

THE JOHNS HOPKINS Vasculitis Center



INNER VOICES

BY JOHN STONE, MD, MPH

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Eight years after hanging up with the transferring doctor, I still remember my thought as the receiver clicked down: “*This is going to be complicated*”. Felicia, a 26 year-old woman employed as a clerk at the city jail, had lupus. Diagnosed several months before in some other place, Felicia had “fallen through the cracks”, coming to rest that Friday afternoon at Johns Hopkins. Details of her medical history, though sketchy, were worrisome: low platelets, falling hematocrit, kidney disease, and an altered mental status. We anticipated her arrival around midnight.

Felicia had been admitted to an outside hospital a couple of weeks before with fevers. Her platelets – the elements of the blood that help it clot – dwindled and then plunged steeply, refractory to transfusions. Her hematocrit, a measure of the number of oxygen-carrying red blood cells, also fell. The clinical picture quickly became muddled – was it lupus? Lupus involving the brain? Lupus associated with vasculitis of the central nervous system? An infection? Or something else? Her records also men-

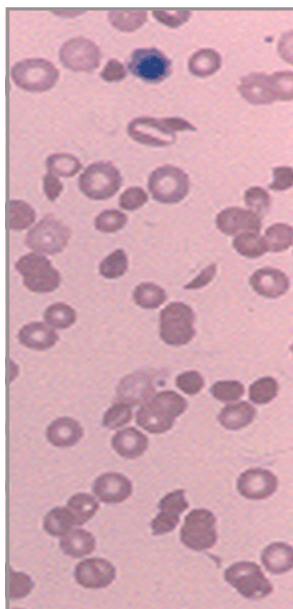
tioned symptoms of schizophrenia; but again, no details. This was going to be complicated, indeed.

When I met Felicia for the first time that Saturday morning, her hair was unkempt and matted, the edges of her mouth unclean with dried spit, her teeth not brushed, her gown in disarray – and her mind apparently oblivious to it all.

Though she was, in medical parlance, “oriented to person, place and time”, her thought processes were profoundly disturbed. Felicia mostly peered straight ahead with a paranoid expression, afraid to look to either side. She had trouble getting the words to begin. Her answers were short and uninformative. Building a therapeutic alliance with her seemed at tall order.

Beyond the medical challenges posed by her medical condition, there were other daunting barriers to trusting each other. To outward appearances, Felicia and I could hardly have been more different in our backgrounds, our cultures, our educations; and, of course, our personal health. Recognizing her altered state of mind,

I wondered just how these first superficial differences might filter through her thoughts. What comfort or, more likely, fear did she draw from encountering this man in a white coat who now seemed to be in charge?

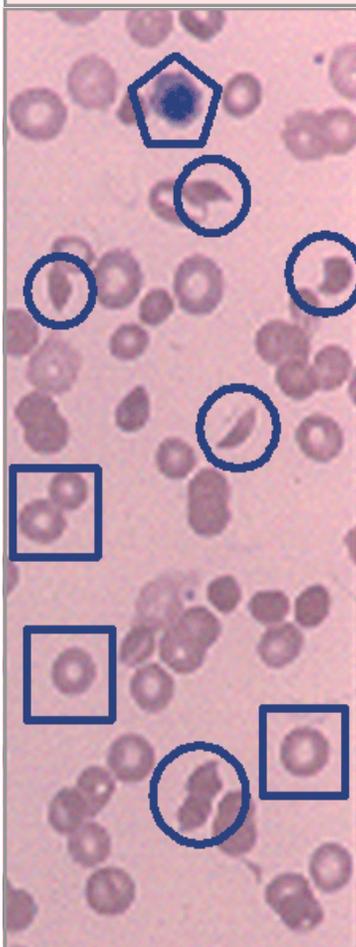


What clues to Felicia’s diagnosis are shown here?

The JHVC Newsletter for 2005 is generously sponsored by an educational grant from Mr. Christos Lambrakis.

INNER VOICES (CONTINUED)

Observe the broken red blood cells on Felicia's blood smear (circled below). These cell fragments were once healthy cells, sliced into pieces by microscopic protein threads that developed in Felicia's blood vessels.



Legend

- - Schistocytes or "split cells"
- - Normal red blood cell
- ◡ - Lymphocyte

If her mental status were not disturbing enough, the results of bloodwork looked even worse. When a patient's platelet count plummets below 20,000 platelets per cubic millimeter of blood (the normal volume ranges from 150,000 to 400,000), one worries about the possibility of a stroke from spontaneous hemorrhage into the brain. Felicia's daily platelet count, charted from data obtained at the outside hospital, was sinking fast: 132,000/mm³... 125,000... 96,000... 39,000... 22,000. Her hematocrit, supported only by transfusions of packed red blood cells, showed a similarly dismal trajectory. We reviewed her peripheral blood smear (Image: cover and same image with detail to the left) under the microscope: a drop of blood spread thinly over a glass slide with a coverslip. At low power, rather than the geometrically pleasing, rounded red blood cells – the normal "bi-concave discs with central pallor" (Left: "donut-shaped" cells) – we found red cells that seemed to have been blenderized: bizarre, irregularly-shaped cells and fragments of cells littered the slide (Left: circled cells). The blood smear told the tale: Felicia had thrombotic thrombocytopenic purpura, better known by sleek but sinister initials, TTP.

One of humankind's more dramatic and frightening illnesses, TTP is a disease of the microcirculation. In medical school, as I struggled to commit to memory the causes of anemia, the mechanism by which TTP destroys red blood cells was unforgettable. Within the small blood vessels of patients with TTP, strands of fibrin (a protein involved in blood clotting) form in the circulation, attaching themselves across the walls of blood vessels. These fibrin strands slice and dice the red blood cells, giving them the appearance of dinner plates shattered on the floor. This red cell shattering con-

verts healthy, oxygen-carrying cells to "schistocytes" (the root, *skhistos*, means "to split" in Greek. The same root, incidentally, forms part of the word schizophrenia, or "split mind"). The consequences of TTP are disastrous and swift: kidney failure, stroke, death.

Though patients with TTP once died in a fury, today TTP is treatable if diagnosed in a timely manner. The therapy – a process known as "plasmapheresis" – resembles hemodialysis. It works, in essence, by siphoning off factors in the blood that are causing harm (some of these are now known to be antibodies), and simultaneously by replacing deficient factors that are essential to the normal flow of blood through small blood vessels. Plasmapheresis requires the insertion of a large catheter into a patient's vein; usually, for the sake of access to a large vessel, one in the neck. As with many desperate therapies that somehow worked, the fact that plasmapheresis was effective in TTP was appreciated long before its theoretical mechanism was understood. In desperate efforts to combat the lupus flare that had probably triggered TTP, we gave Felicia more steroids, piling them on to those she had already received. Additional 1 gram pulses of methylprednisolone were mounted upon the steroids she had already received. And after a long conversation with her family about the urgent need for plasmapheresis, the surgeons inserted a large-bore catheter in her neck so that we could begin treatment. The site at which her line was inserted oozed blood all night, symptomatic of her low platelet count and underscoring just how critically ill Felicia was.

Over the ensuing days, Felicia's mental status declined even further. At first she was just less responsive – somnolent, but still following commands. Later, she struggled mightily in bed, calling for her

INNER VOICES (CONTINUED)

father who had died some years before, and for her Bible. She tried to pull out her plasmapheresis line, and to bite anyone attempting to touch her. Examining her for signs of bleeding, changing her catheter dressing, and drawing blood to monitor treatment became gut-wrenching ordeals. Felicia's nurses, interns, and residents prepared for them like a SWAT team. She had delusions of nosebleeds, and often cried hysterically "something is on my face". She sometimes burst out in salvos of gospel-like singing. An intern wrote in the daily progress notes that he had opened her door to find the patient "singing like a banshee in her room". She required a sitter in the room at all times to protect her from herself. Periods of outbursts required heavy sedation. And over her hospital gown, around her torso and tied in the back, was a restraining vest with heavy cloth ties, for use when all else failed. To help keep her calm, the lights in her room were kept low, the door closed.

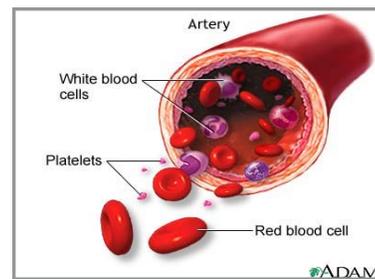
After about ten days of plasmapheresis and prednisone, her blood profile improved. Her platelet count stabilized and began to inch its way up. Her need for packed red cell transfusions diminished. Oddly, though, as we seemed to gain the upper hand on some aspects of her TTP, her mental state grew worse. She began to hear voices. The nurses were the first to pick up on this. They noted that Felicia seldom interacted at all, unless belligerent. During long plasmapheresis sessions, though, she occasionally responded fearfully to input from someone beyond the room. "No!", she would mutter, and without provocation would repeat a louder: "NO!". Pressed for details about these voices – "*Are they frightening to you?*" (Pause. No response.) "*Are they telling you to harm yourself?*" – Felicia usu-

ally looked confused and grew quiet. But eventually she acknowledged that the voices were there. We interpreted this new development in Felicia's care as the emergence of schizophrenic symptoms, and were greatly disheartened.

Generally, I didn't see Felicia by myself. Whenever I was in her room, I was part of a team of caregivers: on rounds, with medical students, social workers, interns, residents; talking with consultants on Felicia's case in low tones; standing at the foot of her bed near the plasmapheresis nurse. Never alone. In part, this was due to the intensive care of a sick patient in a busy hospital. Whenever I was visiting, there was always someone else in the room, too; the nurse, the sitter, the cafeteria worker picking up Felicia's tray, the maintenance staff. But, I knew it was also due to avoidance behavior – on my part. Felicia's violent outbursts had decreased as her blood counts had risen and the round-the-clock sitter had been dismissed, but her capacity for violence, for lashing out, biting, and attacking still seemed present and frightening. Safety in numbers was reassuring.

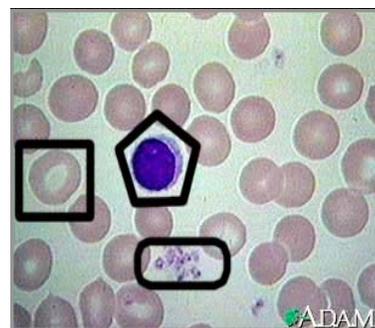
One Saturday afternoon when the hospital was quiet and the house staff had already rounded, I entered Felicia's room alone. I paused at the door, peering through the dimness to see if she was awake. She was. I greeted her softly and smiled, but realized that she probably had trouble seeing my face, backlit by the bright yellow hall lights. Closing the door but leaving the room lights low, I slowly approached her bed. She lay as she had lain for three hospital weeks. I wondered if she had really interacted with a single human being all day.

When talking with patients, doctors-in-



Blood vessel, Vasculitis and TTP:

In vasculitis, inflammation occurs within the wall of the blood vessel, ultimately leading to narrowing of the vessel and insufficient blood supply to tissues. With TTP, in contrast, destruction of red blood cells occurs in the lumen (open tubular space) of the vessel.



(Above) Image of normal blood cells. Notice the "donut" shape of the healthy red blood cells. Also shown, a lymphocyte (the large purple cell) and a platelet.

Legend

-  - Normal red blood cell
-  - Lymphocyte
-  - Platelet

INNER VOICES (CONCLUSION)

training are taught to come close. *“Take your hand off the doorknob... Go in and sit down... Sit at eye level... Touch the patient in a non-threatening way... Hold the patient’s hand”.*

Judging Felicia to be at least relatively calm, I eased my way down and sat on the foot of her bed. My extended hand, resting on her knee through the blankets, was still far enough away for me to pluck and remove it to safety if necessary. Felicia had changed little since her first hospital day. Because of her combativeness, baths from the nursing staff had been few. Her hair remained unkempt, her teeth unbrushed. In response to my warm-up questions, designed to soothe, she uttered only a few unintelligible words. *“Pardon me?”*, I said gently, and leaned closer. There were long pauses in our conversation as I searched for what to say next. Was there any way of getting through to her? Was a human communication possible today? Possibly not. If not, how to extricate myself gracefully, without making her feel deserted or alarmed? The seconds ticked by,

and turned to minutes. I fell back on summarizing for her the progress we had made. *“Your platelet count ... Have you heard the doctors mention that term?... is better. You are still in the hospital and have been very sick, but we have been making some important strides...”*

As I rambled, Felicia stirred and struggled to sit up in bed. Her movement was restricted partly by her tangled IV, her central line, the soft eggcrate mattress, the straps of her restraint vest, and her own weakness. Only then did I think to notice the condition of her restraints: untied. Uncertain of her intentions and mindful of her past attempts to bite, my own inner voices awoke. *“Leave. Just say ‘Good-bye’ and step out before she gets close”.* I was frozen. Slowly she rose from the pillow, staring wildly and grimacing at the effort to hoist herself by the bedrail. The thick wooden door was closed firmly. Even if it had been open, we were at the end of the hall and hardly within earshot of the nurse’s station. The next level of anxiety arose in me, *“Get off the bed! Call for help! Shout! Get OUT of here!”* But something held me there as she drew nearer. She mumbled as she rose. With great effort, she slowly reached out and then lurched toward my side, bringing her head uncomfortably close to my ear. Through a tangle of sheets, blankets, hospital gowns, and an IV, I realized that she was hugging me. Startled, relieved, I hugged her back, welcoming her warm tears on my cheek and white coat.

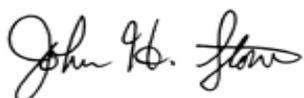
Epilogue:

“It’s the steroids”, pronounced the psychiatrist, whom we had asked to consult on Felicia’s mental health and auditory hallucinations. The prednisone, the psychiatrist maintained, had caused Felicia’s symptoms of psychosis – not the TTP, not the lupus, and definitely not schizophrenia. *“Taper the steroids if you can”*. At the time, I was a junior attending physician, more attuned to the manifestations of disease than to the complications of treatment. Belief in the psychiatrist’s opinion came reluctantly for me. But in the end, thankfully, the psychiatrist was right. We tapered Felicia’s prednisone. Her platelet count, hematocrit, and kidney function remained stable. And, her inner voices fell quiet.

Eight years later, Felicia still works at the city jail. I’ve gotten to know her much better than the chaotic circumstances of her hospitalization allowed. We marvel together at her transformation from those days. She doesn’t remember the inner voices, but believes me when I say she heard them. Her “voices” have not returned, but some of mine have remained. They are not voices of alarm, but rather reminders of the lessons I learned from those weeks of being Felicia’s attending physician. First, some desperately sick patients do recover dramatically and return to full lives, even from dreadful diseases that we don’t entirely understand. Second, never underestimate that a treatment designed to ameliorate may actually complicate the clinical picture. And finally, for patients who are critically ill, the power of human contact – of talking, touching, and yes, hugging – is essential to recovery.

Thrombotic Thrombocytopenic purpura

- Disease in which red blood cells are destroyed within small blood vessels
- Often confused with vasculitis because of its multi-organ system involvement
- Five cardinal features:
 1. Fever
 2. Anemia
 3. Low platelet count
 4. Kidney dysfunction
 5. Altered mental state



KNOW YOUR MEDICATIONS:

PREDNISONE IN TREATING VASCULITIS

BY MATTHEW MARRIOTT, PA AND PHILIP SEO, MD

Prednisone (glucocorticosteroid or “steroids”) is a man-made version of cortisol, a hormone the adrenal gland produces. For a person in good health, approximately 8mg cortisol is released in the body daily in the early morning. That small amount of cortisol impacts everything from mood to the beating of your heart.

Man-made cortisol, better known as prednisone, was discovered in the late 1940’s as a treatment for rheumatoid arthritis. At the time, prednisone was thought to be a miracle drug, but as time has told, it has had its downsides. The mega-doses used in its early days were found to have many undesirable short-term and long-term side effects limiting the use of the medication. Prednisone did not become a commonly used medication until the 1950’s as appropriate dosing was better understood. Until the availability of prednisone, there were essentially no effective therapies for most forms of vasculitis and patients were entirely at the mercy of the disease.

Prednisone works with many different autoimmune diseases because it broadly suppresses the body’s natural immune response. Its effectiveness in curbing the hyperactive immune system of vasculitis patients is why prednisone is a “cornerstone” medication in a vasculitis treatment plan.

Typically, prednisone is used as the first treatment in vasculitis. Often times, it is issued as a large I.V. (intravenous) doses over a few days in the hospital, followed by oral prednisone (inset: right) as continued treatment. Slower acting vasculitides or flare-ups caught early may be treated with oral prednisone and often is combined with an immunosuppressive agent like cyclophosphamide, meth-

otrexate or azathiaprine. Various doses are available for oral prednisone: 1 mg, 2.5 mg, 5 mg, 10, mg, and 20 mg tablets.

Commonly Asked Questions

Q: Why do I need to take prednisone?

A: Prednisone is the fastest acting, most effective medication there is to treat vasculitis.

Q: How long will I be on prednisone?

A: Most of our patients will need at least 6 months of prednisone. Some patients may be able to stop earlier, and some will have to continue on low doses for longer periods.

Q: How soon will I notice a side effect from prednisone?

A: Not all people will experience side effects, though the higher the dose, the sooner a patient may experience them (within a few weeks). Most commonly expressed are weight gain, stomach upset, and difficulty sleeping. As the prednisone is lowered, the side effects tend to be less noticeable. Many side effects resolve once the daily dose goes below 10mg daily. The JHVC has a goal of getting our patients off of prednisone as soon as is safely possible (as short as 6 months in many cases).

Q: What can be done about the side effects?

A: Short-term use of additional medications may make a difference. Medications for blood sugar management, sleep aids, mild anti-depressants and other counter-reactives can be used to help manage side effects. These additional medications can be tapered as the prednisone is tapered, based on instruction from the prescribing clinician.

Prednisone (shown below) in it’s most commonly used form for treatment in vasculitis.



Prednisone’s Possible Side Effects

- Fluid retention (puffiness in hands, feet, ankles and legs)
- Weight gain
- Puffiness around the neck, face, or upper back
- High blood sugar (type II diabetes)
- Skin changes (bruising, thinner skin, delayed wound healing)
- Stomach upset
- Mood swings (noticeable depression, anxiety and/or euphoria)
- Sleep disorders (insomnia, night sweats)
- Susceptibility to infection
- Higher blood pressures
- Blurry vision
- Increased eye pressure (glaucoma)

Rare Side Effects

- Auditory hallucinations
- Severe depression or psychotic episodes (potential harm to oneself or others)

ASK THE JHVC:

ABOUT PREDNISONE—WHAT IS HAPPENING TO ME?

BY STACEY LABAHN, MIM



Cushing's syndrome

(hypercortisolism) occurs when a person's tissues are exposed to an excess of the hormone cortisol or its derivatives, such as taking high-dose prednisone.

What are its symptoms?

- Extreme weight gain
- Moon face (a)
- Excess hair growth (b)
- Acne (c)
- Buffalo hump (d)
- Reddish-blue streaks on the skin (striae/stretch marks) (e)
- High blood pressure
- Tiredness and weakness
- Muscle and bone weakness
- Moodiness, irritability, or depression
- Sleep disturbances
- High blood sugar
- Menstrual disruption in women
- Decreased fertility in men

For many of us, the hard, true reality about prednisone is we must take it, side effects or not. If not taken when prescribed, our health would be quickly compromised by vasculitis.

This article, paired with “Know Your Medications” on page 5, is the first in a two-part series on prednisone. As a patient, knowing the side effects of your medications will help you distinguish between the impacts of disease and the effects of the medication.

Our objective is to help patients be informed participants in their care. The challenge with information articles, though, is one person's acceptable and desired amount of information may be overwhelming, scary, or discouraging to another person. Only you can determine how much information you want or need to know.

What is happening to me?

If you've been diagnosed with vasculitis, odds are you've taken prednisone (glucocorticosteroids) at least once during treatment. As seven year steroid veteran who takes between 15-60mgs daily, I think of it as “the medication we love to hate”. We love it for its effectiveness and life-saving properties, but we hate it for what it does to our physical and emotional well-being. As uncomfortable, disruptive, and even disturbing as prednisone's side effects can be, few can deny the relief that is experienced when as little as a single dose creates a marked and positive difference in our physical well being.

Often, prednisone is the first medication that successfully controls vasculitis. As wonderful as it is to have a medication that works, it creates interesting challenges for patients. When prednisone is effective, it gives us a glimpse that good health is possible again. It confirms we are armed with the right diagnosis and a medication that stops disease progression; so we are ready to start regaining our sense of health.

In the short time between feeling better and determining the impacts of vasculitis on our overall health, we live in fear that this miraculous drug will stop working. We pay attention to things that do not fit in “what is normal”. As the prednisone builds in our system, the side effects start to surface. Combine this with trying to deciphering “what is normal” after vasculitis, and it can be as tricky as figuring out “who dunnit” at the beginning of a murder mystery just as the characters are starting to introduce themselves.

Prednisone complicates matters for patients because of its side effects can mimic symptoms of various vasculitides. Is the muscle weakness and fatigue from the vasculitis or the natural effect of tapering off prednisone? What about the joint pain or “bone pain”, as some patient call the periodic aches in their joints? Muscle cramps? Is the pain round the stomach and chest indigestion from medications or worse? It leaves patients wondering what to note, when to be concerned or not concerned, when to contact the doctor, much less which doctor!

Deciphering the Effects of Prednisone

Many times, these questions can only be answered through experience. By taking and tapering prednisone, you'll start to notice how it effects your vasculitis symptoms. But, additional facts about prednisone will help.

As stated in *Know Your Medications* (page 5), prednisone suppresses the body's natural immune reactions. With vasculitis, the immune reaction to inflammation is one of the culprits of the disease, and prednisone provides an abundance of anti-inflammatory hormone (cortisol). While stopping the progression of vasculitis, it interrupts normal hormone activity in the adrenal gland. Prednisone cre-

ASK THE JHVC (CONCLUSION)

JHVC FACULTY
AND STAFF

ates a state of imbalance resulting in the body reacting as if it has Type II diabetes (inset: below) and Cushing's syndrome (inset: left). The symptoms list for these diseases and the side effects list for prednisone almost mirror each other. It's important to understand that these diseases are not permanent but a reaction to excessive cortisone in your body. Once the cortisone is tapered or stopped, natural adrenal functions will resume in time (which is why tapering is important!). While experiencing prednisone's side effects, knowing how these two diseases are managed can help.

Even though the diabetes and Cushing's syndrome are not permanent, understanding that your body is acting like it has these diseases helps you understand the "mimics", which will help you separate what is prednisone side effects verse vasculitis symptoms. It also offers further information and personal research options to help you understand what you can do about the side effects. By reading about these diseases, you can learn what people with these diseases do to alleviate their symptoms. Often, there are medications that your clinician can prescribe to help counteract some side effects.

Diabetes means that your blood glucose (often called blood sugar) is too high.

What are Type II diabetes symptoms?

- Urinating often
- Feeling very hungry or tired
- losing weight without trying
- Sores that heal slowly
- Dry, itchy skin
- Losing the feeling in your feet or having tingling in your feet
- blurry eyesight

The challenge: Depending on which type of vasculitis you have, each of these symptoms can easily be confused with vasculitis flare-up by patients.

**Dr. Harvey Cushing (1869-1938)**

was the first Chief of Neurosurgery at Johns Hopkins, where he taught from 1900-1912. At

Hopkins, he came under the influence of several famous physicians: the pathologist William Welch; surgeons Howard Kelly and William Halsted; and Sir William Osler, the first Chairman of Medicine at Johns Hopkins.

Early in his career, he designed the first approach to the surgical removal of the pituitary gland, the source of the compound that triggers the body to produce cortisol (as steroid hormone akin to prednisone). Cushing also gained recognition by publishing a landmark textbook, The Pituitary Body and its Disorders. **Cushing's syndrome** is a condition in which the body responds to excessive amounts of cortisol (or, more commonly today, to treatment with prednisone) by the development of characteristic physical changes, described on the left inset of page 6.

Cushing had many other talents outside of medicine, as an athlete, artist, author, and bibliophile. While a surgical resident in Boston, he frequently demonstrated his athletic prowess by performing back flips off the front porch of the Peter Bent Brigham Hospital. He served as a field surgeon with American forces in the First World War, and ironically assisted in the battlefield operation on Osler's son, Revere, who was mortally wounded in France. Cushing's book entitled The Life of Sir William Osler won the Pulitzer Prize in 1926.

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PATIENT PERSPECTIVES: A TRAVELOGUE OF A CLINICAL TRIALS PARTICIPANT

BY JOHN AND RACHEL BROWNLEE

Journeys, like artists, are born and not made. A thousand differing circumstances contribute to them, few of them willed or determined by the will -- whatever we may think.

Lawrence Durrell
Writer/Teacher
1912-1990

My daughter, Rachel, is a patient in the RAVE study (officially known as the Rituximab for ANCA-Associated Vasculitis-ITN021AI). She is an active 17 year-old who was diagnosed with Wegener's Granulomatosis (WG) in 2004. Initial attempts to control her disease seemed hopeful, but that changed when we recently found she had disease progression in her lungs and continuing sinus symptoms. Her lack of remission prompted Rachel and I (her father) to seek a change of treatment. Upon researching, it meant adding cyclophosphamide (cytoxan) to her medications.

Because of her age, I was concerned about the side effects long-term use of cyclophosphamide caused, and sought alternatives. So, the researching continued.

While reading treatment alternatives in medical papers and journals, we found a

"vasculitis buzz" about rituximab, an FDA approved drug for non-Hodgkins lymphoma that depletes a patient's B cells for 6-12 months or even longer. (B cells are the type of cell that transform into cells that make ANCA). The articles I read noted that after pilot clinical trials with Wegener's patients, rituximab had shown promise of decreasing the disease activity and maybe allowing the Wegener's to go into true remission. As promising as this new drug sounded, I am a conservative soul. For me, "new" often equates with hype and fuzzy evidence. So we proceeded with caution.

The RAVE study was attractive because it had a built in safety net: either Rachel

would get the best standard therapy for her Wegener's or she would get the new therapy at the start of the trial. If the treatment to which she was randomized initially was not effective, she would be switched to the other treatment. Once we accepted the protocol or "practice methods" of the study, the logistical challenges came next.

We live in San Antonio, Texas. The nearest research center for RAVE is in Birmingham, Alabama, but the easiest center to access is in Baltimore thanks to non-stop flights offered by Southwest Airlines (SWA). The airline schedule was convenient for our needs, but changing our flight schedule to accommodate possible reactions to medications or other unforeseen problems would be costly. I wrote to Southwest. The Customer Relations Department was very compassionate to our situation. Once Rachel's illness was confirmed by the Vasculitis Center staff, to our surprise and gratitude Southwest Airlines donated four roundtrip tickets and a \$500 flight credit to accommodate schedule changes.

I also found a group called Angel Flights, an organization of volunteer pilots who donate their time and planes to transport individuals and families with medical needs to the hospitals and clinics where care will be provided. Because of the generosity of Southwest Airlines we did not have to rely on An-



Rachel in her RAVE hat— As a participant in the RAVE trial, Rachel is required to cover her hair when she visits the JHVC. The reason? One of the medications used in the trial can cause a specific type of hair loss and the investigating physician is not supposed to know which patients are taking which medications. To prevent "doctor deduction" from occurring, its "hats on!" rather than "hats off!"

Can we be of service?

Please email us at JHVC@jhmi.edu or send your comments to:

The Johns Hopkins Vasculitis Center
c/o Stacey LaBahn
5501 Hopkins Bayview Circle
JHAAC, Rm. 1B.1A
Baltimore, MD 21224

PATIENT PERSPECTIVES (CONCLUSION)

gel Flights, but it is great to know of the existence of this program.

So, RAVE study, Baltimore, and the Vasculitis Center for treatment. Next was the reality of traveling to Baltimore, renting a car and staying in a hotel, in other words: expenses. It looked painful but necessary. A friend who heard of my daughter's illness asked if there was anything other than prayer that she could do to help. I explained the lodging issue. Since Rachel is under the age of 21, our friend suggested we try Hopkins Children's House or Ronald MacDonald House in Baltimore. The Children's House had no room, but the Ronald MacDonald House had availability and welcomed us.

We were pleased to discover that Ronald MacDonald is located in a new section of a very old street in Baltimore. It has great security, good lighting, a guarded parking lot, and is a spacious, cheery house. The staff are kind, helpful, and upbeat. The sleeping rooms are typical of those of a hotel, but the House provides common living areas and kitchens that give plenty of room to gather large groups. Sometimes the Ronald MacDonald House hosts family-style meals prepared by local volunteers. The House also has private space set aside for moments of reflection. These are often helpful to families dealing with challenging medical conditions. There are also many recreation rooms and a computer room for adults wishing to access information on the internet or e-mail friends.

Rachel and I ate many meals there, listened to a youth jazz ensemble, helped make a scrap book with the other patients, went to the gym, and attended an Orioles game, all courtesy of donations given in time or subsidy to the Ronald McDonald House from the local

community. All this is provided for \$15 a night, a goodwill offering of cleaning up after oneself, and helping with a few communal tasks.

Rachel has gone through her induction phase of the study and all looks hopeful for a good response to treatment. Our Baltimore experience was healing in many aspects because of the wonderful staff at The Johns Hopkins Vasculitis Center, The Ronald McDonald House and SouthWest Airlines.

May God bless!



Rachel in early 2005, two months before starting the RAVE trial



Beauty and the Beast, 2005 - Rachel, an aspiring artist, painted her sister and the family dog for a summer art class.

ALL IN THE FAMILY: A GENETICS STUDY IN SYSTEMIC VASCULITIS

BY KAREN GAUSS, RN, MLA AND KATHLEEN BARNES, PHD



DNA Collection Kit for Family members: The Oragene DNA Self-collection Kit is mailed along with 2 postage-paid shipping envelopes, instructions on how to use the kit, and a plastic canister containing the holder for a saliva sample (shown to the right of the canister).



DNA Collection Kit for the person with vasculitis: Blood specimens for DNA are needed from the family member with vasculitis. It can be collected locally or during a visit to the JHVC. If local, the sample collection kit will be mailed to your home. Take this kit with you the next time you visit your local doctor or lab for blood work. They will mail the samples back to the Hopkins Genetics lab.

One of the questions most commonly asked by our patients is, “If I have vasculitis, will my children get it?” Thanks to a private grant from the Lowe family, the JHVC is addressing this question. Determining how and why someone contracts an illness is critical to identifying cures for those with the disease and discovering preventative medicine for those susceptible. This is a very exciting time in vasculitis research, and the JHVC is launching a critical study using advanced genetic research techniques to determine if and what genetic risk factors are associated with vasculitis.

The Family Genetics of Systemic Vasculitis Study (FGSV) at The Johns Hopkins Vasculitis Center, gathers DNA of biologically-connected family members in hopes of finding similarities or differences that help us find indicators for vasculitis. In the FGSV, we are collecting samples from families because DNA coming from the same sources (a mother and father) make seeking genetic uniqueness easier to identify. Our researchers will attempt to identify genetic maps or “linkage” maps by looking

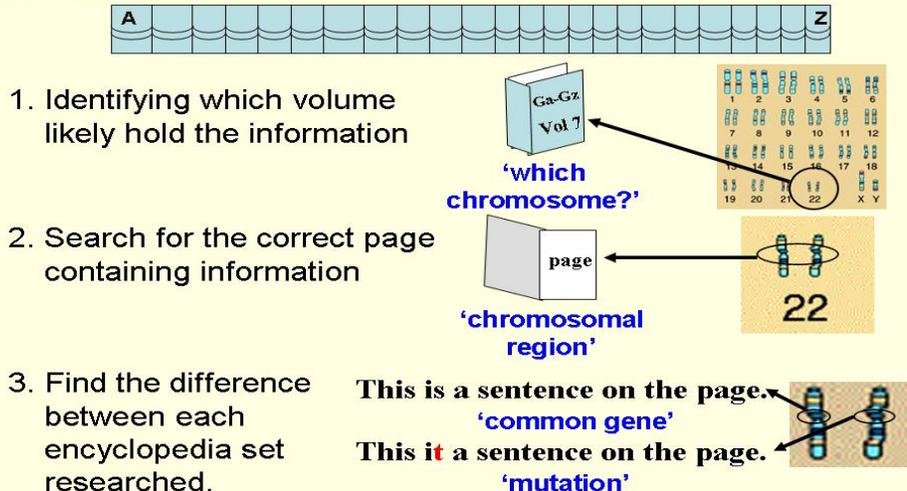
across at large numbers of participants, both with and without vasculitis, that have the similarity of having a person with vasculitis in their direct genetic lineage. Any markers found consistently in the research pool will help support identification and understanding the role of genetics in vasculitis. If a genetic connection can be made, researchers can use this information to further determine what makes the identified part of the gene react in vasculitis.

For a family to qualify for the FGSV Study, the person with vasculitis needs to have two living biological parents or one living biological parent and two living, fully-related siblings. To collect the DNA, the person with vasculitis would provide a one-time blood sample collected either local to the participant or at the JHVC. Family members DNA is provided through saliva, which is gathered in a specially-designed collection container (image: left).

If you are interested in participating in this study, please contact us at 1-800-226-6023.

How we find variants, or genes, responsible for human disease

Find a disease gene is like finding a misspelled word in a set of encyclopedias



JHVC IN THE NEWS FALL 2005

Hellmann Appointed Vice Dean for Hopkins Bayview



David B. Hellmann, M.D., M.A.C.P., Co-founder and Executive Director of The Johns Hopkins Vasculitis Center, a nationally renowned rheumatologist and Chairman of the Department of Medicine at the Johns

Hopkins Bayview Medical Center, has been named the Hopkins School of Medicine's Vice Dean for the Hopkins Bayview Campus.

Since arriving at Bayview several years ago, Dr. Hellmann has played an important role in the dynamic development of the Bayview campus. He has recruited accomplished division directors and attracted other highly regarded individuals to join the medical center's departments.

Dr. Hellmann's patients can be assured he will continue his normal clinic schedule in conjunction with his duties as chairman of the Department of Medicine at Hopkins Bayview, and as the Mary Betty Stevens Professor of Rheumatology in the School of Medicine.

On Dr. Hellmann's appointment, Edward D. Miller, M.D., dean of the School and CEO of Johns Hopkins Medicine commented, "David Hellmann is committed to building upon Hopkins Bayview's strengths and developing programs there that reinforce the bonds between our two campuses. His appointment as vice dean reflects our commitment to the same goal."

Asked to comment on how he will handle his Vice Dean's duties in addition to his other responsibilities, Dr. Hellmann states: "I will stop taking the elevator and start taking the stairs!"

NEW WAYS TO REACH THE JHVC

BY JOHN STONE, MD, MPH AND STACEY LABAHN, MIM

Phone, fax, cell phone, email, instant messaging, text messaging,...as each year passes, new ways to reach people are developed. In the beginning, the new communication seems novel and exciting. Gradually, we accept the new mode as commonplace, as it fits into the many ways we can be reached during the day. Once the communication tool is "every day" for us, we are surprised to find someone who does not use it, and we do our part to educate them on the new communication mode's use and function.

Until recently, medicine has been like "the person who doesn't have an email address or have access to a computer." HIPAA regulations, insurance, computer access and system security have kept clinicians from being able to exploit the full power of the internet to communicate and care for their patients. All this is starting to change.

New web-based systems are becoming available that provide secured, HIPAA compliant communications. The systems are designed for patients to be able to send and receive non-emergency communications, house their basic health information, manage prescription refills, request appointment changes, get referrals or receive lab results. These systems can even do "web visits" where specific question and answer spaces are provided to the patient to gather information. Once complete, the responses are solidified and emailed to their care giver for reply. Sounds like medical science fiction, but these systems are in use currently and gaining sophistication all the time!

The Johns Hopkins Vasculitis Center is currently evaluating secured, internet patient communication centers. We hope to establish new ways of reaching us soon. If you have any questions, please email us at JHVC@jhmi.edu.



Patients using doctor provided web communications systems state greater satisfaction with their care. Some of the reasons stated in the survey were:

- **Efficiency**—It eliminates missed phone calls or "phone tag" where messages are left.
- **Timeliness**—In place of taking time off work, routine questions can be asked through the website and the clinician can provide instruction without an office visit.
- **Privacy**—The ability to receive information in print provides greater privacy. Phone calls, being spoken aloud, can be overheard.
- **Interconnections**—Some systems enable designated family members or care givers to communicate for the patient (such as young children or elderly parents). Records are linked so the clinician knows who to contact. Access for each person's records in the "interconnection" is controlled by the user.



CLINIC CORNER: INSURANCE HAND-OFF

For new patients or returning JHVC patients who have not been to clinic in the last six months: *Don't hang up when Sidone or Cynthia wishing you good day!*

With a goal to make insurance processing more effective, a new insurance verification hand-off has been added as the final step in scheduling a new or returning appointment. It's quick and easy! So, have your insurance card ready when you call for a JHVC appointment.

Appointment Process

1. A Patient Care Coordinator (Cynthia or Sidone) will work with

you to schedule a new or return appointment at our clinic.

2. Once the appointment is set with the JHVC, they will ask you to hold while they transfer you to Hopkins Central Scheduling.
3. A Central Scheduling Representative will ask for insurance information for new patients or verify insurance information on file for currently patients.

This final step confirms your appointment, and will be followed by a reminder letter from the JHVC.



<http://vasculits.med.jhu.edu>
We're on the web @

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The Johns Hopkins
Vasculitis Center

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