

LEAP

Johns Hopkins University
School of Medicine
Division of Rheumatology
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PRECISION MEDICINE

SJÖGREN'S: THIS CHANGES EVERYTHING

CHRONIC CARE

SCLERODERMA AND IFI16 ANTIBODIES:
A DETECTIVE STORY

You may notice a theme to this issue of *LEAP*: change. Our cover story features research in Sjögren’s that has the potential to transform the way drugs are tested in clinical trials, to eliminate some of the time-consuming, trial-and-error process that is so familiar to patients and doctors alike, and even to lead to the development of new drugs that target the molecular mechanisms of disease. Like the Rubik’s cube on the cover, Sjögren’s – like many rheumatic diseases – is a complex puzzle with multiple immune pathways; in turn, each patient’s disease is an individual piece of that puzzle. This research, led by Livia Casciola-Rosen and Alan Baer, is precision medicine on the molecular level. It is exciting, it is revolutionary, and it is definitely a change for the better.

New discoveries by Zsuzsanna McMahan and colleagues may help limit the damage from severe Raynaud’s phenomenon in scleroderma. This work has the potential to help prevent the devastating loss of function from digital gangrene. As part of our ongoing commitment to tell the stories of people – those who work here and the patients we care for, and think about, and are committed to helping – we are sharing the stories of two of our patients who are coping with serious autoimmune disease. They are inspiring to us, and we hope they will be to you, as well.

Also changing: the way you see the doctor. Precision is not limited to molecular pathways. Patients have enormous experience of their disease and lives, and medicine does not adequately access this unique perspective. Clifton Bingham is leading a new program in rheumatoid arthritis, creating and applying new tools to help patients quantify and communicate what’s really happening in their lives – what they’re not doing because of fatigue, for example – and helping physicians to ask questions about things that otherwise might not be addressed during the visit. Thomas Grader-Beck is spearheading a pilot program to help physicians be more responsive to their patients, and to allow patients to keep their physician updated on how they’re doing, in a highly nuanced way.

And finally, there’s the story of how our clinic has been beautifully updated and, in fact, transformed. Those of you who remember how it used to be may have done a double-take the first time you saw it after the makeover. We know that we are providing the same great care, but it is nice to do it in a setting worthy of our patients, our partners in discovery and care. I came across a *LEAP*-related quote by Harvey Mackay that seems to capture how we feel here: “A great accomplishment shouldn’t be the end of the road, just the starting point for the next leap forward.”

Looking forward to new roads of discovery and more good changes,

Antony Rosen, M.D.
Director, Division of Rheumatology
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SJÖGREN'S

PRECISION MEDICINE

Two people with Sjögren's syndrome can have identical symptoms, and one may respond well to a drug and one won't. The results of a recent study by Livia Casciola-Rosen, Ph.D., Alan Baer, M.D., and colleagues tell us why: The Hopkins scientists have discovered that although patients might look similar on the outside, on the inside they don't have the same disease at all. For the doctors who treat people with Sjögren's, and the scientists devoted to finding and testing new medications to modify the disease, these findings, published in the journal, *Arthritis & Rheumatology*, are revolutionary.

standing what is specific to the disease," she explains. The first finding was that only about 60 percent of Sjögren's patients had evidence of high interferon activity; these patients had evidence of more severe disease. The remaining 40 percent had milder disease. "If patients had evidence of high interferon, the different molecular patterns were clinically indistinguishable. Yet our assay was able to convey that these subgroups have very different types of interferon activity going on in the tissue – and that is amazing, because that also means that they're likely to respond to different medications."

The ramifications of this discovery are huge. Take clinical trials, for instance: "To date, most of the clinical trial selection criteria have been based on broad clini-

THIS CHANGES EVERYTHING

TWO OF THE MAIN targets of Sjögren's syndrome are the glands that produce saliva and tears. These are disease-targeted tissues, and they provide a wealth of clues – information not forthcoming in a blood test – if scientists ask the right questions and know how to find the answers. "All patients have inflammatory infiltrates and abnormal secretory functions of their salivary and lacrimal glands, even though the clinical features of the disease can vary," says Casciola-Rosen.

In previous work published in the *Proceedings of the National Academy of Sciences (PNAS)*, Casciola-Rosen and colleagues developed molecular probes that analyzed tiny bits of frozen salivary tissue from individual patients and determined which particular type of interferon pathway was active in each. One probe detects Type I (alpha) interferon, which can be made by

many cells; the other detects the presence of Type II (gamma) interferon, which suggests that certain immune cells, particularly T cells, are active. Although both types share the name interferon – as their name suggests, their job is to "interfere" with the life cycle of a virus or other foreign invader – they are distinctly different.

Molecular subsets: In this most recent work, the scientists set out to learn more about the molecular differences in people with clinically well characterized Sjögren's syndrome. "This is a precision medicine approach," Casciola-Rosen says, "a way of trying to be more precise about what's going on with the patient's disease at a molecular level." They looked at salivary gland tissue samples of 53 participants with Sjögren's, and 29 in a control group. "The controls are really important for under-

cal features, and trials in Sjögren's have been disappointingly unsuccessful" says Casciola-Rosen. "But the subgroups of patients with Sjögren's – who have evidence of activity in different interferon pathways that mark distinct disease mechanisms – are not clinically distinguishable. We are proposing that if you know what's going on in these patients at the molecular level, you may observe much more effective responses to mechanism-targeted therapy." If, say, a woman takes Drug A, which targets Type I interferon, but she only has Type II activity, then she is most likely not going to be helped by that particular drug.

From a clinical standpoint, "we know that there is considerable heterogeneity in Sjögren's syndrome," says Baer. "There may be differences related to age; the younger patient may have greater salivary gland

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enlargement and less dryness – findings opposite to what is often seen in older patients. There are differences between men and women, and differences determined by the types of antibodies a patient might have. Differences in disease patterns such as these suggest that the disease may have come about in different ways. That automatically tells us that we need to individualize treatment.”

Also: “We are frustrated in treating Sjögren’s syndrome,” Baer continues, “since we don’t have any medications that we know reliably alter the natural history of the disease.” Instead, most drugs just treat the symptoms. “So we are eager to have good treatments that can actually affect the outcome of the disease. We’ve accomplished that well in diseases such as rheumatoid arthritis (RA), and we want to expand that wonderful experience to patients with Sjögren’s. Unfortunately, in some of the early trials using some of the medications that were very successful in RA, we’ve been disappointed. They haven’t worked in Sjögren’s.”

But it may well be that “the people who did not respond in those clinical trials didn’t respond because they didn’t have the right molecular profile,” says Casciola-Rosen. “They were the wrong subset.”

What’s unique about this work, says Baer, “is that with a very small amount of salivary gland tissue from a lip biopsy, we’re able to determine information that is very important in that particular patient.” Being able to study the tissue that’s directly affected

by the disease is crucial – and a blessing not afforded scientists studying a disease such as lupus, in which the kidney is affected. It is much easier to take small bits of tissue from the lip than to biopsy the kidney.

What’s next? To build on this groundwork and start a clinical trial of medications that are directed at very specific pathways, and “really test what we’ve shown might be feasible here,” Baer says. “Which is, with careful selection of patients, to do clinical trials of specific agents that are directed at these very specific pathways, and evaluate what would happen.” This is not what goes on in most clinical trials, he notes. “You give a drug, then you get blood samples, but you don’t really have a handle on what’s happening at the level of

the target tissue. What we’re at least theorizing here is that you could obtain very small amounts of target tissue during the course of a clinical trial, and have a very direct look at what’s really happening at a molecular level as the trial is going on – before, during, and after.” In the future, as technology gets better, it might be possible to obtain just a few cells with a needle, instead of taking a small core of tissue.

Even more promising: The scientists feel certain that what they have learned can be applied to other autoimmune diseases. “We are very excited about the possibilities,” says Casciola-Rosen.

Other co-authors of the most recent paper are John Hall, Ami Shah, Lindsey Criswell, Caroline Shiboski, and Antony Rosen. This work would not have been possible, the scientists say, without the Jerome L. Greene Foundation, which established the Sjögren’s Syndrome Center at Johns Hopkins and helped fund this research. Casciola-Rosen and Baer also credit the SICCA cohort for supplying tissue samples. Funded by the National Institutes of Dental and Craniofacial Research, this is an international bank of data collected from more than 3,500 Sjögren’s patients in seven countries. [L](#)



Casciola-Rosen: “We are very excited about the possibilities.”

MY STORY

LIVING WITH SJÖGREN’S

Virginia* is a Sjögren’s patient of rheumatologist Alan Baer, M.D. Recently, she was kind enough to talk to *LEAP* about how she has learned to cope with her illness. She is upbeat, but she struggles with depression; she works at a difficult technical job, although some days the “brain fog” that can accompany Sjögren’s makes her feel that she can’t think. There are good days, and days when she feels like an elephant is standing on her chest. She has learned to take all of this in stride and achieve a balance. Here’s some of what she had to say:

“I was officially diagnosed with Sjögren’s when I was 25, but I’m pretty sure that the illness started before then,” during the spring of her freshman year in college. “I had an unexplained illness that caused extreme fatigue, and my tongue felt weird. It was severe enough that I had to withdraw from that semester, and go home and rest up before I could resume my college career.”

Just before Virginia was diagnosed, she had a couple of rashes; big, raised welts on her forearms, and a butterfly rash on her face. Bloodwork revealed that she had an autoimmune disease, and her doctor later refined the diagnosis to Sjögren’s. “So, depending on how you measure the start date, I’ve had it anywhere from 15 to 20 years. I’m currently 40. That is a long time.”

For Virginia, although she has other problems related to Sjögren’s – eye, mouth, dental, and digestive issues – her main concern is fatigue. “It is my number one most day-to-day disabling symptom, period, hands down,” she says. “I have far less energy than a person my age should have, and with Sjögren’s, the fatigue affects everything – my ability to take care of myself, to have a job, to take care of things like going to the grocery or cleaning the house, to travel, socialize, attend family gatherings.”

The fatigue is unpredictable. “I don’t realize that I’ve crossed the line and overexerted myself until it’s too late, until it’s already happened.” Then, a day or two later, she wakes up with a “fatigue

hangover,” and “it’s like an elephant sat on me,” she says. “The fatigue has really ramped up, and I’m thinking, ‘Oh, my God, I have to get out of bed, but how am I going to do that?’ It’s that kind of fatigue. Although blood and lab tests can measure Sjögren’s activity, “there’s no way to measure fatigue. You have to just feel your way through it day by day.”

The fatigue is its own entity: “There is this ‘Do not cross’ line. You don’t want to push yourself too far on days when you feel good, or you will accidentally overdo it. It’s a guessing game. I’m sort of a Type A person; I like challenges, I like trying to achieve particular things in my job, my family and my community. And when you have to wonder, how hard can I push myself *today*, and how much energy do I have *today*, it gets very hard to build your life with that level of uncertainty.” There may be a couple of weeks in a row when she feels pretty good, and “I’m cruising along and maybe I’m even starting to adjust my life to embrace that energy, and then it’s like somebody pulls the rug out.”

Virginia lives with her husband and one dog. “I’m very lucky,” she says. “I have a good support network, and I’m able to use my husband’s insurance benefits – which is good because I’m only able to work half days right now.”

If it’s possible for the fatigue to have a dark side, it is the mental component. “There is a distinct phenomenon called brain fog, where it’s like your thoughts have slowed down, and your brain isn’t working at its best. On those days, you

“It’s like an elephant has sat on me, and I’m thinking, ‘Oh, my God, I have to get out of bed, but how am I going to do that?’ It’s that kind of fatigue.”

might also be feeling physically fatigued, and that’s a one-two punch.”

Like many people with an autoimmune disease, Virginia has tried a wide variety of medications. When she talks about her disease and her treatment, she speaks of Baer as a partner. After the drug Methotrexate caused some liver problems, they tried Plaquenil for a while. “That stopped being enough, so Dr. Baer and I have tried things like prednisone every other day, but that’s not good for the long term. We’ve tried Benlysta intravenously, and that worked very well until early last year. But then Dr. Baer was concerned that my bloodwork numbers were progressing. So I switched to IV Rituxan this year. It has not had the effects that we hoped, and I’ve been more tired. There’s a lot of trial and error. Many treatments work for a while, but then aren’t enough as your symptoms get worse. Or, you try something different and it doesn’t work for you at all.

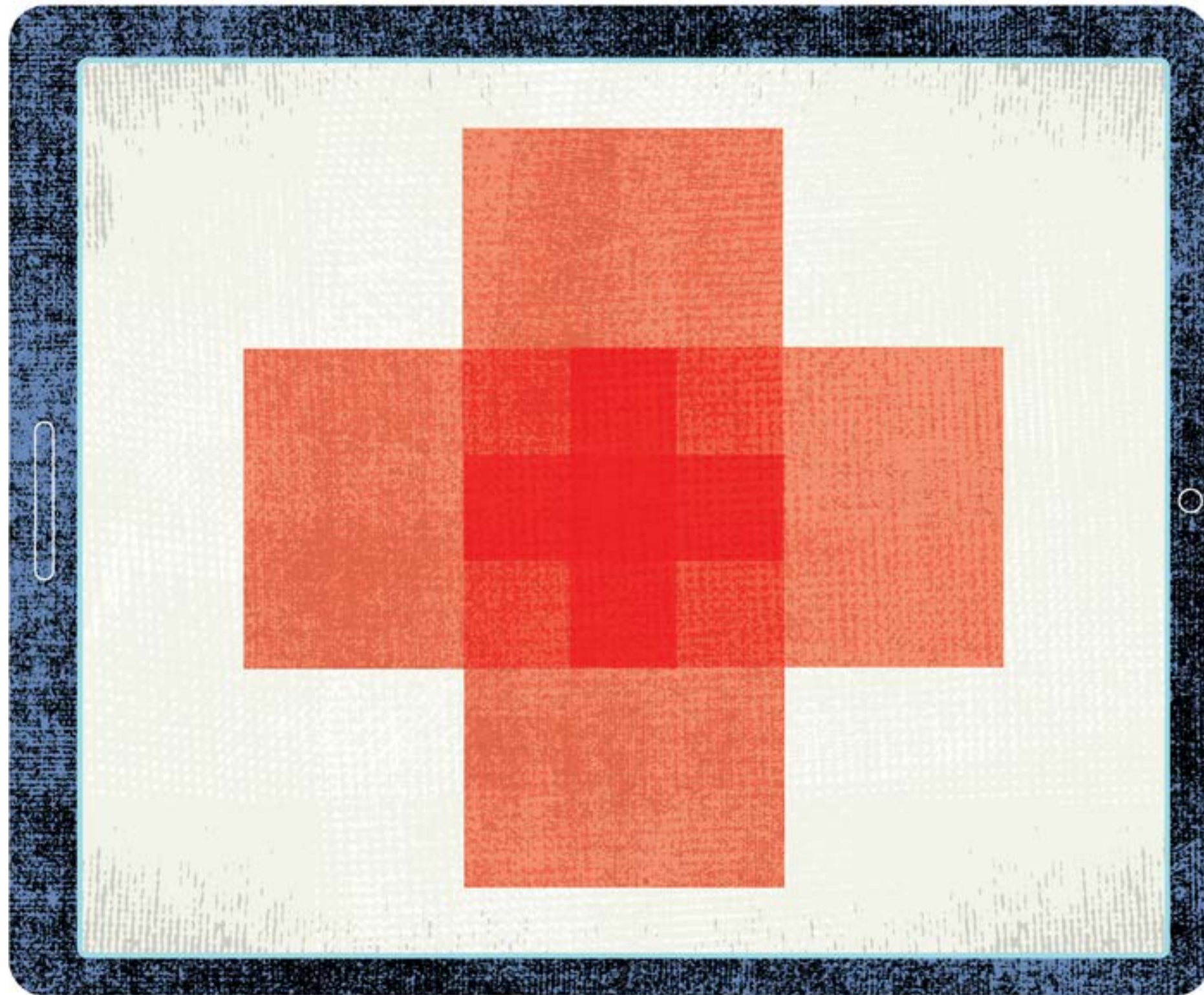
“This is going to sound right out of a PR booklet, but I feel very lucky to live in Baltimore and have access to the Sjögren’s Center, because I think doctors like Dr. Birnbaum and Dr. Baer are really addressing the illness and advancing the research, and trying new approaches and new ideas, and things that will hopefully make a difference.” She sees Baer every six months, and “his appointments with me are always essentially the same thing. He’ll say, ‘How are you feeling?’ I say, ‘I’m really tired. It’s really a problem.’ He’ll say, ‘I know, I know, maybe we’ll try something different.’”

Ultimately, she says, it all comes down to finding that balance “that lets you feel as well as you can and do as much as you can, without wearing you out too much or having a negative health effect. Some days I’m feeling pretty good about the whole thing, other days I’m not. It varies. It’s a moving target.”

**Name changed to protect the patient’s privacy.*

CHRONIC CARE

The way you see the doctor – the way your parents and grandparents and their parents saw the doctor – is about to change. If you’ve been in the Rheumatology clinic lately, you may have seen it changing already. Tablets and iPads have come to the clinic. They have questions for you, things you might forget to mention during your visit, questions your doctor might not know to ask. Your doctor might follow up with an e-mail at home, or might send you a copy of that study you talked about during the visit. Instead of care that has long been episodic – from appointment to appointment to lab test – the doctor-patient relationship is becoming more of a continuum. It’s chronic care for chronic diseases.



Rheumatologist Thomas Grader-Beck, M.D., in addition to treating patients with rheumatoid arthritis, Sjögren’s syndrome, and other rheumatic diseases, is a “physician builder.” He is working with the electronic medical records (EMR) system to find ways to make communication between doctors and patients more personalized, so that physicians can be more responsive to their patients, and patients can keep their doctors updated on how they’re doing in between visits.

“We’ve always gotten patient information on paper,” he says. “We would mail patients questionnaires, or when they come to the clinic, give them questionnaires to fill out. We would document the findings in our notes.” But the system wasn’t perfect; some people never got their questionnaires in the mail, some forgot or didn’t want to take the time to fill them out. “Now with the EMR systems, we can not only collect that information much more efficiently, we can also tailor the questions toward individual patients.” In a pilot project supported by the Ira Fine Discovery Fund, Grader-Beck is giving patients tablet PCs. The information they provide goes right into the EMR, and then when they see their doctor, they can review those answers together. “Also, patients can see how it compares to what they’ve answered in the past and get the perspective of their disease.” He’s studying how the doctors like this process, too – whether they feel that it indeed saves time, whether they get more or better information from their patients than before.

Grader-Beck also foresees more frequent doctor-patient interaction through patient portals, secure, web-based sites that also give patients access to their own health information. “With the development of a patient portal, medical care becomes much more continuous,” he says. “Before, we were able to find out how patients are

“For patients with a chronic disease, we see them every three to four months, but that is really an old model.”



Grader-Beck: Communication is becoming “much more continuous.”

doing at the visit. Now, we have the opportunity to gather information in between visits – when do they have flares, how much pain do they have. The doctor can send patients an e-mail and ask how they’re doing. I do that now with my patients.”

The pilot tablet project is with arthritis, “but I’m also part of the Sjögren’s Center, and I’ve started to develop questionnaires for those patients, as well.” The project is small – there are only four tablets right now – but with more funding, Grader-Beck hopes to purchase as many as 20 tablets.

“I have this vision,” he says. “We have patients with arthritis, lupus, Sjögren’s, and other diseases. Each patient is different; no one patient is like the other. In Sjögren’s, say someone doesn’t have dry mouth or dry eyes. But patients get a standard questionnaire about general things that may not be related to their disease at all. We can develop questions that are much more relevant to them, and ask in much more detail, and then use our visits to get much better information and address what the patients’ concerns are. That’s where I would like to go.”

This is a new idea, and there aren’t really any guidelines for outside-the-office communication. “You can imagine,” says Grader-Beck, “if all your doctors are connected with you, you have a different approach to talking to your doctors.” Communication is becoming “much more continuous. For patients with a chronic disease, we see them every three to four months, but that is really an old model. Nowadays, it is much easier to respond to changes in the patient’s health, and because you are getting more information, you can actually focus the patient care to when it’s needed. It may not always be necessary for every patient to come in every three months; maybe someone needs to come in once a month, maybe not. We can tai-

PATIENT-CENTERED MEDICINE

Rheumatologists can look at numbers until the cows come home. They can determine precise things – such as how many swollen and tender joints their patients with rheumatoid arthritis are dealing with; and which inflammatory markers, as determined by lab tests, are in their blood, and what those numbers are. “Those numbers have been determined by experts as the best outcomes to measure,” says rheumatologist Clifton Bingham, M.D., Director of the Arthritis Center, and of the Center for Patient-Centered Outcomes Research in Rheumatology “But they don’t always reflect what’s really happening in our patients’ lives – what they’re *not* doing because of pain, or fatigue, or depression, or something else. And maybe the whole visit would come and go and we wouldn’t know something was wrong because we didn’t ask the right questions.”

Numbers don’t tell the whole story, and the goals between patients can vary widely, says Bingham. A couple of years ago, Bingham was one of a few in the country to receive Federal funding for a pilot project from the Patient-Centered Outcomes Research Institute (PCORI, pronounced “picori”). In clinic patients with RA, he implemented an interactive questionnaire that tackled not only physical manifestations of disease, but lifestyle issues – activities at home and work, and how satisfied patients feel with their ability to function in various areas. On iPads, patients filled out the questionnaire before they saw the doctor in the clinic, and together, after the regular check-up, they looked at the questionnaire to see what might have been missed in the visit. The questionnaire, Bingham found, “enabled conversation that might not have happened otherwise.”

Recently, he and his team received additional PCORI funding for a second project to expand this work and move it forward. Bingham hopes that what he and colleagues are learning about incorporating patients’ wishes and concerns into their health care plan will expand to include other rheumatic diseases, including myositis and Sjögren’s syndrome, and they plan to expand the research to include other diseases, as well.

“Physicians consistently underestimate the magnitude or impact of symptoms on the patient. We hope that through this work, we can provide clinicians with a better understanding of how a disease is affecting the patient, particularly as the disease changes over time.”

lor the frequency of visits to the patient’s needs. It’s changing dramatically.”

Having more information to work with means that physicians need to be good analysts, able to put it all together – patient-reported outcomes, results of tests, the

physical exam, and what the patient says during the visit. “I think we’ll really increase our understanding of how the patient is overall, on a week-to-week or even a day-to-day basis. The more I know about my patients, the more I can help them.” L

SCLERODERMA AND IFI16 ANTIBODIES: A DETECTIVE STORY

Imagine you are a detective and you make a startling discovery: footprints. Other investigators have noticed them, too, and not made much of them. But you take a different approach, and you realize that these footprints are connected to a whole string of break-ins. Rheumatologist Zsuzsanna McMahan, M.D., a Greene Scholar, is not investigating crimes, but she is studying footprints. These footprints are antibodies to a protein called IFI16 (short for “interferon inducible protein 16”), which is expressed at high levels in the lining of blood vessels. With colleagues Livia Casciola-Rosen, Ph.D., Fred Wigley, M.D., and Ami Shah, M.D., McMahan, who specializes in scleroderma, has been working to understand the clinical significance of these antibodies in scleroderma.

As many as 30 percent of people with scleroderma have these antibodies in their blood, “but they’ve never really been tied to any particular clinical features, other than limited skin disease.” In an initial study, the investigators used the Division’s own longitudinal database of blood samples, collected every six months from thousands of patients. They looked for the presence of these antibodies in 94 patients with scleroderma and in a control group of 47 people without the disease. First, “we found that levels of IFI16 antibodies were significantly higher in the people with scleroderma compared to the healthy controls,” says McMahan; only one out of the 47 people without scleroderma had the antibodies, but 18 percent of the people in the scleroderma group had them.

Then, “we looked to see what the clinical associations were between the presence of this antibody and the features of scleroderma that these patients had.” Not only did having the antibodies seem to go along with limited skin disease, but “with really severe Raynaud’s syndrome,” a vascular problem “so severe that it resulted in digital gangrene.” They also found that the people with anti-IFI16 antibodies had a significantly lower DLCO than those without it; DLCO is the extent to which oxygen is able to diffuse from the lungs into the blood. Low DLCO is considered a marker of vascular disease in the lung, “and this is interesting, because Raynaud’s is a vascular problem.”

These findings were intriguing, but the study was small, and the scientists needed to know more. Next, they carried out a small pilot study “a quick-and-dirty analysis” – to find out whether the antibody levels changed over time, and if this had anything to do with an episode of severe Raynaud’s. “We noticed a trend,” says McMahan. “It seemed that the

“This is all new. It’s still being explored; nobody even knew this association existed before.”

antibodies were highest around the time of an acute ischemic episode or episode of bad Raynaud’s. So we said, ‘isn’t that interesting? Maybe the antibodies correlate with disease activity.’” In a follow-up case-controlled study, “we went back to our database and selected patients with digital gangrene who had blood drawn within six months of the episode. The control group was made up of scleroderma patients with a history of Raynaud’s alone.” There were 39 matched pairs of patients.

Before this work, Italian scientists had published data that showed the presence of these IFI16 antibodies in people with scleroderma, “and they did look for other clinical features associated with the antibodies, but weren’t able to find them,” says McMahan. “Our thought was that maybe they couldn’t find any associations because they didn’t look at the specific timing, and they didn’t examine blood from the time of the event.” In comparing the two groups, the Hopkins investigators found that there is indeed an association between the antibodies and the digital gangrene – that patients with digital gangrene have higher levels of the antibody than patients who don’t have it. Even more interesting, the people with the highest levels of antibodies had the highest risk of gangrene; those with the next highest had a medium risk, and then the next highest had a lower risk.” People who had their blood drawn within six months of the episode of digital gangrene had the highest levels of the IFI16 antibodies, but the levels were still elevated in patients whose blood was drawn within two years. “Antibodies don’t typically go away quickly,” explains McMahan, “so two years is a reasonable window.”

All of these findings were retrospective, looked at after the fact. “It will be important to confirm these findings in a longitudinal, prospective study,” she adds, “to help us understand whether we can one day use IFI16 antibodies as markers of vascular disease in scleroderma.”

What do the findings mean? Are the antibodies a marker – a reflection of an injury? Or do they actively contribute to the vascular disturbance that leads to gangrene? In many autoimmune diseases, such autoantibodies are present in the blood before diagnosis, sometimes years before there are any clinical symptoms. “One of the challenges in scleroderma is that we do not have a comprehensive marker of disease activity,” says McMahan. “We can’t look at a blood test and tell if the disease is active in the blood vessels, GI tract, or heart or lungs. We can’t detect early disease activity in many patients, which means we are limited in our ability to catch a problem early and take steps to treat it. This is all new. It’s still being explored; nobody even knew this association existed before.”

Are the antibodies a marker – a reflection of an injury? Or do they actively contribute to the vascular disturbance that leads to gangrene?

And maybe the most important question of all: could a rise in these antibodies actually predict a vascular event? Could the antibodies be the rheumatologist’s equivalent of a seismograph – to foresee impending gangrene, instead of an earthquake? “If we were able to prove that,” says McMahan, “then potentially, the minute patients start having pain, we could increase the blood flow to the finger with various medications, and think about adding higher-level aspirin or other anti-platelet agents. We wouldn’t do that for everybody, but losing a digit or more can cause a devastating loss of function. If we could come up with a test that would help us predict risk, we might one day be able to limit the damage or even prevent our patients having to go through this.”

These studies were sparked by recent work in Sjögren’s syndrome by Jungsan Sohn, Ph.D., Casciola-Rosen, and colleagues, that identified IFI16 as a common target of the immune system. The investigators looked at scleroderma, and struck paydirt; once again, IFI16 was a target. Future studies of these antibodies may lead to new ways to detect, monitor, and treat symptoms of several autoimmune diseases. [L](#)

LIVING WITH SCLERODERMA

Beth Skinner is a longtime scleroderma patient of rheumatologist Zsuzsanna McMahan, M.D. Recently, *LEAP* had the privilege of talking to her about her illness, but the conversation turned out to be more about her wellness. Because although Skinner has scleroderma, that’s not who she is, and although it has changed her life, she hasn’t let it define her. Here’s some of what she had to say:

“When I was in my twenties, I started experiencing Raynaud’s phenomenon. I didn’t know that was the name for it, I just knew that if I got cold, my fingers and toes would turn white or blue and stay that way. I wasn’t too worried about it until I started developing wounds that wouldn’t heal. Eventually I had to have a toe amputated, and I thought, ‘Wow, that’s a big thing, that’s a big change to my body.’”

What was happening? Skinner, who was in graduate school at the time, went to many doctors, and there didn’t seem to be a clear answer. She was even put on chemotherapy for a while. “I was taking medicine that was making me sick and I’d throw up for days. They didn’t really know what it was.” Possible diagnoses included vasculitis and bacterial infection. “In the meantime, I kept having more and more problems, for four or five years.”

Scleroderma came up as a possible diagnosis. Skinner went to the library. “I remember looking up scleroderma in an old medical book, and being really scared. The book was talking about the mortality rate and different symptoms. It seemed to me not just life-changing but a potentially life-ending kind of diagnosis.”

Skinner earned her degree in communications and got a job at Towson University as a debate coach, and soon afterward, she was diagnosed with scleroderma. “It was scary, but also great to have a diagnosis and finally know what was wrong with me. That’s when I got hooked up with Dr. Fred Wigley and the Scleroderma Center, and that’s how I met Dr. McMahan. Even though I’ve had further health problems, it has helped me to

know that the people I’m working with are the best people in the world that I could be getting medical advice from.”

She has continued to struggle with Raynaud’s. “I have had more surgeries and amputations than I can count. I have three fingers left on my left hand, and none on my right. I had transmetatarsal amputations on both feet, which means that the front half of the foot, all of the toes and across the bridge of my foot, are cut off. That has affected my mobility, my ability to work, to do the things that I like. But because it’s happened over a long period of time – it was not some sudden, traumatic thing – I’ve had some time to adapt to it.

“I remember looking up scleroderma in an old medical book, and being really scared.”

“The thing that I want other people to know is, even if you’re dealing with pretty difficult symptoms and you feel that it’s not within your control, you can learn how to do things differently. I like making jewelry, so I’ve gotten some adaptive tools, and I continue to do some of the things I like: cooking, art projects, things that bring fulfillment to my life. I’ve also been volunteering, trying to use some of my knowledge and expertise to help people.”

Recently, Skinner has been moved by stories of immigrants coming to America, “so I started volunteering with the International Rescue Committee, which helps to resettle refugees.” She hopes she

“The thing that I want other people to know is, even if you’re dealing with pretty difficult symptoms and you feel that it’s not within your control, you can learn how to do things differently.”

can be particularly useful in helping new residents navigate the medical system. “It can be pretty overwhelming; I hope I can use the benefit of my experience. Even though it’s not something I would have wished for myself, I feel it’s something I can use to help people.”

Skinner says that she feels especially fortunate to be McMahan’s patient. “She’s very compassionate, she listens when patients talk. One of the things that’s difficult when you’re dealing with specialists, at least in my opinion, is that sometimes people know so much about the technical, theoretical, medical side of things that they have trouble communicating with patients and explaining, breaking things down, trying to help people understand what’s happening. She’s great with that.”

Last year, at McMahan’s recommendation, Skinner attended a rheumatology conference in Washington, D.C. On the trip, she met U.S. Senator Ben Cardin, and got to talk to him and other officials about how important NIH funding is to people with rheumatological diseases, and “how much we appreciate their support for research that can help make people’s lives better.”

When *LEAP* interviewed her, Skinner was preparing for a trip the next day to Jamaica. If you have been diagnosed with scleroderma, this is what she wants you to know: “No matter what your symptoms are, you can learn to live with those things. You can work with a good medical team, and if you have the support of the friends and family who are around you, it’ll be okay.”

The waiting room, never big enough and always crowded, seemed tiny with the increase in patients; sometimes patients had to stand in the hallway because there was no other place to wait.



NEW CLINIC

TRANSFORMED

What does the clinic of the top-ranked Division of Rheumatology in the country look like? For years, frankly, it looked outdated. It was also not big enough to handle the patients who come to Johns Hopkins Bayview from around the world; there weren't enough exam rooms, the layout made it awkward for patients to get

to phlebotomy to have their blood drawn, and the waiting room was too small. There was a satellite clinic at Good Samaritan Hospital, where our Rheumatology fellows and some faculty saw patients. Although our patients received excellent care, some of our doctors, nurses, and staff were spread a bit thin. The situation was not ideal.

Last year, we got the chance to consolidate the two clinics, and to redesign and enlarge the space we had here at Johns Hopkins Bayview. The only catch was, we had a very small window of opportunity – just a few months – to figure out how to do it, to reconfigure the space and to orchestrate the complicated move out of Good Sam and into Bayview, which took place during one very busy weekend. The renovation happened a few weeks later, during a record-breaking cold snap.

It's all done now, and what a difference: The space is airy, cheerful, and – well, it just flows better. There are three major reasons for this, and their names are Laura Hummers, M.D., Clinical Director; Shannon Bishop, Clinical Manager; and Deann Gavney, Assistant Administrator for Rheumatology.

"We integrated our practice into one cohesive space," says Hummers, "and doing that has also improved the cohesiveness of the Division." With the clinics integrated, work flow is streamlined, and the fellows and faculty precepting them have been given a gift, too: no more commuting. Plus, "there was a lot of feeling that the fellows' clinic was isolated." As Bishop puts it, "it was difficult to get synergy between our centers because of the distance between the two groups."



Three major reasons the renovated clinic is a success, from left: Deann Gavney, Laura Hummers, and Shannon Bishop. OPPOSITE PAGE: Samantha Hurst-McCarroll provides a cheerful greeting in the renovated reception area.

Flow is a huge issue in a busy clinic. Picture, if you will, a military operations hub in a World War II movie: A giant table, with little tanks and airplanes, and staff constantly moving them around to show who is deployed where; the flow is constant. Flow in a well-run clinic requires that same kind of precision, and it

starts even before the reception desk. Patients come in and register, and if they come with family members, they all need a place to sit. Rheumatology patients often need to get their blood drawn to monitor their disease activity, so most visits to the clinic involve several stops: the front desk, the waiting room, the exam room, and phlebotomy. People who are in wheelchairs need some extra space for getting around; if they want to get a drink of water, the water fountain needs to be accessible, too.

Although the basic architectural footprint is the same, the space, which used to be shared with the Department of Medicine, is all ours now, with 16 exam rooms instead of 10, all freshly renovated. In combining the clinics, “our patient volume increased by 30 percent,” says Gavney. The waiting room, never big enough and always crowded, seemed tiny with the increase in patients; sometimes patients had to stand in the hallway because there was no other place to wait. The phlebotomy station, which used to be in the middle of the exam room space – meaning that “patients had to leave the exam room, check out at the front desk, get their orders and go back again,” says Hummers – needed to move and expand, too.

Not a Leisurely Move

“We found out that we would be moving the practice in April of 2014, and that we could need to move by November,” says Bishop. Not only was this a very small window, “this is nothing we’ve ever done before.

We were figuring it out as we went along.” First, there was a budget approval process, which involved getting bids for various jobs, and then many meetings with archi-

tects and administrators. “Somehow we were able to rush it through,” Bishop adds. “Quick turnaround time in an institution as big as Hopkins usually doesn’t happen; somebody was watching over us!”

Once the financial approvals came in, the team figured out something else: “Hopkins doesn’t provide an interior designer,” says Bishop. “None of us had experience in interior design.” The team added picking out textiles to the to-do list, and just kept going. “Originally, we thought we could do the renovation in piecemeal sections, but we couldn’t, so we were working against the time crunch to figure out how we were going to manage patient flow without closing the clinic down.”

The big move of the Good Sam clinic happened on a Saturday. “The day before, one of the elevators wasn’t working,” says Gavney, who was very pregnant at the time. “When we showed up that morning, it was up and running and we felt very grateful – but instead, the plumbing wasn’t working.” It took 12 hours, and that didn’t include unpacking and organizing. Monday was the start of a big yearly rheumatology conference, so “luckily, we had a few days. On Monday morning, all the clinical staff came together to get the clinic cleaned up and back in order for patient care later in the week.” The transition was seamless, says Bishop, “I think everybody was happy to be in one place.”

The renovation of the waiting room and phlebotomy areas happened a few weeks later. The registration desk moved temporarily to the glassed-in atrium just outside the front door. Says Gavney, “We did our best to troubleshoot in advance.” They did not anticipate a record deep freeze.

“It was incredibly cold outside, the coldest February on record,” says Hummers. The atrium is not heated. “The average temperature in there on one of the days was

45 degrees,” says Bishop. “We had brought blankets and space heaters and gloves, trying to keep everybody warm and get the patients in as soon as possible, because every time the doors opened, there was another gust of wind. It was pretty bone-chilling.” Helping to keep the clinic going despite the difficulties was “probably one of the most challenging things I’ve ever had to do in my career, because I felt like so much was at stake. There were so many patients scheduled to come in, we didn’t want to fail them or our staff.”

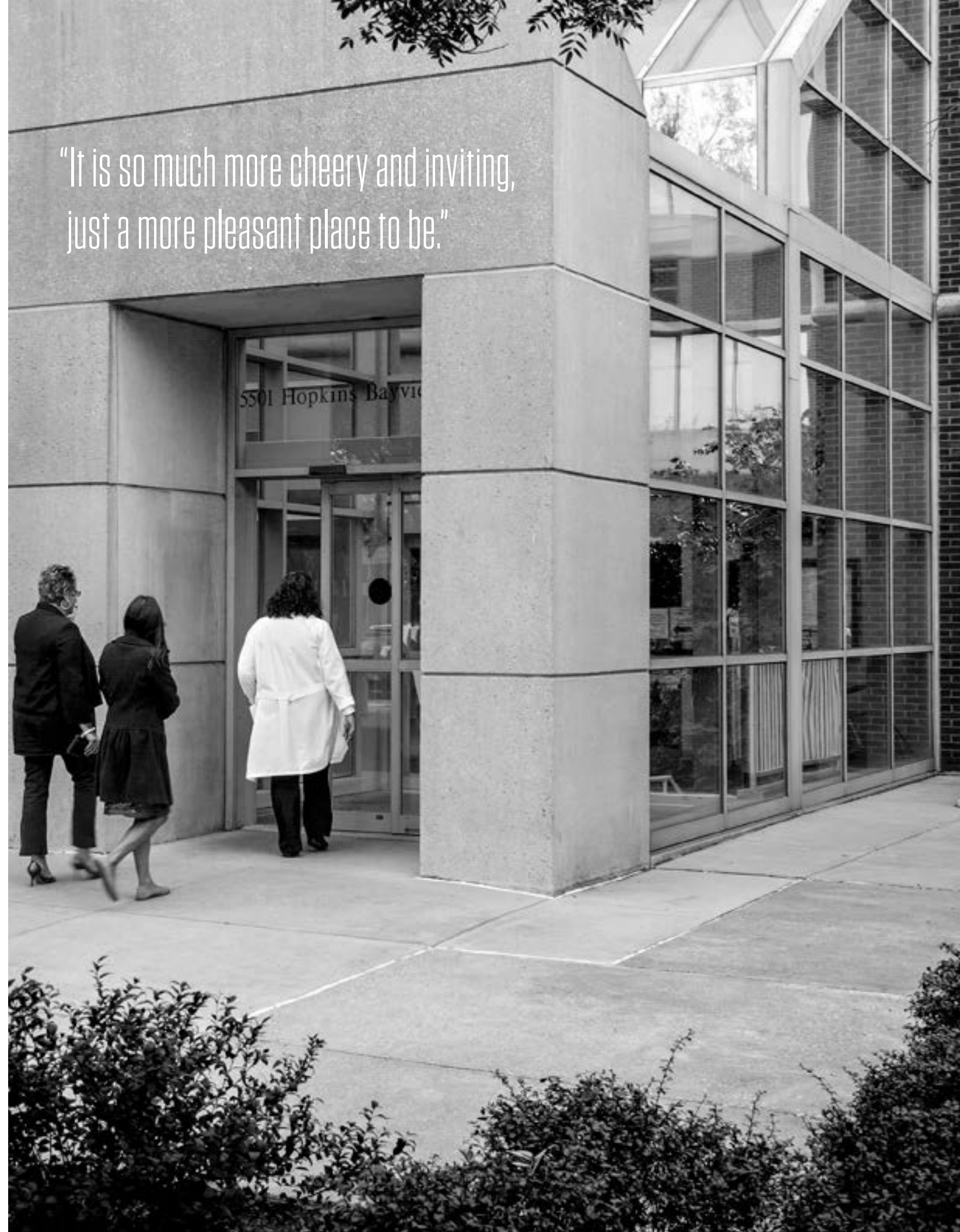
Adds Hummers: “Our staff went above and beyond to make sure the transition went as well as it could. A major stress like this could really bring down a group, or it could get everybody to work together as a team, and that’s what we did.”

The renovation took almost four weeks. On February 28, “we were supposed to move everything back in after a final inspection,” says Bishop. “But there was a snowstorm, so the inspector couldn’t make it.” He managed to get there by the end of the day, approved it, and the clinic was officially finished. “It looks radically different,” Bishop says. “It’s more welcoming. Patients really seem to like it.”

Gavney, who was out on maternity leave during the renovation, says she almost didn’t recognize the clinic when she came back. “When I walked in, I did a double-take. It was so dramatically different. It’s such a transformation; it went from being not very bright and welcoming, outdated, to being much more open and modern, and patient-centered.”

Hummers says that when returning patients see the new space for the first time, “they tell me they were convinced they were in the wrong place. They say that it is so much more cheery and inviting – just a more pleasant place to be.” L

“It is so much more cheery and inviting, just a more pleasant place to be.”





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"...there's got to be a **leap** of faith.
Ultimately, when you're at the edge,
you have to go forward or backward;
if you go forward, you have to jump together."

— Yo-Yo Ma