YOU'RE INVITED TO JOIN US IN WASHINGTON DC!

September is the month for spreading the word. Join us for our first Capitol Hill Day and our 8th Annual Patient/Doctor Meeting. This year’s events will take place in Arlington, VA, just a few miles from Washington DC. Capitol Hill Day will be on Friday, September 23, 2005. The 8th Annual Patient/Doctor Meeting is Saturday, September 24 and Sunday, September 25, 2005. Look inside for registration information for both events!

Capitol Hill Day is an opportunity for our collective voices to be heard by our US elected representatives. Current research funding is in jeopardy and could affect all of us. In order for the NIH to get money to increase medical research, it would need an increase of at least 6%. The House of Representatives has presented only .5% increase proposal in the NIH budget. The Senate is asking for a 6% increase. If they meet in the middle and get 3%, then the only thing that is going to happen is research that is currently being funded will continue to be funded. However, there will be no money for new research. If 3% is not even funded, then some ongoing projects will be halted. Sadly, skin disease research is at the bottom of the list.

The IPF has learned recently that Medicare has cut reimbursement for immunoflourescence tests. This means that basic lab costs will not be covered and labs will be forced to stop immunoflourescence tests. What does that mean for us? With so many doctors unfamiliar with the best way to diagnose and treat pemphigus and pemphigoid, diagnosis could take longer than it does now and patients will suffer. Good labs will be squeezed out and only the second rate ones who are willing to cut costs will continue to be in service. This might mean that our tests could even be compromised.

On page 3 you will find a sample letter for you to send to your Congressional Representative. This sample format can also be used anytime you want to send a letter to your representatives, just change the body of the letter to meet your specific needs. We need your voice to save our lives and the lives of those who have yet to be diagnosed. Join us on Capitol Hill Day and exercise your constitutional right to have your opinion heard.

The 8th Annual Patient/Doctor Meeting will be held at the Hilton Arlington in Arlington, Virginia. Beginning on Saturday at 7:30 am, Support Group Leaders and Telephone Contacts

decided on page 3…
The IPF has taken two giant steps forward. I will no longer be Interim Executive Director. Our new CEO is Scott Peter Leigh. I will now be able to direct my attention towards patient needs as the Director of Patient Services. I want to hear your thoughts on directions we should take in expanding our services to you. You can send me your comments by e-mail (jsegall@pemphigus.org) or online at www.pemphigus.org/contact.html.

Our second giant step - We've moved! We have moved our offices to Sacramento, California. We have a terrific support group and network in that area, and it is the State Capitol so we will be able to use our resources more effectively and efficiently.

These are good changes in our efforts to assist people living with pemphigus and pemphigoid to find the help they need. It is because of you and your support that we can provide patients, friends, families, doctors, and many others with this help.

Our success depends on you – keep supporting the IPF and help save a life.

Lastly, mark your calendars for this September's Capitol Hill Day & our 8th Annual Patient/Doctor Meeting in Virginia!

I am excited to join the IPF family as CEO! But first, let me tell you a little about myself. My name is Scott Peter Leigh and I was born and raised in the Bay Area. I have a rich background in non-profit healthcare organizations as well as teaching ethics to nursing students. My fiancée and I live in the Sacramento area with 2 teenage daughters, 4 cats, 2 dogs, and over 350 walnut trees! In my spare time I enjoy snorkeling, weight-lifting, gardening, and travel.

My first order of business will be developing a Strategic Plan and building "bridges of relationships" with donor companies, individuals and groups. I am here to help. Feel free to email me anytime with your comments and suggestions at Scott@pemphigus.org. Remember, we are all in this together.

What better way to use my compassionate skills at organization, funding, and leadership than to serve individuals with a difficult and often life threatening disease? For years Janet has selflessly led the path towards education, service to patients, and research. I am honored to join her, Will, and all the Board Members as we continue to serve & advocate for all those affected by pemphigus and pemphigoid.

The quarterly is a 501(c)(3) nonprofit organization. Our goals are to increase awareness of pemphigus and pemphigoid among the public and the medical community; to provide information and emotional support to pemphigus or pemphigoid patients and caregivers; to provide referrals to specialists; and to support research into advanced treatments and a cure.

Founded in 1994.

The Quarterly is published: Spring, Summer, Fall and Winter. The material presented in our journal is not intended as medical advice. Readers are urged to consult their physicians before making any changes in their health regimen. The contents of the Quarterly cannot be reproduced or copied without written permission of the International Pemphigus Foundation. The opinions of contributors are not necessarily those of the International Pemphigus Foundation.

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Dear Representative XXX (or Senator XXX):

As one of your constituents, I would like to express my opinion to you on an issue that is very important to me and all of us living with the life-threatening autoimmune diseases pemphigus and pemphigoid.

Pemphigus and pemphigoid affect the skin and/or mucous membranes. Because it is an autoimmune disease, the body will not heal on its own and will often need massive doses of dangerous drugs to obtain some relief. Also, this disease can move quickly and cover an entire body within several weeks. For this reason, the immunofluorescence test is an extremely important diagnostic tool for me and others with the disease. Good labs will be squeezed out and only the second-rate ones willing to cut costs will continue to be in service. With so many doctors and labs already unfamiliar with these diseases and how to perform the test correctly, diagnosis could take longer and patients will suffer needlessly and in excruciating pain.

If you are a member of the Health Care Finance Committee, please reconsider this adjustment in the reimbursement of immunofluorescence testing (CPT 88347), and if you are not on the Committee, please help us fight for our lives.

Thank you for your time and consideration.

Sincerely,

[Your Signature]

[Your Typed Name]
A recent report by Dr. Aimee S. Payne and her co-workers (The Journal of Clinical Investigation 115:888-899, 2005) from the University of Pennsylvania Department of Dermatology and Pathology describes the use of a new technology to genetically characterize and analyze antibodies from a pemphigus vulgaris (PV) patient. This molecular technology, called phage display, allows the cloning of human monoclonal antibodies from a PV patient.

How are human monoclonal pemphigus antibodies cloned?

Pemphigus is a disease that is caused by autoantibodies that bind to a family of molecules called desmogleins (Dsgs). In mucocutaneous PV, Dsg3 and Dsg1 are “attacked” by the autoantibodies. We know from previous work that these anti-Dsg antibodies (also called immunoglobulins) cause the blisters characteristic of pemphigus.

A relatively new molecular biologic technique called phage display, allows the cloning of human monoclonal antibodies from a PV patient. This molecular technology, called phage display, allows the cloning of human monoclonal antibodies from a PV patient.

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A relatively new molecular biologic technique called phage display was used to clone these autoantibodies. In this technique the DNA encoding all antibodies contained in a PV patient’s serum was amplified from the patient’s peripheral blood lymphocytes. The DNA was cloned into phage, which are viruses that infect bacteria. The cloning is done so that each phage (i.e. viral) particle contains the DNA for a single immunoglobulin molecule, and that actual antibody molecule is expressed on the surface of the phage particle. Thus, outside each phage is a monoclonal antibody, and inside is the DNA encoding that antibody.

The pool of phage can be considered a “library” that contains all the monoclonal antibodies that make up the total immunoglobulin pool in the patient. However most of these antibodies have nothing to do with pemphigus, but are immunoglobulins for other purposes (mostly the prevention of infectious disease). How, then, do we select the specific anti-Dsg antibodies from this library of antibodies? The answer is to take the library of phage particles and “pan” them on a plate that contains Dsg3 or Dsg1. The phage that have antibodies against the particular Dsg stick to the plate, and the other phage (that are not related to disease) are washed away.

These anti-Dsg phage can then be grown as individual colonies (i.e. clones) and the DNA inside can be sequenced to determine the gene families from which the immunoglobulins are derived. (Immunoglobulin molecules are derived in a complicated way from scores of individual gene families). Furthermore the cloned phage can be grown in bacteria in such a way that they produce essentially unlimited amounts of the monoclonal antibodies that can be characterized as pathogenic or non-pathogenic in cell culture or mouse models of disease.

In this way, Dr. Payne and her colleagues have produced pathogenic and non-pathogenic PV monoclonal antibodies and shown that they are derived from a very limited number of immunoglobulin gene families.

Why is the cloning of human monoclonal antibodies potentially important?

We know that patients’ sera contain a mixture of antibodies to different parts of the Dsgs, and that not all the antibodies are pathogenic (i.e. that antibodies to some parts of the Dsg molecule do not cause disease). In addition, most antibodies in a patient are not in any way related to pemphigus (i.e. are not directed against Dsgs), but are present to protect the patient from infectious disease.

Currently, therapy for pemphigus depends on generalized immunosuppression to lower total antibodies in blood, but antibodies are necessary to prevent infection, therefore, current therapy is a balance between enough immunosuppression to control disease but not too much immunosuppression which would cause
Continued from CLONING, page 4... susceptibility to infectious disease. In other words, the current therapy is not targeted in any way to immunoglobulins that are pathogenic compared to all the immunoglobulins (most of which are not at all related to pemphigus) in an individual.

By cloning human monoclonal PV autoantibodies, we can determine: a) which specific antibodies are pathogenic and which are not; b) what immunoglobulin gene families are used in anti-Dsg antibodies and anti-Dsg pathogenic antibodies.

These discoveries, then, may allow us to direct therapy just at the immunoglobulin gene families that are used by the pathogenic antibodies (as opposed to all immunoglobulins). In addition, we can use pathogenic monoclonal anti-Dsg antibodies to screen large libraries of peptides to find those that bind the pathogenic antibodies (as opposed to all immunoglobulins). In addition, we can use pathogenic monoclonal anti-Dsg antibodies to screen large libraries of peptides to find those that bind the pathogenic antibodies, thereby identifying specific peptides that might block disease-causing immunoglobulins.

**Hope for the future**

Characterizing pemphigus autoantibodies at the genetic and functional levels will hopefully be a step towards finding more specifically targeted therapies for these severe diseases.

Cloned anti-Dsg3 phage

Sequence cloned DNA (inside phage)

Produce monoclonal anti-Dsg3 IgG

Analyze monoclonal anti-Dsg3 IgG for ability to cause disease

**FUNDING & PATIENT CARE AMONG TOPICS Discussed FIRST IAPO MEETING HELD IN UK**

The International Alliance of Patients Organizations (IAPO) held its first ever Global Patients Congress in London from February 25-27, 2005. Bringing together IAPO’s member-patients’ organizations from around the world, of which the IPF is one, over 70 patient leaders from a diverse range of therapeutic backgrounds exchanged valuable skills and experiences and worked together developing strategies to bring patients to the center of health care systems.

Attendees were from disease specific and cross-disease organizations. Millions of patients were well represented with conditions including Alzheimer’s disease, arthritis and rheumatism, autoimmune related diseases, Crohn’s disease, cancers, diabetes, endometriosis, genetic conditions, haemophilia, headache disorders, heart conditions, HIV/AIDS, interstitial cystitis, Marfan syndrome, mental health, multiple sclerosis, myotonic dystrophy, osteoporosis, rare diseases like pemphigus and pemphigoid, other skin diseases, and ulcerative colitis.

Peter Foldes, our volunteer in Paris, represented the IPF at the meeting. Here are Peter’s impressions of the meeting.

“Initially I was somewhat dubious about the aims and purpose of such an organisation as IAPO, despite being bombarded with documentation. But apart from the location, a hotel near Heathrow airport, which I suppose was chosen to accommodate the 106 delegates from various destinations who mostly flew into London, the event was well organised and the diversity of those attending interesting. It also drew representatives from the World Health Organisation, the European Commission in Brussels, the Pharmaceutical Industry and the medical professions.

The event was organised on Friday around the AGM of IAPO, which I did not attend and an international patients group meeting in which I was involved. The issue of international and national was a major issue for most groups as above all they competed for sources of funding, essentially from pharmaceutical companies. I am happy to report that pemphigus associations and particularly the IFD do not suffer from these conflicts as there is a common interest to regroup as many people as possible because of the rareness of the illness.

The issue of conflicting groups for the same illnesses due to clashes of personalities was an issue.

On Saturday and Sunday the meetings were split into a plenary meeting with key note speakers and working groups. In the plenary meetings the idea of patient centred care was the main issue. This would seem obvious but not always easy to put into practice. The medical professions idea of “identify and fix” tended to go against this ideal, despite denials. There was also concern, for particular chronic or rare illnesses that the patient knew more about their illness than the medical profession on the defensive and did not necessarily lead to good relations.

In terms of those attending the majority came from the US, Canada and Europe. The exception was from Australia, Uruguay, Pakistan and Nigeria. Third world countries were very poorly represented; where the health care issues were dramatically different. In Nigeria there are 4 million people with Aids, but medicine for 14 thousand, mostly from Trust funds in the States. In Pakistan apart from the problem of the absence of doctors in the countryside where 80% of the population live, the hierarchical family structure can be disruptive in the case of illness. A grandfather with Alzheimer’s totally disrupted the family life until it was correctly diagnosed.

The other issue was the growing number of generic organisations or groups like chronic pain diseases or even rare diseases which cut across existing organisations. At times it was a mystery to know why and for what purpose these groups existed. In the case of the grouping of rare diseases there was some logic, but others just duplicated existing groups and above all were a drain on the limited resources available.

As a conclusion it was a well structured and interesting event.”
WHEN A DOG TAKES CHARGE....

**TAKING A BITE OUT OF PF**

by Kirsten R Bellur

If Krista could talk, I am sure she would like to share her story of remission with everyone. But as she is a beautiful, black, 130-pound pedigreed Newfoundland dog descending from a pure bred family of show dogs, she will let me tell her tale.

Krista did not follow in the paw-steps of her family and go into show business; instead she was adopted and joined the family of my friends, Birthe and Verner Stenderup, and became a “valley girl” - for sure - totally. She lives on a farm at the foot of Bear Mountain, the Tehachapi Range, in the sunny San Joaquin Valley of California. She enjoys the good life and long walks in the vineyard, through the carrot, potato, and onion fields or whatever other vegetable is in season. She finds time to play with her friends Musekat (mouser), and her sister Gidget - both black cats. When it gets too hot in the valley, she takes long naps on a cool tile floor. She is an extremely kind and affectionate dog, therefore, because of her sweet disposition, she is dearly beloved by all in the valley.

A year ago or so, I visited the Farmer in the Dell, when Krista was not feeling well.

I found my friends so very concerned, the wounds on her legs had again returned. The sadness was written all over the sweet face of Krista, caused by the great pain of the reoccurrence of blisters.

The Stenderup’s had taken their pet to the Vet., And she was once more back on Pred.

I could relate to Krista’s pain for sure, I had been like her, but knew there was a cure.

I told my friends: “I do believe, your poor pooch has pemphigus indeed - not something else as first perceived.

Please return to the vet with your pet; do request her to kindly change view, so Krista can again be as good as new.”

An article - “Pemphigus in Dogs” - they would show the vet, if Krista had PF her meds should be imuran and pred instead.

With a taper schedule to follow, Krista should heal, and soon recover.

The Stenderup’s returned to the vet with their pet. The vet confirmed that Krista had PF instead. She changed the meds to meet Krista’s needs, and soon there after her skin was beautiful indeed.

Then, gradually tapered her drugs according to plan and before too long, she was drug-free, healed, and happy again.

A year later I am pleased to tell, there is a very happy ending to this dog’s tale.

Krista is a jolly tail-wagging dog once more, she is man’s best friend and by all adored.

Living happily ever after with the Stenderup’s on the farm - enjoying remission – PF-symptom-free – is an added charm.

Therefore to this tale the moral should be:

Early detection does lead to early remission in pemphigus indeed. •
2005 IPF PATIENT/DOCTOR MEETING, Washington D.C., September 23 - 25, 2005

CAPITOL HILL DAY/ANNUAL MEETING REGISTRATION FORM

NAME: ____________________________________________  AGE: __________________

ADDRESS ____________________________________________  # of Previous Conferences Attended _________________

CITY ____________________________________________ STATE __________________ ZIP ______________

COUNTRY (if other than United States) __________________________ PHONE ____________________________________

EMAIL ____________________________________________ ARRIVING ___________ DEPARTING ____________

*I require special assistance (please attach a written description for all persons with special needs) Yes ☐ No ☐

Will you be staying at the Hilton Arlington? Yes ☐ No ☐

If NO, then where will you be staying? ____________________________________________

You are responsible for making your own hotel reservations by calling (703) 528-6000 or (800) HIL-TONS. Don’t delay! Be sure to say you are a part of the International Pemphigus Foundation group to get your special room rate. Reservations are limited, and are on a first-come, first-served basis. Reservations must be made by Tuesday, August 30, 2005.

Hotel Confirmation # __________________  * Names* of additional persons sharing room ___________________________

CAPITOL HILL DAY: Please make arrangements for me to meet my legislators or staff on FRIDAY, SEPTEMBER 23, 2005. Yes ☐ No ☐

U.S. Senators ____________________________________________and ________________________________ ☐ Please find out for me

U.S. Representative ____________________________________________ ☐ Please find out for me

REGISTRATION - EARLY REGISTRATION IS ENCOURAGED

Registration must be received by the IPF no later than September 9, 2005.

<table>
<thead>
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<th>Event</th>
<th># of people</th>
<th>Price</th>
<th>Total</th>
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<tr>
<td>8th ANNUAL PATIENT/DOCTOR MEETING REGISTRATION FEE</td>
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<td>$85.00</td>
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<tr>
<td>Saturday Night Dinner</td>
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<td>$40.00</td>
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<tr>
<td>CAPITOL HILL DAY (includes transportation to/from hotel/capitol and breakfast)</td>
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<td>$50.00</td>
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<tr>
<td>Optional Scholarship Donation (help someone in need attend this year’s meeting)</td>
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<tr>
<td>☐ I request scholarship assistance. Please accept this donation to help defray costs. *</td>
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GRAND TOTAL $ __________________

* If you are requesting scholarship assistance, please contact the IPF prior to sending in your form.

Check ☐ Money Order ☐ Visa ☐ Master Card ☐

Card # __________ – __________ – __________ – __________ Expiration Date: _____ / _____

Name on Card ______________________________ Signature ______________________________

PAYMENT MUST ACCOMPANY THIS FORM. ALL PAYMENTS MUST BE MADE OUT TO THE IPF IN U.S. DOLLARS. MAIL PAYMENT AND COMPLETED REGISTRATION FORM TO INTERNATIONAL PEMPHIGUS FOUNDATION, 828 San Pablo Ave., Suite 210, ALBANY, CA 94706.

Please list names and ages of other guests who are attending.

Name ______________________________ Age __________________ Special assistance? ☐

Name ______________________________ Age __________________ Special assistance? ☐

Name ______________________________ Age __________________ Special assistance? ☐

Name ______________________________ Age __________________ Special assistance? ☐

Name of person(s) with Pemphigus/Pemphigoid ______________________________

Cancellations are not refundable after September 16, 2005.
# EVENT & SESSION SIGN-UP SHEET

Use an X to mark the events you plan to attend and use the letter code for your preferred Session Choices. Session Choices Descriptions are provided after the registration kit.

<table>
<thead>
<tr>
<th>Johnny Example</th>
<th>Self</th>
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## FRIDAY, SEPTEMBER 23, 2005 – CAPITOL HILL DAY

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<tr>
<th>Time</th>
<th>Event</th>
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<tr>
<td>11:00 am</td>
<td>Capitol Hill Visits</td>
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<tr>
<td>7:00 pm – 10:00 pm</td>
<td>Meet &amp; Greet</td>
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## SATURDAY, SEPTEMBER 24, 2005 – MEETING/DAY I

<table>
<thead>
<tr>
<th>Time</th>
<th>Event</th>
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<tbody>
<tr>
<td>8:00 am – 9:15 am</td>
<td>New Patient Orientation and Information Session</td>
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<tr>
<td>11:30 am – 1:00 pm</td>
<td>Morning Age-Specific Break Out Session</td>
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<td></td>
<td>1 - Children Living with Pemphigus</td>
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<td>2 - Young Adults</td>
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<td>3 - Adults</td>
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<tr>
<td>2:15 pm – 4:30 pm</td>
<td>Afternoon Concern-Specific Break Out Sessions</td>
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<td>A - Caregivers Workshop</td>
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<td>B - Pain Management &amp; Wound Care</td>
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<td>C - How To: Insurance and Disability</td>
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<td>D - Dental &amp; Oral Care</td>
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<td></td>
<td>E - Chronic Illness &amp; Nutrition</td>
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<tr>
<td>6:30 pm</td>
<td>Reception (cocktails &amp; hors d’oeuvres)</td>
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<tr>
<td>7:00 pm</td>
<td>Dinner</td>
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Are you available to volunteer some time to help the IPF? Please indicate which type of volunteer work you prefer and the times & dates you are available (for example, when you are not attending a session). You will be contacted prior to the meeting with a volunteer schedule.

- Registration Assistant: Greet attendees and distribute registration packets.
- Capitol Hill Day Volunteer: If you are joining us for CHD, help others locate and use the Metro.
- Foods & Disease Session Leader: If you are a certified nutritionist, we need a session leader.
- Short Term Child Care: We need someone to watch children for a short amount of time.

Name: ___________________________ Phone: ___________________________

Available on (day/time): __________/________, __________/________, __________/________
WILL YOU BE AFFECTED?

from the National Health Council

Medicaid: A Quick History

Since its creation in 1965, Medicaid has been the sole source of affordable, comprehensive health care coverage to millions of people with chronic diseases and/or disabilities. Today more than 52 million Americans receive Medicaid benefits. Medicaid has, among other things, funded innovations in the delivery of health care and functioned as the nation’s primary source of long-term care financing. Medicaid is part of the Centers for Medicare & Medicaid Services, a division of the US Department of Health & Human Services.

Medicaid Plays Key Role for People with Chronic Diseases and/or Disabilities

The role of Medicaid in providing necessary health care coverage to people with chronic diseases and/or disabilities cannot be underestimated. Although low-income children and their parents make up 75 percent of those enrolled in Medicaid, they account for only 31% of the program’s expenditures. 69 percent of Medicaid spending is used in caring for people with chronic diseases and/or disabilities.

Services Covered by Medicaid

There are two levels of services within the Medicaid program, those that states are required by federal law to cover and those considered “optional” or additional services as determined by each state.

Benefits that states must provide include inpatient and outpatient hospital services, physician, midwife, and certified nurse practitioner services, lab and x-ray coverage, nursing home and home health care for individuals aged 21 and older, family planning care and supplies, and rural health clinics. Falling into the “optional” category are such benefits as prescription drugs, clinic services, hearing aids, and dental care.

How Federal Funding Cuts Hurt People with Chronic Diseases and/or Disabilities

Congress is considering proposed federal budget cuts to the Medicaid program for FY 2006 of between $45 and $60 billion over 10 years. Deep federal funding cuts or caps to the program will greatly strain already strapped state Medicaid programs and could force states to make changes that will have a disproportionate negative impact on patients with chronic diseases and/or disabilities.

So called block grants and caps are a dramatic change from the federal government’s current and long-standing commitment to providing ongoing resources to states to help finance Medicaid. The current federal matching system provides a stable and open ended funding mechanism for state Medicaid programs, which is necessary to ensure access to care for patients with chronic diseases and/or disabilities.

Alternative funding structures like block grants, which are one time only allocations to states to fund public programs, do not provide the stability necessary to run a program like Medicaid that meets the ongoing needs of millions of vulnerable Americans each day.

CONGRESS CONSIDERS $45B-$60B IN MEDICAID CUTS OVER 10 YEARS

DONATE NOW! SUPPORT OUR EFFORTS!

Contribute to the International Pemphigus Foundation and receive the IPF Quarterly. It has the latest information on research, plus valuable, pertinent and accurate information for those who live with pemphigus and pemphigoid and their caregivers. And best of all, your contribution not only covers the cost of your newsletter, but also goes to support all of our patient services as well as the research that could eventually cure pemphigus and pemphigoid.

Donate today!
WHY IS THE MORTALITY OF BULLOUS PEMPHIGOID GREATER IN EUROPE THAN IN THE US?

BULLOUS PEMPHIGOID MORTALITY RATES

by Jean-Claude Bystryn, M.D. and Jennifer L. Rudolph, M.D.

Originally published in the Journal of Investigative Dermatology, March 2005

What is the prognosis of bullous pemphigoid (BP)? The answer is critical to provide meaningful information to our patients. Unfortunately, the answer is confusing.

The reported mortality of BP is much greater in Europe than in the US. Mortality, one year after initiation of therapy, ranges from 19% to 41% in Europe but is only 6% to 12% in the US. This difference is supported by an increasing number of reports which are remarkably consistent in their observations. Five European studies report the 1-year mortality as 19%, 29%, 30%, 40%, 41%, respectively; whereas three reports from the US report it as less than 6%, 11%, and 12%.

In our own unpublished observations on 135 sequential BP patients treated in a tertiary referral center in New York City during the past decade, mortality at one year was less than 10%.

Dr. Pascal Joly and his colleagues in the March 2005 issue of the Journal of Investigative Dermatology (JID) offered several possibilities to explain this difference in outcome. These included differences in treatment, ethnic differences, age, study size, and patient selection bias.

There is an obvious difference in treatment, as the majority of patients reported from Europe are hospitalized for treatment whereas this is the exception in the US. The immediate suspicion that complications of hospitalization ARE the cause OF higher mortality in Europe is, however, refuted by Dr. Ignacio Garcia-Doval and colleagues, who indicate that the greatest mortality in their patients occurred after hospital discharge.

Other aspects of treating BP in Europe and the US are generally similar, and so do not provide a ready explanation for the differing mortality rates.

Nor does difference in age provide a likely explanation for the higher mortality reported in Europe. The average age of patients in two European (Rzany et al, 2002; Garcia-Doval et al, 2005) and one American study (Colbert et al, 2004) is identical, i.e., 77 years. Nonetheless, mortality was still three to four times higher in the European studies.

American studies are generally smaller than those conducted in Europe. However, the consistency of the reports, i.e., mortality in all European studies, is higher than in all American studies, and the clear nature of the end-point (death) suggest the reported differences between European and American studies are real.

We suspect the most likely explanation for the differences in BP mortality between Europe and the US is a patient selection bias, as initially suggested by Dr. Neil Korman (1998). The published evidence suggests that BP studies in Europe were conducted in patients who were generally sicker than those in the US, as was initially argued by Dr. Korman (1998). Whether this is because patients with more severe disease or comorbidities are not treated in tertiary medical centers in the US, as suggested by Dr. Joly et al (2005), or conversely because such patients are preferentially referred to tertiary centers in Europe as we suspect, the results will be the same mortality will appear worse in Europe than the US because it is based on a subset of patients with BP who are sicker and/or have more comorbidities.

Lastly, the true mortality of BP cannot be discerned from any of these studies, since none is adjusted for the expected mortality of age-matched patients with similar comorbidities.
The Division of Dermatology at Duke University Medical Center is recruiting patients for a clinical study using rituximab (Rituxan®) to treat patients with bullous pemphigoid who continue to have blisters, skin ulcers or other disease activity despite therapy with 17.5 mg/day or more of prednisone therapy. Eight patients will be treated with intravenous infusions of rituximab on day 1 and day 15 and followed monthly for 6 months and then every 3 months for a total of 1 year.

The purpose of this trial is to determine the safety of rituximab for the treatment of patients with bullous pemphigoid. Secondary clinical endpoints include the number of days to cessation of new blister formation, the ability to reduce prednisone to 25% of the initial dose by week 24 and bullous pemphigoid antibody levels at week 24.

Patients must be at least 18 years of age with a clinical diagnosis of bullous pemphigoid and ongoing disease activity on 17.5 mg/day or more of prednisone. Patients that are taking other immunosuppressive drugs such as azathioprine (Imuran), mycophenolate mofetil (Cellcept), methotrexate, or cyclosporine A will undergo a 4-week washout phase before beginning the trial. Pregnant or nursing women are not eligible to participate in the trial. All potential participants will have a screening visit which will include a review of their medical history, physical examination and obtaining of a blood sample to determine their eligibility to participate in the study.

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