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## The Johns Hopkins Vasculitis Center: Origins

You may remember all the TV commercials during Michael Jordan's heyday that exhorted people to "be like Mike". Well, I never really wanted to "be like Mike", but I did want to be like Mary Betty – **Dr. Mary Betty Stevens**, that is. Dr. Stevens is the reason I wanted to become a rheumatologist, and she's the reason I became interested in vasculitis.

Dr. Stevens was the rare person who inspired everyone fortunate enough to meet her. Her patients adored her, her students were awed by her, and her colleagues admired her. A tall lady with a booming voice, Mary Betty ("Marty", to her friends) always made a dramatic first impression. As her patients and students quickly learned, her intellectual and humanistic qualities were even more impressive: her dedication, warm bedside manner, and vast knowledge of medicine made her one of Hopkins' finest physicians.

As a junior medical student I had the privilege of spending 4 weeks working side by side with Dr. Stevens and her staff. Before I started that rotation, I was uncertain what kind of doctor I wanted to be or what field I would pursue. But quickly I learned from Dr. Stevens the personal rewards of getting to



**Mary Betty Stevens, M.D.**

know patients and their families well. She also impressed upon me the need for research, so that the treatments of tomorrow would be better than those of today. Finally, I learned from Dr. Stevens that it takes a team to provide patients the best care, and that team includes the administrative staff, receptionist, nurse, physical therapist, and others.

Of the more than 100 diseases in rheumatology, Dr. Stevens was especially interested in two: vasculitis and lupus. Not surprisingly, I, too, became very interested in these. Over time, I saw that vasculitis had received

*(Continued on page 2)*

### Inside this Issue:

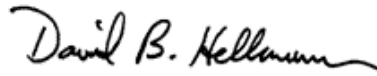
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less attention from other doctors. So in the mid-1980's I began concentrating my efforts on patients with vasculitis. I started several research studies and began recruiting others to the field. The big break came in 1997 when I was able to recruit **Dr. John Stone** back to Hopkins. John had been one of my residents some years earlier, so I knew well of his unusual talents. With John's energy and creativity, we officially established the Johns Hopkins Vasculitis Center in 1998. Thanks to the efforts of many, the Center has flourished.

Sadly, but perhaps fitting in some ways, Dr. Stevens died several years ago, the day after she retired.

But in a real sense, Dr. Stevens' spirit lives on. In 1995, I was honored to become the Mary Betty Stevens Professor of Medicine, an endowed Chair named after this great woman. Her spirit also lives on through all of the wonderful people working at the Vasculitis Center. In our conduct every day in trying to help patients with vasculitis, I encourage everyone associated with the Vasculitis Center to "be like Mary Betty", one of Hopkins' all-time greats.



*David B. Hellmann, MD  
Executive Director, The Johns  
Hopkins Vasculitis Center*

**Welcome  
Chasity Harrison!**



The Vasculitis Center is delighted to welcome **Chasity Harrison** as a full time clinic coordinator. Last month, Chasity recently graduated summa cum laude from Towson University with a double major in Speech Pathology/Audiology and Mass Communications.

**Vasculitis Center Patient Statistics**

The Johns Hopkins Vasculitis Center is one of few centers in the world committed exclusively to the care of patients with vasculitis. As a result, patients come to the Center from all over the world for consultation on their conditions and advice on treatment. The Center is also committed to finding better treatments for these diseases. Many patients come to our Center in Baltimore to participate in research studies of new treatments. Regardless of patients' reasons for coming to the Vasculitis Center, the doctors and staff of the Center are dedicated to diagnosing these diseases swiftly, treating them effectively, and discovering why they begin.

In the past 4 years, nearly **1,000 patients** have been evaluated by physicians at the Center. The table to the right specifies the 10 diseases most commonly evaluated in our Center. Wegener's granulomatosis and giant cell arteritis are the types of vasculitis seen most frequently (23% and 15% of the total, respectively). In all, however, nearly 20 different diagnoses comprise the group of diseases known as "vasculitis". Many of the more rare forms of this disease are included in the "Other" category in the table. This category includes Buerger's disease,

cutaneous leukocytoclastic vasculitis, Henoch-Schönlein purpura, Cogan's syndrome, urticarial vasculitis, and others.

<b>Disease Type</b>	<b>Percentage of Total</b>
Wegener's granulomatosis	23%
Giant cell arteritis	15%
Polyarteritis nodosa	15%
Central nervous system vasculitis	7%
Takayasu's arteritis	6%
Behcet's syndrome	6%
Cryoglobulinemia	3%
Polymyalgia rheumatica	3%
Churg-Strauss syndrome	3%
Microscopic polyangiitis	2%
Other	19%

*Table 1. The 10 diseases seen most often at the Vasculitis.*

## Vasculitis and the Dickersons: A Family Affair

In November of 1993, when **Todd Dickerson** was 20, he developed symptoms of an upper respiratory infection, fevers, aches, and began to cough up blood. He quickly found himself in a hospital in Delaware, initially diagnosed as having pneumonia. Within days, he had bled extensively into his lungs and was found to have a blood clot in his leg. He was transferred to the Intensive Care Unit at Johns Hopkins, where he first met Dr. David Hellmann. Dr. Hellmann immediately recognized the fingerprints of Wegener's granulomatosis: upper respiratory tract symptoms (which had first appeared harmless) followed by bleeding from the lungs and evidence of inflammation within the kidneys. After the diagnosis was confirmed by lung biopsy, Todd immediately started on aggressive treatment consisting of daily cyclophosphamide and high doses of prednisone.

Within days, Todd's breathing status had improved. He was discharged from Johns Hopkins Hospital after two weeks, and went home to Selbyville, Delaware to recuperate. He returned to Hopkins frequently to see Dr. Hellmann. Eight months later, Todd finally discontinued the last of his treatments. In 1996, Todd married and is now the father of a little girl! He works full time as a carpenter with a local company. Eight years after the start of his illness, he remains in complete remission.



**Todd Dickerson**

Several years later, in April of 1997, Todd's grandmother, **Betty Dickerson**, developed excruciating headaches on the right side of her head. The pain became so intense that she essentially became disabled. Every bone and muscle in her body hurt. When she attempted to eat, she experienced jaw pain so severe that she was unable to chew her food.

She was referred to the Johns Hopkins Vasculitis Center, where she met Dr. John Stone.

After reviewing Ms. Dickerson's symptoms and performing a physical examination, Dr. Stone suspected the diagnosis: giant cell arteritis. The telltale finding was the presence of a tender, thickened temporal artery, just above and in front of Ms.



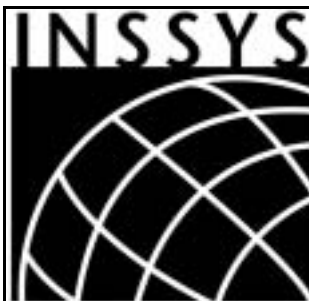
**Betty Dickerson**

Dickerson's right ear. That afternoon, Dr. Stone asked Ms. Dickerson to undergo a temporal artery biopsy (a simple outpatient procedure in which a portion of the artery is removed for examination under the microscope). The biopsy confirmed the diagnosis of giant cell arteritis. Ms. Dickerson began treatment with prednisone that evening, and felt dramatically better 48 hours later. Within a couple of weeks, all of her symptoms were gone. After a year of treatment, she was able to discontinue her prednisone completely. She has been off prednisone entirely for nearly 3 years now.

The occurrence of 2 cases of vasculitis with the same family raises an interesting question: *Does genetics have anything to do with vasculitis?* More specifically, "Will my children get this disease too?" In fact, the Dickerson's story is unusual. Although vasculitis occurred in both grandmother and grandson in the Dickerson family, familial cases of vasculitis are rare. Vasculitis Center patients are usually reassured that the chance of one of these diseases occurring in a family member is low. Nevertheless, when the causes of vasculitis are understood, it is likely that genes will be found to play some role. The likelihood that many genes contribute to these diseases makes singling out the ones responsible a challenge. We will report further on a Vasculitis Center project to investigate the role of genes in vasculitis in an upcoming issue.

## American College of Rheumatology Conference

Each year, Rheumatologists from around the world gather to review new clinical and research findings at the American College of Rheumatology (ACR) Conference. This year's conference was held in San Francisco, CA. Faculty and staff of the Vasculitis Center made several presentations about ongoing research at the Center. **Dr. John Stone** and **Misty Uhlfelder** presented the work of an international group of vasculitis investigators at nine centers in a poster entitled "A Giant Cell Arteritis-Specific Quality of Life Instrument". At a concurrent session, **Dr. Michael Regan** gave an oral presentation of the recently completed study entitled "*Chlamydia pneumoniae* and Temporal Arteritis: Failure to Detect the Organism by Polymerase Chain Reaction in 90 Cases and 90 Controls".



The official logo for  
INSSYS: International  
Network for the Study of  
Systemic Vasculitides

At the same time as the ACR conference, the International Network for the Study of the Systemic Vasculitides (INSSYS) convened its 10<sup>th</sup> annual meeting to discuss the results of ongoing international collaborations and to plan new studies. INSSYS is a

worldwide organization consisting of physicians (Rheumatologists, Pulmonologists, Nephrologists, and Ear/Nose/Throat specialists), scientists, nurses, and research staff whose work relates to vasculitis. This year, the INSSYS meeting was attended by members from 14 countries in North and South America, Europe, and Asia. Drs. Gary Hoffman (Chairman) and John Stone (Vice-Chairman) directed the meeting. Dr. Stone provided an update on the Wegener's Granulomatosis Etanercept Trial (the largest INSSYS project to date) coordinated at Johns Hopkins. Misty Uhlfelder outlined plans for the completion of the giant cell arteritis quality of life project. Finally, Dr. Michael Regan proposed a new treatment study of high-dose cyclophosphamide in patients with refractory vasculitis (*see details in the story below*). With 14 new

research projects presented, the 2001 meeting was the most exciting INSSYS gathering since its inception in 1992.

## High Dose Cyclophosphamide (CYC)

Oral CYC (Cytoxan<sup>®</sup>) remains the most effective medication for the treatment of many severe forms of vasculitis. The CYC regimen most commonly used now includes using a combination of low dose, daily CYC and prednisone. This approach has many potentially serious side effects that limit

**Immunoablative  
CYC may "re-boot"  
the immune system,  
hopefully inducing  
vasculitis cures.**

its use. (For more about CYC, see this issue's Clinic Corner). In addition, vasculitis often flares once CYC is discontinued.

The Johns Hopkins Vasculitis Center is now evaluating a different approach to the use of CYC in vasculitis. This new approach is designed to control the disease quickly, yet (it is hoped) decrease the number of side effects and lead to lasting disease remissions. The new approach involves the use of CYC in "immunoablative"

doses. "Immunoablative" means that CYC is given in sufficiently high doses to completely suppress the bone marrow's production of red and white blood cells for 2-4 weeks after treatment (Oncologists often use this kind of treatment for some kinds of cancer). The rationale behind using this type of therapy in vasculitis is that once the bone marrow recovers, the "re-booted" marrow may no longer produce cells that cause inflammation in blood vessels.

This type of treatment, which has been used in several other types of autoimmune disease, was pioneered by the oncologists at Johns Hopkins.

**Dr. Michelle Petri**, a Hopkins rheumatologist, has used the treatment in lupus. Early results of this approach to the use of CYC are encouraging.

**Dr. Michael Regan** and his collaborators at The Sidney Kimmel Comprehensive Cancer Center at Johns Hopkins are leading this new study.



## Thank You For Your Recent Contributions to The Vasculitis Center

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*Johns Hopkins is an active center for research. Research is essential to the discovery of causes and the development of cures for vasculitis. Much of this research is made possible by private support. If you would like to support vasculitis research, please send your tax deductible contribution, payable to The Johns Hopkins Vasculitis Center (1830 E. Monument Street, Suite 7500, Baltimore, MD 21205). Questions about philanthropic giving may be directed to Mandy Masterson at (410) 614-6005.*

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*Senior Clinic Coordinator, Judy Harrison, with Vasculitis Center patient Christine Harris. Ms. Harris, whose Wegener's granulomatosis is in remission, is a graduate student in psychology at Rutgers University.*

## Clinic Corner

Cyclophosphamide (CYC) is one of the "big guns" used to treat systemic vasculitis. We prescribe CYC when vasculitis threatens either the patient's life or the function of a vital organ. Below, we have listed a number of important ways to prevent the side-effects of CYC. Each point is crucial:

1. Get blood & urine tests every 2 weeks. Ensure that your doctor receives the test results and adjusts the CYC dose appropriately
2. Drink 8 tall glasses of water a day
3. Take CYC in the morning, not the evening
4. Take the prescribed antibiotic to prevent pneumonia (Bactrim or Dapsone)
5. Make sure that you receive the following vaccines:
  - the influenza (flu) vaccine every year
  - the pneumococcal (pneumovax) vaccine once every 5 years
6. Follow-up with The Vasculitis Center for clinic visits as instructed

We have developed a pamphlet that provides more information about CYC and avoiding its side-effects. You may also wish to read about CYC on our website:

<http://vasculitis.med.jhu.edu>

### In Upcoming Newsletter Issues

- A "Commonly Asked Questions" Section
- Proteomics: The doctor's tea leaves?
- A study of genes and vasculitis

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