

Perspectives

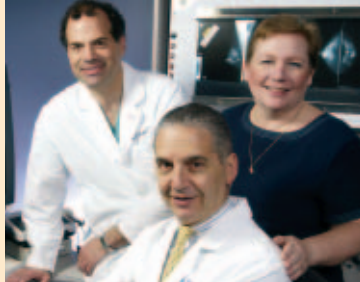
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THE JOHNS HOPKINS *Cutting* Edge



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An Operation No One Else Would Tackle

The tumor on 18-month-old Maison Jackson's face had ballooned to the size of a softball. Appearing around the little boy's first birthday, the mass had been steadily enlarging week by week, distorting his mouth, inflating his left cheek and closing his left nostril. By the time his parents were referred to Anthony Tufaro, a plastic surgeon specializing in head and neck surgery, the tumor had inched dangerously close to Maison's left eye.

At first, physicians had told the Jacksons that the growth was simply part of the port wine stain Maison had been born with. The reddish facial birthmark, characterized by an overabundance of capillaries just beneath the skin's surface,

worsening facial disfigurement that was also afflicting their son, the Jacksons heard only that there was nothing to be done for it.

“The moment I saw Maison,” says Tufaro, “I knew he needed an operation. He had what's called an ossifying fibroma—a rare tumor that's usually benign but very aggressive. It was so large that it had displaced his upper teeth and jaw. He couldn't eat, he couldn't breathe, he was snoring. He was so dysfunctional from the tumor, we had to do something.”

Tufaro knew immediately that the sheer size of Maison's fibroma was worrisome—“We weren't sure it wasn't cancer,” he says, “and we didn't know if we'd have to remove his eye and socket.” But what presented one of the toughest technical challenges was the fact that the tumor was sitting amid a bed of dilated blood vessels. Not only would Tufaro have to completely remove the boy's upper jaw in order to excise the massive growth, he would have to do so with the risk that the child could bleed to death during the operation.

To solve that problem,



“He plays, he's happy, he's ornery like any 2-year-old,” says Tammie Jackson of her son's recovery from his operation. “Dr. Tufaro was a godsend.”

Tufaro turned to interventional radiologist Sally Mitchell. Mitchell first used magnetic resonance imaging to make the tangle of blood vessels visible, then assisted Tufaro during the four-hour procedure by floating coils in the vessels to limit blood flow. Even so, says Tufaro, “I had to go slow. One blood vessel feeding that tumor

was the size of my finger.”

Tufaro was able to remove virtually all of the 5-centimeter mass, which to everyone's relief was noncancerous. “Maison will probably need further reconstructive surgery down the line,” he says, “but he bounced back one two three. He's growing. He's thriving.” ■

Mary Ann Ayl

The Surgeon Speaks

“There Weren't a Lot of Options”

Because of my training, I see a lot of unique tumors and a unique cadre of patients. I



was in private practice as an oral surgeon for seven years, but then I decided I wanted to do more. I went back to school for my M.D. and went on for training in general, plastic, and head and neck surgery. So when I approach any operation, I draw on all those specialties. For example, it's intuitive for me to have children fitted for braces before they have tumor surgery to minimize the effect on developing teeth. And as a plastic surgeon, I want to make sure the end result looks as good as possible.

I've seen—and removed—a lot of big tumors, but never before in a child the age of Maison Jackson. And I had never seen a tumor in the middle of so many blood vessels.

It wasn't easy to say, Sure, I'd like to operate. You don't want to have to totally take out half of a baby's upper jaw. But there weren't a lot of options. Maison was so distorted from the tumor that I actually felt good about undertaking his operation.

But I was glad when the operation was over. I am humbled every day in the OR. I treat every operation with respect, because anything can happen when you make an incision in someone's skin.

Anthony Tufaro

“He was so dysfunctional from the tumor, we had to do something.”

ANTHONY TUFARO

typically signals the presence of Sturge-Weber syndrome, a congenital neurological disorder that can include such symptoms as seizures, glaucoma and excessive blood vessel growth on the surface of the brain. When he was 8 months old, Maison did have two seizures but ever since then has responded well to medication. And by the time his tumor became visible, he was already being treated for glaucoma. Yet, despite the progressively

Perspectives



From Julie Freischlag
Director of Surgery

Vive la Difference!

In the small Midwestern towns where I grew up, most people looked and acted much like me. But when I moved to Chicago, Los Angeles and Milwaukee, I learned all about “diversity.” To me, the buzzword simply means people who are “different” types. It’s a natural occurrence. In fact, if you don’t have diversity in your surroundings, you have an artificial situation. Our world is diverse, and I think it’s a better place because of it.

Surprisingly, my introduction to diversity began with my role as a woman training in surgery in big cities in the 1980s. I was suddenly in the minority. But it didn’t take long to realize I loved stepping into a world of people from different backgrounds—all dedicated to patient care.

Surgery has been slow to accept women and minorities. Maybe that’s because once, only wealthy white males could afford medical school, or perhaps doctors felt more comfortable being around people like themselves and excluded everyone else. Thankfully that changed as more opportunities arose for educating women, minorities and foreign students.

Today, you can look around the Johns Hopkins Department of Surgery—one of the best in the world—and find men and women from the United States, India, Bermuda, Korea, the Philippines and many other countries. We’ve finally reached a point where it doesn’t matter where you’re from, as long as you’re good at what you do. Different isn’t wrong; different is just different.

And we can take better care of our foreign patients when someone from our team can speak their language. We’re nicer to patients and to each other. We learn different approaches to the same problem. We’re more creative and productive.

And when we’re open-minded, we can relax and appreciate our diversity.



Ted Tsangaris (left) and Lillie Shockney await Nagi Khouri’s analysis of a patient’s digital mammogram. The new technology allows earlier detection of tumors.



Surgeon Maurice Nahabedian says for patients like Felisa Maldonado, breast reconstruction means a better quality of life.

Teams at Work: Focus on Breast Cancer

A Place to Call Home

An unprecedented gift fixes the nagging problem of scattered services—and more.

The Project: Two years ago, when surgical oncologist Ted Tsangaris arrived at Hopkins to become director of the Johns Hopkins Breast Center, he found a superb group of services in place and a top group of specialists. But Tsangaris also saw room for change. In his view, the field of breast disease—the leading cause of cancer in women—set the gold standard for a team approach to care. Hopkins had mastered that concept, but, to Tsangaris’ dismay, patients—many weak from chemotherapy—were forced to shuttle between the Weinberg Center, Outpatient Center and two hospital floors to receive the full range of care. His goal: to find a way to bring all these services together.

The Players: About a dozen surgeons, radiologists, oncologists, nurses and geneticists make up the center’s diagnostic and treatment core group. The “breast surveillance team,” headed by nurse practitioners Sue Appling and Kyle Terrell, provides screening and education. Another key player, Nagi Khouri, director of breast imaging, uses high-resolution digital mammography to detect tumors, even in the densest breast tissue.

Once a woman has been diagnosed with breast cancer, Tsangaris, Julie Lange or Lisa Jacobs use minimally invasive techniques, whenever possible, to remove the malignancy. Chief of Clinical Oncology Nancy Davidson collaborates with Associate Director for Clinical Practice John Fetting on bringing the latest research advances into treatment with chemotherapy. When women opt for breast reconstruction,

Maurice Nahabedian uses a highly sophisticated technique called the DIEP flap, which only a handful of surgeons nationwide have mastered, to create a more realistic form.

But the hallmark of the Breast Center is its unflinching emotional support. Director of Education and Outreach Lillie Shockney, a breast cancer survivor herself, and a network of dedicated volunteers are always available to mentor patients facing surgery, chemotherapy and radiation.

The Results: Last fall, the Avon Foundation made a \$10 million gift to Hopkins. And with that donation, Tsangaris could plan the Center he envisioned. The new Johns Hopkins Avon

Foundation Breast Center—on the fourth floor of the Outpatient Center—will unite experts and technology to coordinate treatment from detection through diagnosis, surgery and postoperative care. By February 2005, those services will be housed in one beautifully decorated site. Meanwhile, a portion of the Avon gift is supporting research to detect and silence genes that contribute to breast cancer development.

Patients love the idea of getting all their care in one place, Tsangaris reports. Hundreds of them attest to the excellent care at the center. “We have some of the best people in the world,” Tsangaris says. “Now we’ll also have an environment we can call our own. I look at it as a reward for our patients.” ■

Let’s Meet: Endocrine Surgeon Alan Dackiw

A year ago, Alan Dackiw was at The University of Toronto Medical Center performing hundreds of thyroidectomies and adrenalectomies and seeing 30 patients a week. Today he’s forsaken his Canadian roots and accepted an offer from the Department of Surgery as an endocrine surgeon.

Dackiw (pronounced *DAK-cue*) is fast building a practice at Hopkins. He specializes in minimally invasive surgery to remove endocrine tumors of the adrenal, thyroid, and parathyroid glands as well as the pancreas. An M.D., Ph.D., Dackiw studies familial endocrine tumor syndromes, intraoperative nerve monitoring and endocrine organ development and its relationship to cancer. He’s fascinated by how the adrenal and thyroid glands develop and the mutations that cause endocrine syndromes, and he’s set his sights on finding a consistently effective antitumor drug.

Despite a rise in thyroid cancer (perhaps because of more radiation in the environment, Dackiw suggests) “we’re getting better at diagnosing it.” Clinicians are finding more nodules, and pathologists are looking harder at tissue, he explains. Hopkins embodies that vigilance, he observes. “I’m struck by the sense of unity here toward patient care.” ■



Epigenetics: The Latest Antidote to Cancer

Surgical oncologist Nita Ahuja once believed that excising tumors was the most satisfying part of her job. But increasingly she's drawn to her lab, where she explores new frontiers in molecular biology—methylation and its reverse—to silence or turn on gene function.

For more than a decade, Ahuja has studied ways to thwart colorectal and breast cancer growth.

But she's never been as excited about her work as she is right now.

Removing tumors isn't enough any more, she says.

Instead, Ahuja wants to harness epigenetics, changing the information carried by genes

without altering their sequence, to improve treatment. "The future," she says, "is all about looking at genes and molecular biology—and then finding out how they

affect the patients in the clinic." That idea has a name too: translational research.

Hopkins' Department of Surgery has one of the largest dedicated translational research teams in the country, according to Chief of Surgical Oncology Steve Leach. The new model surgical clinician/scientist combines knowledge of a patient's cancer history with genetic research. "Nita is the perfect

example of the kind of superbly trained surgeon-scientist who is contributing to the expanded research effort within our department," he says.

Not long ago, explains Ahuja, scientists thought mutations—changes in genes' building blocks—were the only cause of cancer.

Now they know that sometimes people can acquire changes in gene expression that don't affect DNA sequence. But these epigenetic changes can turn off tumor suppressor

Picking up these altered epigenetic patterns early in sputum, stool or blood samples from cancer patients can tell researchers how aggressive the cancer is.



Nita Ahuja uses DNA methyltransferase enzyme (backdrop) to manipulate gene function.

genes just as mutations can, encouraging tumor growth.

Picking up these altered epigenetic patterns early in sputum, stool or blood samples from cancer patients can tell researchers how aggressive the cancer is. Ahuja is quick to say that detecting cancer before it happens is a long way off. But assessing a person's potential for chemotherapeutic response is a more attainable goal—maybe within the next five to 10 years.

Using epigenetics, especially methylation, already has worked for curing certain leukemias. In some clinical

trials, doctors were able to reactivate or turn on "good" genes so that aggressive chemotherapy could stop the blood disorder in its tracks. In other studies, chemotherapy wasn't even necessary. But finding prognostic epigenetic markers or therapeutic targets for solid organ tumors has proven much more difficult. Still, Ahuja and her team are collecting data to see whether tissue sample research helps direct treatment options for colon and breast cancer patients.

Most troubling to Ahuja are the questions she and her

team have to ask themselves: "Could we have foreseen that a patient would have developed a tumor? Should that person have gotten chemo earlier?" Clinical trials are answering those questions by measuring outcomes.

As for the future, Ahuja says the "holy grail" would be having the ability to use drugs that demethylate to turn genes back on. "We've learned so much about genes over the last 30 years. I have no doubt we'll continue to find innovative ways to control the spread of disease. That's why we go into medical research." ■

On the Job

Deb Baker: Manager, Surgical Advanced Practice

"Good NPs haven't forgotten they're nurses first."

Nurse practitioners emerged in the 1960s—from a need for experienced RNs to assist overextended physicians in rural communities. Since then, NPs have transformed nearly every realm of private and academic medicine throughout the United States. Deb Baker, recently hired to supervise 25 NPs and seven physician's assistants, talks about the role these professionals play in the Department of Surgery.

We all know that NPs require superb training, but what do they do that's so valuable?

Our NPs are an integral part of the team and have many roles: writing orders, educating patients, prescribing, discharge planning. We spend the most time with patients—we've earned their trust. Surgeons and nurses trust us too.

Have they had trouble being accepted within the team?

No, not in surgery. On the contrary: It used to be so frustrating trying to find a surgeon

available to sign orders. Now an NP or PA is always on the floor to do that job. Occasionally I'll see some tension when a new NP joins the team and works with a seasoned R.N. In such cases, new NPs have to prove themselves.

What makes a good NP?

Good NPs haven't forgotten they're nurses first. The biggest asset is nursing experience stressing a holistic approach. These nurses have an intuitive sense that something might be going wrong for the patient,



like a pending heart attack.

But don't surgical NPs require additional skills?

Yes, most need acute care certification. They should excel at working with acutely ill patients in their specialty (e.g., colorec-

tal). That also means interacting well with families. And many of our NPs do research.

What are your biggest challenges?

Teaching people how to parcel the work. NPs and PAs work beside residents, who are rotating for just a month. Interns want practice doing the urgent things that NPs and PAs often do. It requires flexibility and communication. But the bigger challenge is recruiting more inpatient NPs and PAs, which we need badly. ■

Of Kilns and Kidneys

To Margery Pozefsky, shaping a piece of pottery is not unlike starting a new program. In both cases, the inspiration starts with a need. In pottery, it may be for a decorative vase. In the case of the Johns Hopkins Incompatible Kidney Transplant Program (InKTP), which she founded, her inspiration was a desire to help.

In 2000, doctors discovered Pozefsky needed a kidney. Her husband, Thomas, wanted to donate one to her, but their blood types didn't match. Transplant surgeons James Burdick and Lloyd Ratner (who has since left Hopkins) encouraged her to ask other family and friends to step forward. But Thomas, a Hopkins internist, was willing to donate a kidney to anyone who matched his blood type. What if, Mrs. Pozefsky wondered aloud, we could be paired with another couple in the same situation and swap kidneys?

Ratner told Pozefsky it was a great idea, but Hopkins lacked



Margery Pozefsky at home with her functional creations.

the resources to build a database and the nursing support to screen potential altruistic donors and recipients. "Maybe one day," he said, "but you need a kidney now." And so,

Pozefsky's son Kenneth Peyton—a perfect match—insisted on donating a kidney to his mother. Pozefsky feared for her son's health, but the two of them did well.

Meanwhile, Pozefsky couldn't stop thinking about all the other people awaiting kidneys. Born into a philanthropic family, she understood that good ideas require financial support. (She's vice chair of the Baltimore Symphony Orchestra, head of the capital campaign for Baltimore Clayworks and

past president of the League for People with Disabilities.) And today, thanks to her donations, InKTP has helped hundreds of people receive kidneys.

Pozefsky recalls the morning she watched three donor/recipient pairs meet for the first time on national TV as Transplant Center Director Bob Montgomery explained InKTP's mission. "It was a fabulous moment," she says. Afterwards, she went to work in her studio and inched closer to filling her kiln. ■

If you'd like to make a gift to the Department of Surgery, contact Boi Carpenter-Mellady, director of development, at 410-516-5483 or bmellady@jhmi.edu. To no longer receive information about supporting Hopkins' Department of Surgery, write to her at One Charles Center, Suite 421, 100 N. Charles St., Baltimore, MD 21201.

HOOP SCOOP

In front of a crowd of more than 400, a group from the Department of Surgery, organized by Charlie Yeo, hit the hardwood at Villa Julie College on March 26 to benefit the children of the Police Athletic League. They took on the Harlem Ambassadors, a spin-off of the Harlem Globetrotters, in a game that was three parts b-ball, one part comic routine. "We lost, but we didn't embarrass ourselves too much," says co-captain Eddie Cornwell, who broke his pinky in the fray but claims it hasn't affected his OR performance. "You learn to do it with four fingers," Cornwell says. "I broke the other one last year playing basketball. It's an addiction—what can I tell you?"

FOND FAREWELL

Jean Queen, one the last members of the technical staff who worked with cardiac surgery pioneer Vivien Thomas, retired after 45 years at Hopkins Hospital. Thirty-five of those years she spent in the Surgery Research Laboratory. At a recent farewell party, friends shared reminiscences about Queen's pivotal role.

AWARDS AND HONORS DEPARTMENT

Chief of Thoracic Surgery Steve Yang and his division presented three papers at the recent American Association for Thoracic Surgeons meeting. AATS—which has an international membership of 1,143 premier cardiothoracic surgeons—selects only 16 abstracts from more than 3,000 entries. That means almost a quarter of this year's selections originated from Hopkins' Thoracic Surgery. Yang was also recently voted Medical College of Virginia's most outstanding alumnus. And, this year's American Orthopaedic Association's volunteer clinical faculty award will go to Gershon Efron. Finally, Lillie Shockney, director of breast cancer outreach and education, was chosen as one of Maryland's Top 100 Women of 2004. The award is given annually by *The Daily Record* in recognition of women who make a significant impact on the state.

ONE MORE FOR THE PORTRAIT GALLERY

As it does for all retiring chairmen, Hopkins commissioned an official portrait of former Chief of Surgery John Cameron. It was dedicated in January. Cameron (JHM '62), who was chair for 18 years, is most famous for performing the complex Whipple procedure to stave off pancreatic cancer. More pancreatic surgeries were done here during his reign than anywhere else in history. Cameron just completed his 1,000th successful Whipple.

JOHNS HOPKINS MEDICINE

THE **CuttingEdge**

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