Zeroing in on Sickle Cell Retinopathy

There’s no time to waste
Music’s Soothing Powers

Though cataract surgery is commonly performed and highly successful, the idea of eye surgery still provokes anxiety in many patients. Wilmer cataract surgeon Christina Prescott, M.D., Ph.D., is researching alternative therapies to improve the patient surgical experience. “Patients are not under general anesthesia,” she explains, “so the more relaxed and comfortable they are, the smoother the surgery will go and the better their experience will be.”

In a current randomized study, she is investigating whether listening to music during surgery reduces anxiety—and hence the level of anesthesia needed. Because most cataract patients have separate surgeries, one for each eye, Prescott has a natural control group. Patients are given an iPod for both surgeries but only choose music for one. Once both surgeries are complete, she compares the patient’s heart rate, blood pressure, amount of anesthesia used and the results of a questionnaire (aimed at measuring anxiety) that is filled out before and after each procedure. The study is ongoing, so results aren’t yet final, but Prescott says that patient feedback to date has been promising.
Teach Your Children Well
Gene de Juan Jr. has honored his father’s commitment to teaching with an endowed professorship that supports the important work of running Wilmer’s residency program.

A Bigger Cause
Newly equipped with a dual infusion of funding, Akrit Sodhi and colleagues are focusing on oxygen deprivation at the cellular level in their quest to find treatments for devastating eye diseases.

Zeroing in on Sickle Cell Retinopathy
Adrienne Scott is building on research that revolutionized the field half a century ago to help patients whose condition has gone underfunded for too long.
Dear Friends,

The Wilmer Eye Institute knows a thing or two about partnerships! In this issue of Wilmer, you will read about remarkable lasting partnerships, including some spanning half a century or more.

Possibly the longest ongoing relationship revolves around Mr. Albert Turner. Now in his second century of life, this Wilmer Board of Governors member was recently recognized by Johns Hopkins University President Ronald J. Daniels. At a celebratory event, President Daniels acknowledged Mr. Turner’s partnership with Wilmer faculty members Walter Stark and now Yassine Daoud, as well as faculty members in other departments whose work has been accelerated by Mr. Turner’s generous support.

We also celebrate the four-decadelong alliance that began when Howard and Elaine Brownstein made a gift to launch the research career of a young assistant professor, Gerard Lutty, who was then facing the daunting challenge of convincing the National Institutes of Health that his new ideas were worth exploring. Today, Dr. Lutty is one of Wilmer’s most internationally recognized scientists, and the Brownsteins’ gift has been subsequently matched by many millions of funding dollars from NIH.

Wilmer’s two newest endowed chairs—the Branna and Irving Sisenwein Professorship and the Helen Larson and Charles Glenn Grover Professorship—reflect collaborations unique in Wilmer’s history. They resulted from the extraordinary generosity of private citizens who made Wilmer part of their estate plans and matching awards from the taxpayers of the state of Maryland via the E-Nnovation Initiative.

Research to Prevent Blindness (RPB), the largest private supporter of vision research, continues after five decades to be a generous catalyst on important research projects here at Wilmer and Johns Hopkins. Wilmer is thrilled to announce three new RPB awards to propel forward some of our most exciting projects.

You’ll also read about partnerships among faculty members that span generations, including at least one case in which the career of the senior scientific partner was well-established prior to the birth of the junior partner! The availability of “seasoned” professors to serve as collaborators and mentors to assistant professors who are decades younger is a particularly valued advantage enjoyed by those who come to Wilmer to launch their academic careers. These coalitions among faculty members, generous donors, and private and public agencies will bear fruit in improving how we care for and prevent vision loss in the future in patients with sickle cell disease and macular degeneration.

To all who serve as partners in furthering the work of the Wilmer Eye Institute to preserve, protect and restore vision around the world, I offer my sincere appreciation.

Peter J. McDonnell, M.D.
William Holland Wilmer Professor and Director
AFTER A LIFETIME of building things—houses, shopping malls, office high-rises and whole communities almost entirely from scratch—Albert Turner knows a thing or two about strong foundations. He is, after all, 101 years old. He began building family homes at age 21, before heading off to World War II, and he is now recognized as one of the preeminent builders in the national capital area.

Turner long ago ceded day-to-day management of his company, Carrollton Enterprises, to his son, Glenn Turner, and to his grandson, Ian Kelly. Nonetheless, Turner endeavors to make it to the office at least a few times a week.

Among Albert Turner’s proudest recognitions is as a philanthropist, and one of his favorite causes is the Wilmer Eye Institute at Johns Hopkins. This year, Turner will be enshrined on Johns Hopkins’ Founders’ Wall, which honors an esteemed group of benefactors who have surpassed the remarkable threshold of $7 million in gifts to Johns Hopkins.

Turner first turned to Wilmer when he began to develop cataracts many years ago. He was a patient of Walter Stark, M.D., for years and is now in the care of Yassine Daoud, M.D.

“My grandfather has been coming to Wilmer for decades, and they’ve helped him retain his vision past the century mark. He is very grateful,” says grandson Kelly.

“Turner gives so generously to Wilmer, Kelly says, because he knows the potential of the research that goes on there and because he believes everyone, regardless of means, should be able to enjoy the level of care for which Wilmer is known.

“Like one of his buildings, he wants to ensure Wilmer is standing on a strong foundation,” Glenn Turner adds.

“My grandfather has been coming to Wilmer for decades, and they’ve helped him retain his vision past the century mark. He is very grateful.”

—Ian Kelly
WHEN RANDY CRAVEN, M.D., became chief of Wilmer’s Bethesda clinic less than one year ago, the job was both exciting and daunting. There was, of course, the opportunity to lead and grow Wilmer’s largest market outside of Baltimore, one that happens to serve the considerable patient population in the national capital area. But the job was not without its challenges. Space was limited, and the technical infrastructure had pressing needs as well.

Fortunately, Craven, who brought to the job a background in clinic management, is more than up to the challenge. His plans include growing into a new office in order to triple the office’s space (later in 2018). There will be a broadening of surgical capabilities, and he plans an expansion of primary eye care services to help more patients.

“Having a full-service clinic just outside D.C. really helps our patients, for whom a single trip to Baltimore is a half-day affair at the very best,” he says.

Craven is not alone in his work to build the very best clinic he can. He is supported by many others, including clinicians and staff members, and by Wilmer board member and Bethesda resident Liz Dubin, whose late husband, Dick, had been treated for macular degeneration at Wilmer.

“They really saved his sight, and, after that, I joined the board,” says Dubin.

Dubin has been a generous supporter of Wilmer over the years and, in particular, the Bethesda office. Recently, she purchased a new Optos retinal camera for the Bethesda clinic. To Craven’s knowledge, Wilmer’s Bethesda clinic is now the only ophthalmology facility in the Washington, D.C., area with this camera, which offers doctors a much wider view of the retina and thus aids in better diagnosis.

“Liz was very straightforward and asked me how she could help,” Craven says. “I said we needed this camera. She has been tremendously helpful in our ability to bring cutting-edge services to Bethesda.”

Many doctors are not aware that many people, like Dubin, are eager to help, he says. “Sometimes, all you have to do is ask.”
That’s Classified

Rob Chun lends his visual assessment skills to athletes competing in the International Paralympics.

AS AN OPTOMETRIST working at one of the foremost eye centers in the world, Wilmer’s Rob Chun, O.D., has plenty of challenging assignments to tackle. A few years back, he was given a big one. That’s when a mentor in Chicago nominated Chun to become one of a handful of certified international vision impairment classifiers for the International Paralympic Committee.

The Paralympics organizes global events for all sorts of sports for people with a range of physical impairments, from amputees to the blind. Chun travels the world three or four times a year to attend various Paralympics events, where he evaluates and classifies the competitors according to their visual abilities.

“I have a background in visual function and electrophysiological testing, which helps properly assess the athletes’ visual capabilities. That particular combination is important to the International Paralympic Committee. It was a natural fit,” Chun says.

Chun is now one of just four such trained and certified classifiers in the United States and one of just 60 or so in the world. There is a great need for such services, he says. “The Paralympics has a lot of events at different levels of talent and age. We evaluate and classify athletes so that people with similar visual abilities are competing against one another,” he says.

In March, Chun made his way to Rio de Janeiro, Brazil, for the Para-cycling Track World Championships. He has worked events ranging from track and field and skiing to a soccerlike sport called goalball, created specifically for blind participants. In goalball, the para-athletes are actually blindfolded to ensure a more equitable level of impairment to all.

As for the personal rewards of this work, Chun enjoys traveling and meeting doctors and athletes from around the world, but he also sees the impact on his youngest patients back home. “I talk about it with them. I use it as motivation. Learning that others with vision impairment can compete in sports really inspires them to live a full life,” Chun says.
Making a Difference

Founded in 1960 by ophthalmologist Jules Stein, Research to Prevent Blindness (RPB) funds, coordinates and promotes vision research in the United States. In the course of pursuing its mission of “preserving vision, restoring sight,” RPB provides funding to 37 scientific institutions, including the Wilmer Eye Institute.

Wilmer Director Peter J. McDonnell, M.D., is delighted to see these grants awarded to Wilmer researchers and their colleagues who are working on projects poised to make a radical difference in the lives of patients. The 2018 awardees and their projects are:

**Jordan Green, Ph.D.**
RPB Dr. H. James and Carole Free Catalyst Award for Innovative Research Approaches for Age-Related Macular Degeneration

The abnormal growth of blood vessels wreaks havoc on the retina and is often the root of a range of eye diseases—from wet age-related macular degeneration (AMD) to diabetic retinopathy. One current treatment inhibits the protein called vascular endothelial growth factor (VEGF), but it has drawbacks. Green hopes the new treatment he is working on can do more for patients—perhaps even restore vision they have lost. “It’s a drug that works through a different mechanism, so it’s not directly blocking VEGF. Instead, it’s disrupting a different target that then leads to blocking the activity of VEGF and non-VEGF growth factors, and activates an additional factor to stop vessel leakage as well,” Green says.

The drug is a peptide, which is a piece of a protein. “We’re developing a controlled release system so we can have one injection deliver this peptide drug in a way that it’s active for six months or longer,” says Green.

“This award means a great deal to me and my lab for our project. It enables us to do cutting-edge research at the interface between ophthalmology and a field outside of ophthalmology—biomedical engineering—and to conduct interdisciplinary research with collaborators to create new treatments that can make a difference,” says Green.

**Ian Pitha, M.D., Ph.D.**
RPB Career Development Award

Pitha focuses his research on a part of the eye that takes up a lot of surface area but hasn’t taken up a lot of space in the minds of researchers until the past couple decades. “The sclera is the white part of the eye—the wall. It’s what gives your eye structure,” says the ophthalmologist. Led by Wilmer’s Harry Quigley, M.D., researchers have found that the sclera, which plays a central role in controlling the effect of eye pressure on the optic nerve, is implicated in glaucoma. Pitha’s work builds on this by seeking to discover how the cells of the sclera—particularly fibroblasts—are organized and how they behave when subjected to different stressors. The hope is eventually to create a drug that can manipulate the behavior of the sclera to respond to eye pressure more effectively and protect people from glaucoma.

“The funding from RPB is great,” says Pitha. “It’s really good to get this funding earlier on because you just need a little time to figure things out.”
Gregg Semenza, M.D., Ph.D.
RPB Sybil B. Harrington Innovation Award for Macular Degeneration

Injections of anti-VEGF medications have revolutionized treatments for such eye diseases as AMD and diabetic macular edema because they inhibit new blood vessel growth. However, anti-VEGF medications do not help all patients. Enter: HIF-1 (hypoxia-inducible factor 1) protein.

“We’ve shown in mouse models that when we treat the mice with a drug that blocks HIF, we block all of the increases—not just in VEGF but other angiogenic factors also,” says Semenza, who discovered HIF-1, for which he won the Albert Lasker Award for Basic Medical Research in 2016.

The C. Michael Armstrong Professor of Medicine and director of the vascular program at the Johns Hopkins Institute for Cell Engineering, Semenza is partnering with several doctors at Wilmer to translate HIF-1 inhibitors into safe, effective medications for patients with the help of funding from RPB.

“When you get your discovery to the point of trying to translate it to the clinic, this is when it’s most difficult to get funding from NIH,” says Semenza. “To be able to have funds that we can specifically use for that purpose is really important.”

Quigley Honored by ARVO

Harry Quigley, M.D., the A. Edward Maumenee Professor of Ophthalmology at the Wilmer Eye Institute, has received the Joanne G. Angle Award from the Association for Research in Vision and Ophthalmology (ARVO), the preeminent eye and vision research organization in the world. “This award recognizes outstanding leaders who have made significant, continuous contributions to ARVO in support of its mission to advance research worldwide into understanding the visual system and preventing, treating and curing its disorders,” notes the organization.

Quigley served as executive vice president and editor-in-chief for ARVO. During his tenure, he extended ARVO’s annual program of oral and poster presentations by one day in order to accommodate cross-disciplinary symposia—a design change still in effect 30 years later.
IN 1979, WHEN Gerard Lutty, Ph.D., was a young scientist, fresh from his master’s thesis and eager to make his mark on the world of ophthalmic research, he learned quickly that funding his research would be tough going. It is not an uncommon situation for young researchers, regardless of the relative promise of their research ideas. Donors often want to fund well-developed ideas advanced by scientists who have earned their stripes. At that stage of his career, Lutty had nothing but promise.

It was at this point that the renowned Arnall Patz, M.D., then director of the Wilmer Eye Institute, took the young Lutty under his wing. Patz would escort Lutty around, introducing him to his patients and letting Lutty look into their eyes and learn about their eye disease. Unbeknownst to Lutty, many of those patients were also donors.

“This young man is going to cure your disease,” Patz would tell the patients without hesitation.

The gesture was more than a touching compliment to Lutty. It was just the infusion of faith he needed to spark his career. If confidence has a transitive property, Patz and Lutty were living proof. Soon, several of those patients became benefactors to the as-yet-unproven researcher they had just met, launching him on his way.

“Arnall was a lovely man who, for a basic scientist like me, was nothing less than a hero,” Lutty, now Wilmer’s G. Edward and G. Britton Durell Professor of Ophthalmology, says today, decades into a remarkable research career. “His belief in me made me feel great.”

One couple Lutty met back in those early days was Howard and Elaine Brownstein. Howard Brownstein was a patient of Patz and a member of Wilmer’s Board of Directors. He suffers from diabetic retinopathy and was immediately taken with Lutty’s description of the young scientist’s research approach. Soon, he was funding Lutty’s new approach to diabetic retinopathy, which is caused when blood vessels in the retina begin to die.

“Many people thought it was just the high glucose or nasty things in the blood,” Lutty says of the state of the science at the time. But he had a different notion.

His hypothesis was that cells in the lining of the blood vessels were somehow making chemicals that attracted white blood cells: Thus, the body’s own immune cells were sticking to the blood vessels and blocking blood flow through the blood vessels, which eventually killed the blood vessels and caused blindness.

Lutty’s theory proved true when his team discovered a particular type of white blood cell specifically associated with the decline of blood vessels in the retina. It was an important advance that may never have happened without support from the Brownsteins.

Rachel Brownstein Koch, Arnall Patz, Howard Brownstein and Elaine Brownstein in Wilmer’s Portrait Room several decades ago

“Dr. Patz told me he needed funding to keep Dr. Lutty at Wilmer. Dr. Patz had saved my vision, so, of course, I listened. It was a
no-brainer to provide the financing,” Howard Brownstein says.

Over the years, the relationship between the Brownsteins and Lutty has blossomed in new and surprising directions: Their daughter, Rachel Brownstein Koch, would work with and co-author a peer-reviewed paper with Lutty. She earned her master’s degree in health policy and management at Johns Hopkins.

In the three-plus decades since he first met the Brownsteins, Lutty has become one of the world’s foremost experts on the molecules that stimulate new blood vessel growth in the eye—as well as those that stop that growth, a promising strategy for halting the growth of tumors.

“I think ophthalmology is so important because the work that’s being done these days doesn’t only affect the eyes, it affects other areas, including oncology and even brain cancer,” Howard Brownstein says when asked why he funds research like Lutty’s.

It all goes back to that day Patz made a simple introduction, says Howard Brownstein, who would also formalize his respect for Patz when he made the lead gift to establish the Arnall Patz Distinguished Professorship. It is a permanent chair currently held by Lutty’s colleague Kannan Rangarajan, Ph.D., who works with cutting-edge nanoscale drug delivery mechanisms.

“Howard has always been so generous to me and to Wilmer,” Lutty says, “It’s really a model patient/researcher relationship.”

“Arnall was a lovely man who, for a basic scientist like me, was nothing less than a hero.”

—Gerard Lutty
Teach Your Children Well

Gene de Juan Jr. has honored his father’s commitment to teaching with an endowed professorship that supports the important work of running Wilmer’s residency program.

GROWING UP IN Mobile, Alabama, Gene de Juan Jr., M.D., learned a lot from his ophthalmologist father, Eugene Sr.—perhaps more than his father ever realized. The elder de Juan was a respected physician in and around Mobile, where his work brought him into the sphere of William Holland Wilmer (namesake of the Wilmer Eye Institute) and Edward Maumenee, both of whom made names for themselves much farther north.

While Eugene de Juan Sr. was a noted doctor, he was much more than that. He was founder of the first eye bank in Alabama and a committed volunteer in the community. He was also a sought-after teacher who welcomed medical residents from as far away as New Orleans, 150 miles to the west, according to his son.

“My father was an active and generous man who took the time to teach not just me, but many others. It was a different time, when relationships were more important than they are now,” says the younger de Juan, a noted ophthalmologist in his own right. After completing his residency at Wilmer in the early 1980s, he served nine years as co-director of the Wilmer Eye Institute’s Vitreoretinal Service in the 1990s, before moving on to the University of California, San Francisco, where he is now the Jean Kelly Stock Distinguished Professor of Ophthalmology.

He was so moved and grateful for his father’s legacy that he endowed a professorship at Wilmer in his father’s name, specifically to support education: the Eugene de Juan, M.D., Professorship in Ophthalmic Education.

“I grew up in that environment and saw how important teaching was to my father and to the profession,” he says. “Though my father never wanted or sought attention, I wanted to honor that tradition.”

Two Wilmer physicians who have benefited from the professorship are Michael Boland, M.D., Ph.D., the current director of Wilmer’s residency program, and Divya Srikumaran, M.D., the former director. Both know well the advantages a funding stream like the de Juan Professorship affords and see the results of that philanthropy every day when working with residents.

“Training the next generation is perhaps the most important way to im-
pact the future of ophthalmology, but the role of residency program director has become more time-consuming and rigorous over time,” says Boland.

There are more frequent and demanding reviews of how the residency program is meeting its goals, for instance, and the ever-present responsibility of maintaining the program’s currency with the latest medical advances. On top of it all, there are the pressing recruitment challenges of finding and attracting the very best trainees to Wilmer year after year. It can be overwhelming to handle all of these duties on top of a heavy research and patient care load.

“The de Juan professorship allows me to commit fully to leading the residency program and maintaining the best training program possible for the next generation of ophthalmologists,” Boland says.

Srikumaran concurs in that assessment and says an endowment is also important for its symbolism. “Gene de Juan was the beneficiary of the Institute’s commitment to teaching excellence. This professorship is a perfect embodiment of the two great influences in his life—his father and his Wilmer education.”

For his part, Gene de Juan Jr. has been deeply gratified to see the endowed professorship bear fruit. “There’s a wonderful tradition of education at Wilmer, and I was happy and proud to make that contribution in my father’s memory,” de Juan says. “It’s so important to encourage teaching.”

Training the next generation is perhaps the most important way to impact the future of ophthalmology.

—Michael Boland

Nancy de Juan, Gene de Juan Jr., Eugene de Juan Sr., Wilmer Director Emeritus Edward Maumenee and Sue Maumenee

In Wilmer’s surgical wet lab, from back to front: Divya Srikumaran, resident Angeline Nguyen, Michael Boland and resident Ravi Pandit
A Bigger Cause

Newly equipped with a dual infusion of funding, Akrit Sodhi and colleagues are focusing on oxygen deprivation at the cellular level in their quest to find treatments for devastating eye diseases.

Photo by Chris Myers

THOSE WHO KNOW mountain climbing are familiar with the condition hypoxia. At high altitudes, the air is thin. It contains less oxygen, making it hard to breathe. Side effects include headaches, dizziness, confusion and nausea. In the worst instances, it can kill. The body’s response to hypoxia is to emit a cry for help in the form of a protein known as hypoxia-inducible factor, or HIF, for short. HIF implores the body to increase production of the protein erythropoietin, which is tasked with the job of increasing the delivery of oxygen to the body part that is hypoxic.

At lower altitudes and on a smaller scale, hypoxia can have a similar effect in the eye. Hypoxia plays a central role in most forms of blindness. When cells in the eye sense a decrease in oxygen, they do not give up easily, but rather again summon HIF for help. HIF directs the eye to produce more blood vessels to increase the supply of oxygen by increasing expression of another protein, vascular endothelial growth factor. However, the vessels created in this process are often leaky. The oxygen-starved retina quite literally drowns as a result of these incompetent blood vessels, and it fails.
“In the retina, the big culprits from this kind of effect include wet age-related macular degeneration—AMD—diabetic eye disease, retinal vein occlusions, sickle cell disease and retinopathy of prematurity,” says retina specialist Akrit Sodhi, M.D., Ph.D.

Sodhi is co-founder of a new company that will exploit a novel understanding of the root causes of such diseases to develop drugs that might one day slow or even cure them. HIF has proven so promising that Sodhi and two Johns Hopkins colleagues, renowned researcher Gregg Semenza, M.D., Ph.D., and biomedical engineer Jordan Green, Ph.D., formed a company, HIF Therapeutics, which is garnering considerable interest in medical and technology circles, even prior to the release of a marketable product.

Sodhi is a noted authority on retinal and choroidal neovascular diseases. Semenza discovered HIF and was the first to describe its regulation of erythropoietin and vascular endothelial growth factor. Green is an expert in cutting-edge drug delivery, a key to getting pharmaceuticals into difficult-to-reach areas of the eye. Together, their goals are to learn more about how HIF works and to develop HIF inhibitors that might help control the growth of new blood vessels and aid in the creation of new drugs.

The promise of HIF-related research, however, stretches beyond ophthalmology. Sodhi says. The knowledge flowing from HIF Therapeutics will join the growing body of understanding of angiogenesis, the process by which the body generates new blood vessels. HIF has become a promising research target for other fields, including oncology, dermatology, rheumatology and cardiology. HIF could prove effective at controlling or stopping the growth of new cancer tumors.

Inspired by HIF Therapeutics’ potential and the stellar reputations of the lead investigators, the state of Maryland recently awarded Sodhi one of its highly prized E-Nnovation grants. E-Nnovation is an initiative meant to fuel scientific and technical research and jobs within Maryland.

The E-Nnovation grant matched a private donation from the Branna and Irv Sisenwein Charitable Foundation. The combined funding of $2.8 million will create an endowed professorship for Sodhi that will allow him to focus much of his attention on HIF and its role in eye disease.

The Sisenweins had been longtime friends of Wilmer Director Peter J. McDonnell, M.D., who met them 30 years ago when he was starting his academic career in Los Angeles. The Sisenweins suffered from severe retinal diseases that ultimately robbed them both of useful vision. McDonnell, a cornea specialist, met them by chance when they came to an appointment with another faculty member, and that lucky encounter in a hospital elevator blossomed into a long and fond friendship between Branna and Irving.
Sisenwein and Peter McDonnell and his wife, Jan.

Through the years, the Sisenweins would often turn to McDonnell for advice and for help getting their friends and acquaintances to the right doctors. “If they learned that someone in their town of Palm Springs, California, had an eye problem that was not responding to treatment or if the person did not have insurance to pay for care, Branna would be on the phone to me immediately. ‘Now, Peter, darling,’ she would say to me, ‘how are we going to help her?’” says McDonnell. Eventually, they established their memorial fund to support numerous charitable causes, including ophthalmic research.

“My aunt and uncle were passionate about ophthalmology and the retina specifically,” says Elin Gursky, who oversees the Sisenwein Charitable Foundation. “They were quite impressed over many years’ association with Dr. McDonnell, and his faith in Dr. Sodhi’s work was quite persuasive.”

“If successful,” McDonnell says, “Dr. Sodhi’s work would be a substantial leap forward in terms of managing people with these very severe and common conditions.”

The form of macular degeneration that Branna Sisenwein suffered from is precisely the type HIF Therapeutics will target. Though the Sisenweins did not live to see a cure for their diseases, that did not lessen their resolve.

“They remained two of the happiest, most optimistic people that I’ve ever known,” McDonnell says. “Faced with severe vision loss, they didn’t spend one minute feeling sorry for themselves. Instead of feeling defeated, Branna and Irv committed themselves to helping others and doing all they could to prevent other people from experiencing the same loss of vision that they shared.”

The professorship the Sisenweins endowed is highly valued by a young scientist like Sodhi, McDonnell says. Grants from the National Institutes of Health and other organizations often include clearly defined uses for the money; researchers are not allowed to veer from those stated purposes to pursue their latest, potentially game-changing ideas. In contrast, the Sisenweins’ funding comes without strings attached and allows a promising researcher the freedom to follow hunches wherever they might lead.

“That sort of intellectual freedom was exactly what the Sisenweins had in mind for their foundation. In the past, they had given brick-and-mortar type gifts to Wilmer, but with their foundation, they wanted more,” Gursky says.

“They wanted to support high-risk, high-reward bench research, like Dr. Sodhi’s,” she says. “This wasn’t about them; it was about a bigger cause.”
Zeroing in on Sickle Cell Retinopathy

There’s no time to waste. That’s the mantra of Wilmer donors and researcher Adrienne Scott, who is building on research that revolutionized the field half a century ago to help patients whose condition has gone underfunded for too long.

By Jessica Wilson | Photo by Chris Myers

Howard Woolley laughs a lot when describing his late wife, Gail Campbell Woolley, a longtime journalist who began her career with the Washington Star and the Baltimore Sun, and later joined the public relations team for Mobil Oil Corporation. “Gail was a lot of fun,” he says. “Also, very honest. It was sort of like the Jack Nicholson line, ‘You can’t handle the truth.’ You didn’t want to be Gail’s friend if you couldn’t handle the truth.”

In her memoir, Soar, published posthumously, Gail explained the reason for this. “I can be pretty blunt. I am direct because I cannot stand to waste time I do not have.”

When she was 7 years old, she was diagnosed with sickle cell disease and told she would live to only 35. She proved that doctor wrong and made it to 58, packing more into her life than most of us could hope to—snorkeling in the Great Barrier Reef, going on safari in South Africa, working as a journalist on assignment in Russia. “Determined” underestimates her commitment to making sure sickle cell disease never slowed her down.

In 2014, Gail and Howard were in Brazil when an experience threatened her momentum. She had a sickle cell “crisis”—the name of the severe pain associated with the disease—and had to be airlifted to The Johns Hopkins Hospital.

“When Gail was airlifted to Hopkins and they saved her life, they noticed that she was having vision issues, and they sent some people over from the Wilmer clinic,” says Woolley. At this point, Adrienne Scott, M.D., a retina expert at the Wilmer Eye Institute, entered the picture.

“Gail suffered a hemorrhage in her eye from an advanced stage of sickle retinopathy and lost vision from it,” says Scott. “We were able to operate on her eye, and she was able to regain some of the sight back in that eye.”

Gail Woolley and Scott discovered they had several things in common—primary among them the desire to see a ramp-up in sickle cell research, specifically in the area of sickle cell retinopathy, or the damage done to the retina by the sickle-shaped red blood cells.

Adrienne Scott and Wilmer Director Emeritus Morton Goldberg
cells caused by the disease. Here, a prepared mind (Scott had developed a plan to better identify individuals at risk for losing vision from sickle cell retinopathy) met an unstoppable force: Gail Woolley, who began a philanthropic partnership with Scott, which her husband has carried on since her death.

**WILMER HAS A** storied history in the field of sickle cell retinopathy research thanks largely to Director Emeritus Morton Goldberg, M.D. Goldberg developed the staging system for sickle cell retinopathy during his residency. “The staging of the disease as well as knowledge of its natural untreated course are important because they help the doctor decide whether or not to intervene therapeutically and, if so, when,” says Goldberg. “There are always some risks for every treatment. It’s good to avoid treating unless you actually have to treat.”

When speaking of Goldberg, Scott says, “He revolutionized the field and described a lot of the retinal changes that happen with sickle cell retinopathy.” She points out that he did not have the imaging tools available today, so he accomplished this feat “because of his clinical acumen and observation.”

Fifty-two years later, the Goldberg staging of proliferative sickle cell retinopathy remains the standard for clinicians to classify the progression of a patient’s disease. “Proliferative sickle cell retinopathy, in which abnormal blood vessel growth occurs, is the most common sight-threatening complication of sickle cell disease and can lead to vision loss from bleeding in the eye or retinal detachment,” says Scott.

There are multiple types of sickle cell disease, and a person’s likelihood of developing vision problems depends on the type. The most common forms of sickle cell disease are hemoglobin SS and hemoglobin SC. An interesting aspect of the disease is that while hemoglobin SS patients experience the most severe systemic symptoms, they have a lower risk of vision loss. The prevalence of proliferative sickle cell retinopathy for patients with hemoglobin SS is 20 to 30 percent, while the prevalence for patients with hemoglobin SC, who have fewer symptoms overall, is 60 to 70 percent, according to published literature.

**EVEN AS THE** Goldberg staging system has stood the test of time, technology has advanced quite a bit. “Now we have such interesting and novel and exquisite imaging techniques,” says Scott, referring to optical coherence tomography angiography (OCT-A). OCT-A provides images of blood while it is flowing, which means doctors can identify exact locations where blood has stopped flowing—an invaluable clue for tracking sickle cell retinopathy.

Scott’s research focuses on using these images to track the progression of sickle cell retinopathy in finer detail and further refining the Goldberg staging system with this new information. She believes such detailed images will support her hypothesis that sickle cell disease, and a person’s likelihood of developing vision problems, depends on the type. The most common forms of sickle cell disease are hemoglobin SS and hemoglobin SC. An interesting aspect of the disease is that while hemoglobin SS patients experience the most severe systemic symptoms, they have a lower risk of vision loss. The prevalence of proliferative sickle cell retinopathy for patients with hemoglobin SS is 20 to 30 percent, while the prevalence for patients with hemoglobin SC, who have fewer symptoms overall, is 60 to 70 percent, according to published literature.

It’s an underfunded charity, so if you target your philanthropic dollars into the sickle cell space, you can impact the quality of people’s lives in a meaningful way. —Howard Woolley
A Name to Remember

During a retinal exam of a patient with sickle cell disease, an ophthalmologist looks for abnormal retinal blood vessel growth. The patterns such growth takes indicate which stage, from I to V, the disease has reached. One of the patterns ophthalmologists look for is a “sea fan lesion”—so named by Wilmer’s Morton Goldberg, M.D., and Robert Welch, M.D. “It describes the largely two-dimensional branching proliferation of pre-retinal blood vessels,” says Goldberg.

At the time he was designing the staging system, in 1966, Goldberg was an active skin diver. “I looked at those sea fans and thought—they look just like what I see in the retina of sickle cell patients,” he says. Goldberg decided to name the pattern a sea fan lesion for “ease of remembrance.”

It worked. In a recent editorial from the *Journal of Vitreo-Retinal Diseases*, Editor-in-Chief Donald J. D’Amico, M.D., writes, “Over the years, I have noted that this is the only lesion that is never forgotten by residents and fellows as they seek to master the deities niched in our retinal pantheon.”

Memorable names and accuracy have combined to keep the Goldberg stages of proliferative sickle cell retinopathy relevant and useful. “It’s to my absolute amazement that nothing has changed in that classification over 52 years. It’s not often that you can do something that stands the test of time,” says Goldberg.
THE MERLAU FAMILY’S support for the age-related macular degeneration research of Jim Handa, M.D., is a “tribute,” Handa says, to “altruism.”

At a world-class eye center like the Wilmer Eye Institute, it is not unusual for grateful patients to become donors. But those patients often have a personal stake in the research they choose to fund. This is not the case with Jo and Ken Merlau of Winnetka, Illinois. Ken has a treatable genetic condition that years ago brought the couple to the care of Wilmer’s Handa. The Merlaus came to respect Handa and his work so much that they chose to fund his research into a disease that does not currently affect their close-knit family: age-related macular degeneration (AMD). “That’s a very unusual thing for a donor to do,” Handa says, “but age-related macular degeneration is a big problem for a lot of people. It’s a tribute to the Merlaus’ altruism that they have done this.”

AMD is a degradation of the macula, the part of the eye that sees fine detail. With more than 10 million Americans suffering from AMD, it is the leading cause of blindness in those over 60.

The doctor-donor relationship between Handa and the Merlaus goes far beyond research and funding. “I would say it’s become a friendship over the last 20 years since I first met them,” Handa says.

In contrast to Handa’s reserved, professorial demeanor, Jo Merlau is a ball of energy. She speaks quickly in a distinct Chicago accent about why her family chose to fund Handa’s AMD research. “By any objective measure, Wilmer is No. 1 in research, which translates into the most effective clinical care, defining treatment protocols for the future,” Jo Merlau says. “Dr. Handa’s research pioneers potentially game-changing research early on, which we are happy to be a part of.”

The Merlaus’ interest in Handa’s research includes the way Handa has chosen to build a highly diverse team of researchers in his laboratory. “Somewhere along the way, the Merlaus and I had a conversation about the cultural makeup of the lab,” Handa says. “We agreed that we wanted international representation, and the Merlaus have since funded an international scholarship to further that goal.” Handa proudly says his team includes researchers from China, India, Poland, Venezuela and Nigeria. “We think medicine has a global impact, and we think it’s important to be inclusive,” Jo Merlau says. “Jim’s reputation has led to the international community finding him. It’s a testament to who Jim Handa is. He at-
tracts the best people to Wilmer from across the world.”

In his AMD research, Handa is pursuing a couple of intriguing avenues by trying to figure out the root mechanisms of the oxidation that kills cells in the retina and causes a loss of vision. “We’re looking at the genes involved in how the retina protects itself from oxidation and how that ability fades as one ages,” Handa says.

While his research holds promise, there are no guarantees, and Handa is reluctant to hype himself or the work. He prefers instead to underpromise and overdeliver.

Jo Merlau is less reserved when speaking about Handa himself. “We think Jim Handa is one of the best doctors around, and we want the whole world to know more about him and his research,” she says. “ Clinically, intellectually, personally and how he cares about his patients—that became my driving force for giving. He’s in the rare air.”

Age-related macular degeneration is a big problem for a lot of people. It’s a tribute to the Merlaus’ altruism that they have done this.

—Jim Handa
Board of Governors Meeting
Nov. 1, 2017 / Wilmer Eye Institute’s Robert H. and Clarice Smith Building

Wilmer Director Peter J. McDonnell, M.D., and Chairman of the Board Sanford Greenberg, Ph.D., welcomed Wilmer’s Board of Governors for an update on the Institute and presentations by faculty members. Jeffrey Mumm, Ph.D., gave a talk, “A ‘Fish Eye View’ of Regenerative Therapeutic Strategies for Blinding Conditions,” about his work with zebrafish and how the immune system is implicated in this species’ remarkable capacity for regeneration. Fasika Woreta, M.D., M.P.H., spoke about the past, present and future of the Wilmer Eye Trauma Center as its newly appointed director.

Dedication of the Helen Larson and Charles Glenn Grover Professorship in Ophthalmology
Nov. 1, 2017 / Wilmer Eye Institute’s Robert H. and Clarice Smith Building

Jeffrey Mumm, Ph.D., received the Helen Larson and Charles Glenn Grover Professorship in Ophthalmology. During his talk, he spoke about the instrumental role his wife, Meera Saxena, Ph.D., has played in his research and in the company they both founded, Luminomics Inc. He also discussed his lab’s ongoing work, which is focused on understanding how neural circuits are formed, how they function and how they can be regenerated in order to develop new therapies for retinal regeneration.

Left to right: Board of Governors members Mary Bartkus, Becky Stirn, Allan Jensen and Claire Jensen, Senior Director of Development Libby Bell, and Wilmer Director Peter J. McDonnell

Left to right: Nalani Mumm, Jeffrey Mumm, Malayna Mumm and Meera Saxena
Wilmer faculty and staff members came together with Wilmer alumni in New Orleans for a reception held during the annual meeting of the American Academy of Ophthalmology (AAO). The world’s largest association of eye physicians and surgeons, AAO is a global community of 32,000 medical doctors that sets standards for ophthalmic education and advocates for patients. More than 50 Wilmer faculty members made presentations during the multi-day conference, demonstrating Wilmer’s leadership in the field of ophthalmology worldwide.

Wilmer Reception
at the American Academy of Ophthalmology Annual Meeting

No. 11, 2017
New Orleans, Louisiana

Wilmer residents at AAO, left to right: second-year resident Angeline Nguyen, third-year resident Kathleen Jee, second-year resident Inna Stroh and second-year resident Angela Zhu

Left to right: Rubens Belfort Neto, affiliate professor of ophthalmology at the Federal University of São Paulo; Andrew Carey, assistant professor of ophthalmology at Wilmer; and Samuel Yiu, associate professor of ophthalmology at Wilmer
What’s New and What’s Next in Glaucoma?

Nov. 18, 2017 / Wilmer Eye Institute’s Robert H. and Clarice Smith Building

Pradeep Ramulu, M.D., Ph.D., chief of the Glaucoma Division, along with a team of experts from the Glaucoma Center of Excellence, hosted a research update and lab tour. Friends of the Glaucoma Division and interested parties gathered to learn firsthand about the work of the division’s doctors and researchers related to optic nerve regeneration, ways to avoid falls and injury, efforts to replace glaucoma drops, and studies on new surgical techniques for glaucoma patients.
Vision for the Future

In 1925, the nation’s first university eye clinic to combine eye patient care, research and teaching was established, thanks to the generosity of friends and former patients of William Holland Wilmer. Your legacy gift will ensure Dr. Wilmer’s legacy continues through education, treatment and pioneering research. Consider these opportunities to leave a meaningful legacy while taking into account your personal goals.

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Seek advice from a tax professional before entering into a gift annuity agreement. Johns Hopkins gift annuities are not available in all states.
Dedication of the Branna and Irving Sisenwein Professorship in Ophthalmology

April 19, 2018 / Wilmer Eye Institute’s Robert H. and Clarice Smith Building

Akrit Sodhi, M.D., Ph.D., received the Branna and Irving Sisenwein Professorship in Ophthalmology. After Wilmer Director Peter J. McDonnell, M.D., spoke about his relationship to the late Branna and Irving Sisenwein and their unwavering devotion to improving the lives of people with vision impairment, their niece, Elin Gursky, spoke about her aunt and uncle’s zest for life. Sodhi explained how his work with HIF inhibitors has the potential to accomplish the Sisenwein’s goal of stopping damage from macular degeneration.

Legacy Society Luncheon

April 19, 2018 / Wilmer Eye Institute’s Robert H. and Clarice Smith Building

Karen Wimberly spoke at Wilmer’s Legacy Society Luncheon, which honors members of the Johns Hopkins Legacy Society. The latter celebrates individuals who make a life-income gift or a bequest to the institution. Wimberly expressed her gratitude to Wilmer’s Jennifer Thorne, M.D., Ph.D., for her treatment of Wimberly, and David Knox, M.D., for his treatment of her mother, Grace Bakerjian.
Live the Mission: A Look Through Our Lens

April 20, 2018 / Wilmer Eye Institute

Wilmer Director Peter J. McDonnell, M.D., hosted a half-day event that offered an opportunity for Wilmer supporters to learn directly from our leading clinician-scientists and researchers. Attendees saw firsthand how collaboration advances ophthalmic knowledge and care for individuals with vision loss at home and around the world. After observing a mock Grand Rounds, featuring Wilmer’s Eye Trauma Center, attendees participated in two of the three following workshops: surgical training in our wet lab and simulation center, a low-vision device demonstration at the Lions Low Vision Center, or a laboratory tour and presentation by researchers in the Center for Nanomedicine.

Board of Governors Meeting

April 19, 2018 / Wilmer Eye Institute’s Robert H. and Clarice Smith Building

After a welcome from Chairman of the Board Sanford Greenberg, Ph.D., and an update on the Institute, the Board of Governors listened to presentations from two faculty members. Akrit Sodhi, M.D., Ph.D., who had just received the Branna and Irving Sisenwein Professorship, discussed his work with HIF inhibitors. Then Michael Repka, M.D., M.B.A., the David L. Guyton, M.D., and Feduniak Family Professor of Ophthalmology and chief of Wilmer’s Pediatric Division, presented on the challenges and opportunities in the field of pediatric ophthalmology.

In Wilmer’s Center of Excellence for Ophthalmic Surgical Education and Training wet lab, Nicholas Mahoney, assistant professor of ophthalmology, helps Live the Mission participant Eric Warren Goldman try his hand at suturing.
Wilmer Eye Institute
Board of Governors

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Novelist Booth Tarkington, considered one of America's greatest living authors in the 1910s and 1920s, held court with newspaper reporters at the Wilmer Eye Institute after undergoing cataract surgery in January 1931. Delightedly demonstrating that he could now “distinguish color and form,” the two-time Pulitzer Prize for Fiction winner “proved to [the] newspaper men [sic] that his 14-year battle against blindness, [sic] had been crowned with success.”