrics* 2002;110:184-186).

How prevalent and effective are medical home models today? In a 2005-06 telephone survey, Solomon noted, 47 percent of families of such children said they had a medical home (National Survey of Children with Special Health Care Needs, *Pediatrics* 2009). In another survey of some 83,000 families of children with and without special healthcare needs, 57 percent of the families reported having a medical home (*Pediatrics* 2011;127:604-611). Of those families with a medical home, 1.6 percent said they had unmet medical needs, while 6.4 percent of those with no medical home reported unmet medical needs.

Also, in a retrospective review of 30 studies from 1986 to 2006 looking at medical home activities like care coordination, care planning and cultural competence, researchers found some positive short-term outcomes regarding effectiveness, efficiency, timeliness and family centeredness of care. Long-term positive outcomes included health and functional status and family function (*Pediatrics* 2008;122:3922-e937).

"So children in a medical home do have fewer unmet medical needs," Solomon said. "We think the medical home improves patient care, but we need more research to prove it."

To learn how to implement a medical home model in your practice, visit www.medicalhomeinfo.org.

Our Medical Home

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telephone—though the office encourages face-to-face visits whenever possible. Being a group practice open seven days a week with some 16 pediatricians and nurse practitioners at four locations, Annapolis Pediatrics makes itself as accessible as possible to patients, especially those with special healthcare needs.

"If I was in solo practice I wouldn't be here until 9 p.m. every night and working Saturdays and Sundays, so my patients would have to go somewhere else," Rice says. "This way they're still in their medical home."

In another step Rice reaches out to families and specialists in the community and places like Hopkins to collaboratively develop ways to manage complex patients. In spring 2011, for example, Rice participated in a panel of pediatric cardiologists and parents of children with congenital heart disease at Hopkins Children's annual Pediatric Trends conference.

"For us," Rice says, "it's mostly getting more comfortable with managing these complex patients, knowing what we can handle and when to defer to the specialist, and improving communication to deal with those areas in between."

Developing a medical home is a tedious, painstaking process, Rice concludes, with myriad medical issues, not to mention reimbursement codes for coordination of care. A good business office, he quips, helps. Perhaps new technologies and strategy meetings with easels lining the walls would help in developing a medical home model, he concedes, but at the end of the day best practices still come down to dedicated care.

"The idea of a commitment, that this is my patient and I'm going to see this through," Rice says, "is still the most important thing. Technology is never going to replace that." ■

Taking Specialty Care to the Patient

Pediatric surgeon **Jeffrey Lukish** recalls walking by the original Anne Arundel Medical Center in Annapolis as a young midshipman at the Naval Academy. Knowing he wanted to go to medical school after graduation, he said to himself, "I'm going to go in there and volunteer and see what it's all about."

So he did. Interestingly, two decades later he finds himself returning to that same hospital, albeit a newer one, as part of Johns Hopkins Pediatric Specialty Care at Anne Arundel Medical Center. With pediatric gastroenterologist **Christine Karwowski** and pediatric pulmonologist **Peter Mogayzel**, Lukish aims to bring Hopkins specialty care to the patient rather than the patient to specialty care in an urban hospital.

"Our goal is to provide outstanding expert care where children are sick, to take our expertise to the community," Lukish says. "It can be stressful for families to travel to cities like Baltimore or Washington, D.C."

Lukish will see patients on the first and third Wednesday of each month, and perform surgeries on the second and fourth Wednesday. Most of the procedures will be straightforward ones like appendix, gallbladder, hernia and soft-tissue operations, with a minimally invasive approach.

"All of these procedures can be done without making incisions or splitting muscles, which is safer for patients," Lukish says. "Also, they can go home the next day."

Jeffrey Lukish, M.D., one of three Johns Hopkins pediatric specialists now providing care at Anne Arundel Medical Center.

Mogayzel, who will see patients with general pulmonary problems like asthma the first and third Thursday of each month, also cites patient satisfaction goals in establishing the clinic.

"It's more convenient, especially for patients from the Eastern Shore, to be seen locally at Anne Arundel," Mogayzel says. "The bulk of our pulmonary care will be done there while more complex chronic conditions will be treated at Johns Hopkins Children's Center in Baltimore."

Gastroenterologist Karwowski takes care of children and teens with a wide variety of gastroenterology and nutrition-related issues and conditions, with a focus on inflammatory bowel disease. She will see patients at Anne Arundel the first and third Monday of every month, beginning Aug. 15.

"I am excited to see a new patient population at Anne Arundel," says Karwowski. "It is a terrific opportunity for me as a physician, as well as for area families."

For more information or to make an appointment at Johns Hopkins Pediatric Specialty Care at Anne Arundel Medical Center, call Gastroenterology at 410-955-8769; General Surgery at 410-502-6649; or Pulmonology at 410-955-2035.

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