



ANTIPHOSPHO...WHAT?

APS Foundation of America, Inc. Newsletter

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What is "CAPS"?

Written by: **Thomas L. Ortel, M.D., Ph.D., Professor of Medicine & Pathology
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A 'catastrophe' refers to something that is disastrous, potentially associated with a large loss or extreme suffering. Catastrophic Antiphospholipid syndrome (also called catastrophic APS, or "CAPS") describes a rare complication that can develop in certain patients with APS. Typically, patients with APS have an increased risk for clots affecting the larger veins (causing deep vein thrombosis or pulmonary embolism) or arteries (causing a stroke or heart attack), or recurrent miscarriages. Catastrophic APS, on the other hand, characteristically involves tiny blood vessels in more than one organ system, such as the brain, lungs, and/or kidneys. These clots can rapidly develop over the course of several days to weeks.

Most patients with APS will never develop catastrophic APS; this extreme presentation occurs in fewer than 1% of all patients with APS. Several factors have been identified that may precipitate the development of catastrophic APS, including infections, prior surgery or trauma, stopping anticoagulant therapy, and pregnancy-related complications. Why these triggers cause catastrophic APS in some patients but not in others is currently not known. Although precipitating events can be identified in most patients, up to a third of the patients with catastrophic APS have no clear 'triggers' prior to their initial presentation.

Of those patients who develop catastrophic APS, about half have a history of prior deep vein thrombosis, stroke, or other symptoms of clots. However, some individuals may have had no manifestations of APS prior to their initial presentation with this potentially life-threatening complication. A similar syndrome can sometimes be seen in patients who don't have APS, referred to as "thrombotic storm".

The clinical presentation is usually complex, which can make this syndrome very difficult to diagnose. The organ systems most frequently involved initially include the heart, lungs, brain, and kidneys. Patients may present with a stroke

or heart valve defects, kidney failure requiring dialysis, or pulmonary emboli or hemorrhage. Many patients present with abdominal pain, which may be due to blood clots affecting various organs in the abdomen (for example, portal vein or mesenteric vein thrombosis). Although clots in the small vessels are most commonly described in this syndrome, these patients may also develop clots in large vessels, such as pulmonary emboli and strokes. An important component of the clinical manifestations of catastrophic APS is the systemic inflammatory response, which most commonly results in the acute respiratory distress syndrome (also known as ARDS). This inflammatory response can be as dangerous for the individual patient as the diffuse blood clots.

Laboratory testing in patients with catastrophic APS may reveal a low platelet count (thrombocytopenia) and evidence for diffuse activation of the clotting mechanism, referred to as disseminated intravascular coagulation, or DIC. Bleeding complications, including cerebral hemorrhage (bleeding into the brain), can develop in patients with severe DIC. Destruction of red blood cells, referred to as hemolysis, may be identified in a third of individuals with catastrophic APS. These patients may have a small number of schistocytes, or red cell fragments, that are seen in a related disorder, thrombotic thrombocytopenic purpura (also known as "TTP"). Anticardiolipin antibodies are frequently detected, but may not be present initially in all patients.

A variety of different treatments have been tried, but even with the most aggressive care, as many as half the patients with catastrophic APS will die. Important aspects of treatment include the early initiation of anticoagulant therapy, antibiotics for any potential infections, and steroids to treat the inflammatory component. Plasmapheresis, or

"Most patients with APS will never develop catastrophic APS; this extreme presentation occurs in fewer than 1% of all patients with APS."

Are You Mapped?

Do you have a pin in our map?? If not, please add it. We are watching in amazement to see the clusters of APS people forming. Our Frappr map is located at: <http://www.frappr.com/apsfa>

Patient Stories & Articles Needed!

We are in need of patient stories to feature in our newsletters. Every APS patient has a story to tell and yours could be shared with the entire APS community.

We also need related articles such as book reviews, poems, recipes, interest articles, quotes, etc.

If you are interested in sending us your story, please write to articles@apsfa.org and we will send you our guidelines.

Without your help our newsletter cannot be a success!

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Letter from the President



Spring is almost here and we will be getting ready for APS Awareness Month in June. It is mind blowing to think in June we will turn 4 years old!

I would like to thank everyone who donated to decorate our 2008 Giving Tree! Because of your generous donations, we were able to raise \$2650.00! The Giving Tree holds a special meaning for the members of the APS Foundation of America, Inc and the community it serves.

The APS Foundation of America actively works with our medical advisors and their respective facilities to get the education out about APS. We have been contacting various newspapers and media sources to get the word out about APS and the foundation. We will be really busy during June and we will need YOUR help.

You will notice that the APS Foundation of America, Inc. and the APS Friends & Support Forum are both HONCode Certified. The HON has been created for improving the quality of information intended to both patients and medical professionals for facilitating quick access to the most relevant and up-to-date medical discoveries. The HONcode is the oldest and the most used ethical and trustworthy code for medical and health related information available on Internet. The HONcode is designed for two target audiences: the general public and the web publisher, actively involving the site owner in the process of accreditation.

On a different note, we have been getting complaints about the doctors list so I have decided to address this issue here. Please make sure you have read and understand the disclaimer. The APSFA does not endorse any of these doctors, they were simply suggested to us by others. The doctors on this list may not be "experts" in APS. It simply means someone along the way has had luck with them. You may not have the same luck. Please be sure to call the office to ensure that the doctor will take your health insurance and is accepting new patients. We would also like to add, if you find a doctor has moved, is no longer seeing APS patients, or had a bad experience, please contact us. This list only works if you, the patient, report the changes and experiences. We aren't trying to black ball anyone, but speaking up is how the information contained on this site stays as accurate and useful as possible.

Once again, I hope this newsletter finds you in the best of health and with a perfect INR level.

Sincerely,

Tina Pohlman

President & Founder

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The Two Year Road To My APS Diagnosis

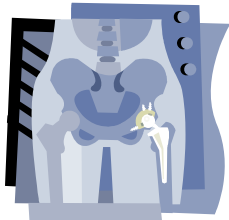
Written by: Kevin McAllorum

My Labor Day weekend in September 2006 started out weird and from there the rest of my life would be changed. Driving to work that day, I was oblivious to the blood infection that had run down my arm. Still to this day don't know if it was a spider bite or mosquito but it does not matter anymore. I got to work that day and my boss stopped me in my tracks and sent me home to see a doctor. I being the oblivious one did what I was told. Went to the doctor. Got a tetanus shot and a cortisone shot and some antibiotic cream. Still no big deal, so I did not worry. I was feeling kind of tired and knowing that we had a Labor Day party to prep the house for, I went to sleep early that Friday night. This is where the fun begins. I got up in the middle of the night and needed something to drink because the house was hot. I slid out of bed, went to stand up and fell flat on my face on the floor, my hips hurt so bad that I could not believe the pain that was searing through my brain. Ok, I slept wrong. I pinched a nerve. No big deal. I struggled leaning on the wall the whole way to the kitchen not waking the family. I went back to bed and adjusted myself so I would not sleep the same way. When I got up in the morning I could not walk, my wife started in on how it was a reaction to the tetanus or the cortisone and on Saturday called the Doctor. He advised her it was nothing of the sort, don't worry, it will pass.

Well that whole weekend I struggled to walk, had the party and tried not to move too much to not let my guests know there was a problem. The pain passed after about 6 or 7 days. I was happy. However for the next 9 months, the pain would come and go. On Memorial Day 2007, I worked around the house physically and we had some issues with teens painting the cars in the driveway, so I chased one in the early morning hours. When I came back to the house, my hips hurt again, and bad. My brother was diagnosed and had had surgery on both of his hips at the age of 33 and I was 39 at this time. My wife started to worry that I had the same disease, Avascular Necrosis of the hip. I told her there was no way, it's not genetic. But she decided to make me an appointment with the orthopedic doctor in Gainesville

Georgia anyway. So we went, he agreed with me on the fact he did not think it was Avascular Necrosis but he scheduled an MRI just to be sure.

Here is where my life went downhill, it was AVN. I had a severe case in both hips and my knees were starting to hurt as well. Two siblings having this disease— that is not supposed to happen. Both bad cases and by the look of the MRI, it was unusual. So we decided to take action. Surgery Number one was scheduled for 8/16/2007. A Core Decompression with a metal rod inserted for stability, it felt better for about 3 weeks but it took 8 weeks to recover



so I gave it time. The next Surgery was on 11/16/2007. I had been in a wheelchair since August and I spent the entire holiday in a wheelchair and using crutches. In January 2008, I began to walk with the cane, but I was still having pain and struggling and my knees started to kick into overdrive.

I went for an MRI on the knees— again, it was AVN. It's spreading, a disease that does not spread, nor run in families, is spreading, and that was a new one for the books. I went in every few months until I could not take the pain in my hips anymore. So in July of 2008, I had total hip replacement on the left side of my body. We found out that the AVN death was continuing. And in September I had the right one done as well. The right one, as I was told, was squished worse than a ping pong ball. While recovering, my knees really stepped it up. I could not believe that pain could be this bad, and I just had to live with it. I had just gone through four surgeries and I was worse than ever before. My hips felt great, but my knees were so bad. I'll admit that suicide had crossing my mind to get out of the pain I was in. I could not eat, sleep, walk, stand, or do my PT from surgery. Nothing. I felt completely useless. I lay in bed and wondered every moment of the day if I did the right thing.

When I went back to the orthopedic surgeon he said that I have some sort of blood disease that's causing this. He did not know what it was but before we do any more surgeries and

try and fix this, we need to get to the bottom of what is really going on.

So off to the cancer center I go, just before Thanksgiving 2008. I gave a pint of blood and they tested it. They tested all different kinds of things; meanwhile, I am still in pain. They found nothing.

So then, I was referred to a Rheumatologist. At this point I am ready to give up. I am still in pain, and it seems like I cannot get answers. By this time I have had about 50 X-rays, 5 blood tests, 4 MRI's, a complete bone scan, seen 5 different doctors, hospitals, and the list seems to go on and on. And I still don't have an answer.

So I'm off to another doctor wondering what they will they find. I have to go through more X-rays, another MRI, and a lot of blood tests. This is getting old to say the least. Because these it's now the Monday before Christmas, this set of tests will take 3 weeks until I can get the results. Three weeks!

The three weeks are finally up and I go in. The Doctor walks in and says, " I know what is causing your AVN, your ticks, and possibly other issues you talked about". "Actually I have two answers". I jokingly say, "Nothing" and "Nothing". She says no, your AVN is being caused by something called Antiphospholipid antibody syndrome and the extremities that are experiencing arthritic pain is being caused by Rheumatoid Arthritis.

Well, after almost 2 ½ years I had an answer. I did not know what Antiphospholipid antibody syndrome was, but I also didn't care. I broke down in tears right then. I had an answer and the doctor was hopeful that we could treat it. I didn't care what the treatment was. I had already lost bones and my hips (which are now completely metal on metal), and I'm getting ready to lose my knees, and maybe more parts but now we know what this is and we have a plan and I have a treatment.

I look forward to starting my treatment, but I worry about my other family members especially my brother who is five years younger and has AVN as well.



Medical Quackery

Submitted by: Tina Pohlman

Sure-cures for medical problems, whether chronic or terminal, drain millions of dollars from consumers' wallets each year. They also keep thousands of consumers from appropriate medical treatment.

TV infomercials, and newspaper, magazine, radio and internet ads often make fabulous claims, use official-sounding titles and testimonials from many satisfied customers. Today's snake oil sellers try to convince you they've discovered new solutions to age-old problems. They may hint that the federal government is keeping people from a product that has cured thousands in other countries. Some use national advertising to deceptively sell "miracles" they can't produce—potions and products for health, beauty, vitality and happiness.

The Bureau of Consumer Protection says there are ways to tell which health-related products are legitimate and which are not. For example, learn to recognize worthless products by the typical phrases often used to promote them.

- Does the ad promise "a quick and easy cure"?
- Is the product advertised as effective for a wide range of ailments or for undiagnosed pain?
- Does the promoter use key words such as "miraculous," "exclusive," "secret," or "ancient"?

- Is the product advertised as available from only one source, requiring payment in advance?
- Does the promoter use undocumented case histories that sound too good to be true?

Don't rely on promises of a "money-back guarantee". Be aware that many fly-by-night operators will never be there to respond to a refund request.

Protect yourself by taking the following precautions:

- Don't trust your health to a salesperson, ad or TV infomercial.
- Don't believe claims of a "secret" or "miracle drug" that works on a wide variety of ailments.
- Don't buy medical devices, bracelets or other products without consulting your doctor or an appropriate health professional.
- Be skeptical of claims of excessive or immediate weight loss or recaptured youth or sexual function.
- Don't buy any product based on the

seller's claim that the purchase will be covered by Medicare or other insurance.

Always discuss your medical problems with your family physician. If you can't get help or information you need, switch doctors—don't start buying cures through the mail or 800 numbers.

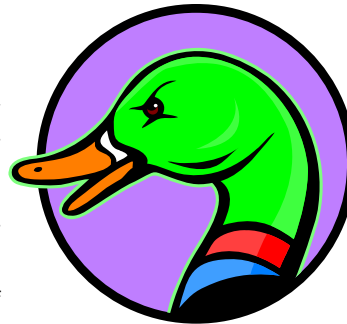
Why Health Fraud Schemes Work

Health fraud, or quackery, is a business that sells false hope. It preys on persons who are victims of diseases that have no complete medical cures, such as arthritis, multiple sclerosis, and certain forms of cancer. It also thrives on the wishful thinking of those who want short-cuts to

weight loss or improvements to personal appearance. It makes enormous profits because it claims to offer quick cures and easy solutions to better health and personal attractiveness.

While the health fraud business causes widespread economic harm, most harmful are the ones that turn people away from proper medical diagnosis and treatment of serious illnesses. In addition, some bogus products themselves may be harmful.

Again, when you have a question about the value of a product, ask your physician or pharmacist.



(Continued from page 1)

plasma exchange, is a treatment that has been used successfully in some patients. This treatment consists of removing the patient's plasma and replacing it with plasma from normal donors, and may need to be repeated several times (or more) before the syndrome is brought under control. Other therapies that have been used, with mixed levels of success, include intravenous immunoglobulin, cyclophosphamide, rituximab, and a variety of other agents.

For those patients who survive an episode of catastrophic APS, most do well with chronic anticoagulant therapy. Recurrent catastrophic APS, while uncommon, can occur. However, some patients may have recurrent 'triggers' without developing

recurrent catastrophic APS. The cause(s) of this syndrome, and the best treatments, remain a mystery.

Reference List

RA Asherson, et al. Catastrophic antiphospholipid syndrome: international consensus statement on classification criteria and treatment guidelines. *Lupus* 2003; 12: 530-534.

RA Asherson, et al. Catastrophic antiphospholipid syndrome. Clues to the pathogenesis from a series of 80 patients. *Medicine* 2001; 80: 355-377.

CS Kitchens. Thrombotic storm: when thrombosis begets thrombosis. *Am J Med* 1998; 104: 381-385.

D Erkan, et al. Long term outcome of catastrophic antiphospholipid syndrome survivors. *Ann Rheum Dis* 2003; 62: 530-533.

S Bucciarelli, et al. Mortality in the catastrophic antiphospholipid syndrome. Causes of death and prognostic factors in a series of 250 patients. *Arthritis Rheum* 2006; 54: 2568-2576.



APS Community Says Goodbye To Leading Researcher

Dr. Ron Asherson, 1938–2008



Dr. Ron Asherson - OBITUARY

Dr Ronald A Asherson died suddenly in May 2008 at the age of 73 in Johannesburg, South Africa. Dr. Asherson was the leading researcher in Catastrophic Antiphospholipid Antibody Syndrome (CAPS)

and was also starting to do more research on Microangiopathic Antiphospholipid Antibody Syndrome (MAPS).

Dr. Asherson will be greatly missed. One of Dr. Asherson's colleagues, Ricard Cervera, wrote a beautiful tribute on Dr. Asherson.

"Ron Asherson was serving as Honorary Deputy Director of Clinical Research at the Lupus Unit, St Thomas' Hospital, London,

when I joined Graham Hughes' team in 1989, in a postdoctoral fellowship that became the most exciting and gratifying period of the professional and personal lives of many overseas fellows that trained – and became good friends – at the unit at that time. Many of the currently well accepted clinical associations of the antiphospholipid syndrome (APS), including the most severe variant of the disease – the 'catastrophic' APS, were described in these very productive years, always with the clever clinical vision and quick paper writing of Ron.

Until the end, Ron continued visiting patients in his private practice clinic, writing papers, planning new projects and receiving a lot of love and respect. In his last e-mail to me, just 2 days before his sudden death, he

wrote – in addition to the many comments to the articles that we were writing together at that time (i.e., his last 'obsessions': the re-lapsing catastrophic APS and the microangiopathic APS): '...people have been wonderful... I had no idea how much love and respect I have managed to acquire in my life and this strengthens me greatly... We will be in touch in a few days. Ron'. But we were not. He fought tenaciously against the disease, but also provided an example to all by continuing with a full work load, sustained by his faith and never yielding to despair. His life and the manner of his death will remain a lasting memory to all who had the great fortune to know him."

You may find this in full at: <http://tinyurl.com/ashersonobit>

Is There A Replacement for Warfarin Coming Soon?

Written by: Ashley Garcia, Pharm D Candidate

University of Colorado at Denver

Reviewed by: Al Lodwick, RPh, MA

Most people who have antiphospholipid antibody syndrome (APS) need to be on medication to lessen the chance of a blood clot forming. For most people with APS this means warfarin. Warfarin is the medication that has the most data and has been used the longest in APS.⁵ It is important for people on warfarin to get their INR checked regularly. Most people on warfarin should assure that the INR remains in the 2.0-3.0 range. This is the therapeutic range that offers the greatest protection from a clot with the least risk of bleeding. If you have had a clot while on warfarin with an INR between 2.0 and 3.0, some experts recommend keeping your INR between 3.0-4.0.¹

Warfarin has been the standard of care for APS for years. New medications are being tested for clot prevention. You may have heard of medications that are going to replace warfarin. Some may consider these to be "wonder drugs" These medications could be used in the future for the treatment of APS. However, the clinical trials underway are not enrolling people with APS. It is also important to know that this means these drugs are new and will not have been proven safety nor effective for

APS. It is important that people with APS expect to continue taking warfarin probably at least for five more years.



One of these new medications is rivaroxaban. This medication is still in phase III trials and not currently available for doctors to prescribe in the United States. This medication is taken once daily like warfarin is. One reason people are excited about this medication is because less monitoring (blood testing and doctor visits) may be needed when taking rivaroxaban.^{2,3} More research needs to be conducted in order for the FDA to allow this medication to be on the market in the United States.

Another medication that is still in the trial phase is dabigatran. This medication is currently approved for clot prevention in Canada and Europe. There are clinical trials occurring in the United States to show that dabigatran is effective in preventing blood clots. This medication is also taken once daily like warfarin, but may need less monitoring.^{2,5} More research needs to be conducted in order to ensure this medication is safe. Studies also need to be done comparing dabigatran and warfarin.

While there are a few new medications in clinical trials for clot prevention, warfarin is still the best option for people with APS. It is important to monitor INR ranges regularly and keep communication open with your doctor and pharmacist. People with APS are at increased risk for clots in the legs, in the lungs, for strokes, and heart attacks. Warfarin therapy will help to prevent these events from occurring.

References:

1. Ruiz-Irastorza G, Hunt BJ, Khamashta MA. A Systematic Review of Secondary Thromboprophylaxis in Patients with Antiphospholipid Antibodies. [Arthritis Care & Research. Vol 57 \(8\)](#), Pgs. 1487-1495.
2. Francis CW. New issues in oral antithrombotics. [Hematology Am Soc Hematol Educ Program. 2008;2008:259-65](#)
3. Rivaroxaban: An Oral Direct Inhibitor of Factor Xa. American Journal of Health-System Pharmacy. Aug 15 2008. Vol 65 (16). Pgs 1520-1528
4. APS treatment. APS Foundation of America Inc. website <http://www.apsfa.org/aps.htm#7> Accessed Dec. 20, 2008.
5. Dabigatran Etxilate Fact Sheet. Boehringer Ingelheim Inc. website. http://www.boehringer-ingelheim.com/corporate/news/information_packs/documents/bi_vte2008_dabigatran_etxilate_backgrounder.pdf Accessed Dec. 20, 2008.



Patient To Patient: Lovenox® Shot Tips

Written by: Tina Pohlman

You have been handed these shots. I know you are scared. But, I know you can do it. And with these extra tips it should make things go a little smoother for you.

The extra things you will need:

- Flexible style blue ice pack – cold but not frozen
- A quart size zip lock bag to put the cold pack in
- An old pillow case to put the cold pack in to protect your skin from frostbite
- Rubbing alcohol swabs with benzocaine (available at Wal-Mart or Walgreens in the Diabetic Supply section) preferably or rubbing alcohol swabs
- Curad™ Sensitive Skin Spots band aids (And to be on the safe side regular band-aids and waterproof ones in case Murphy's Law likes you.)
- A sharps container.

The main thing I stress is numb your skin before the shot with a cold blue pack. You can also numb your skin after, but numbing your skin before is really key. It does two things. One, it reduces bruising and you won't find yourself running out of bruise free places to inject in a few days. Second, once numbed, it doesn't hurt as bad to give yourself the shot.

With enough time with the blue cold pack on your skin, you won't even feel it too much. It makes it a whole lot easier to do if you are nervous about the shots. When I can't feel my target zone anymore I know I'm ready.

While you are waiting for your skin to get cold, get your bandaid ready and all your other supplies ready to go.

Now for actual shot techniques on giving yourself the Lovenox® shot. As soon as you take off the cold pack, wipe a wide area with rubbing alcohol so it will be dry by the time you inject. You want it to dry because it will sting if the alcohol gets on the needle during the injection. (**Note:** If you are able to find the rubbing alcohol pads with the benzocaine in them, you want it to still be damp. The benzocaine will follow the nee-

dle in and reduce the sting even more.) Then unpackaged the syringe and if you are giving yourself the full dose of the prepackaged syringe pull back just a little on the plunger before pulling off the cap. (Do not twist the cap, it will bend the needle.) This prevents the drip of Lovenox® from forming on the tip of the needle. Air bubbles are ok and preferable. They help distribute the Lovenox® into the fat, so **don't** get rid of the air bubble.

If you have to purge out some because your dose is less than the syringe contains, slowly purge the Lovenox® pointing the needle **down**. I point it into the garbage can that is lined with plastic. The goal is to keep the bubble in there.

Don't touch the needle. You want a dry needle. No Lovenox®, no alcohol. This is because as the needle goes through layers of skin any Lovenox® spilled will cause bruising, alcohol will make it sting. The goal is to get all the Lovenox® in the fat layer UNDER the skin, where there should be no bruising if all goes well. The little air bubble should be the **last** thing that goes in.

Now pick your target zone. I try to avoid place my blue jeans will rub, like my waist and zipper area. Now here comes the part that takes the courage. I promise you after a few shots you will be a pro at this.

Pinch your inch. Some people say to go in fast. I go in as fast as I'm comfortable. The needles are dull straight from the package so you'll have to jab quick initially to break through skin if you have thick skin. Try to get it in the first try because each try after that dulls the needle more, however, if it hurts real bad it's ok to pull out and stick it into another spot that has been numbed and wiped down with alcohol. That's why it's good to wipe down a large target zone. You probably hit a nerve in that spot and there's no sense in continuing if it's going to

hurt. After a while you'll learn which spots are the best for your body and that won't happen as much. Do your best to avoid stretch marks, surgical scars, bruises and those little surface veins.



Don't push the Lovenox® in fast, go at a steady pace and if it starts to sting wait a few seconds before continuing. You want to go slow so your body can absorb the Lovenox® as you inject. If you go fast it tends to spill up the needle shaft opening...going into the layers of skin and causing bruises. Besides, it does us no good there. It needs to be in the fat to work, so take your time. At the end of the shot, count to ten

and then quickly pull out. The counting to ten is to make sure the Lovenox® has had a chance to move away from the needle, preferably being pushed by that last air bubble.

Do not rub the spot. I am finding holding a little pressure with that damp rubber alcohol swab with benzocaine helps keep it from bleeding. If you are bleeding or wearing nice clothes that day, put a Curad Sensitive Skin Spot bandage over the injection site (or a bigger bandaid in case Murphy's Law is in effect). I put one over the injection spot so I don't lose track of where I injected. I call it a place marker. Even if you are not allergic to regular bandages, you want the sensitive skin ones because they come off without pulling. Removing a regular bandage will increase bruising, and if you do end up bruising in this spot will hurt real bad too. The sensitive skin ones just peel off no problem. You may also want to fold over a little part of the band aid for better grip for removal later.

If your injection site is sore or stinging, put the cold pack back on until it stops hurting and make a note to cold pack it longer next time. Otherwise put the cold pack back in the refrigerator and you are all done.

Push out the safety shield and dispose of the syringe in your sharps container.

Relax for a few minutes to let that Lovenox® move away from the injection site.



Sorting Out the “Alphabet Soup” of Antiphospholipid Antibody Syndrome: How Do APS, CAPS, and MAPS Relate?

Written by: Gale A. McCarty MD FACP FACR

Chief of Rheumatology and Immunotherapy/Center for Lupus and APS
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Abbreviations are convenient shorthand for communicating general concepts, but as always, the devil is in the details. At the APS Foundation of America, we try to provide a glossary (dictionary) of terms for quick reference and broad guidelines about different forms of APS as they have evolved. How these syndromes were sorted out represents the work of many dedicated physicians and researchers from various specialties over time.

APS: Antiphospholipid Antibody Syndrome. A syndrome, is a set of symptoms (problems about which a person seeks medical care), signs (findings on a medical exam, and special laboratory test results *that tend to occur together*). Not every patient will have the exact same features, and some of the features are found to be present over a period of time, but may come and go. APS has had many names (or synonyms) over the years, initially being called Anticardiolipin Antibody Syndrome (ACA) because antibodies to cardiolipin (a negatively charged phospholipid) were defined by the aCL ELISA test developed by E Nigel Harris and A Gharavi in London. APS has also been called APLS (APL Syndrome) or ACLS (anticardiolipin syndrome) in some parts of the world.

At a minimum, APS usually means that a patient has: a) a blood clot in an artery, a vein, or a very small set of vessels that connect arteries to veins called capillaries, that can be imaged, and b) a positive laboratory test done by a standardized immunoassay and/or a standardized coagulation-based test. The nature of these tests has been discussed in prior Newsletters to which our readers are referred. Most of the healthcare world thinks of arterial or venous thromboses to start, as clots in these larger vessels are easy to image by ultrasound, arteriography, or venography.

Capillaries are so small that they are hard to image, yet for some APS patients, those areas are where their problems are, giving color changes in fingers, and toes, or hidden problems in body organs. Just as soon as a group decides to develop criteria or guidelines for the identification/diagnosis of a syndrome, most of the actual document written is about exclusions to the criteria. The 1999 Sapporo Criteria, the first set of guidelines, were designed to characterize general features of APS as a minimum “symptom, sign, and special lab test result” set, primarily to be used to obtain similar groups of patients for studies, *not to make the diagnosis in any single given patient*. A look at the Sapporo Criteria shows that the explanations of the definitions and the exclusions (reasons to not make the diagnosis of APS) are longer than the actual criteria. A few years later, the Sydney modification of the

Sapporo Criteria further discussed how things changed, yet in some ways remained the same.

CAPS: Catastrophic APS. Guidelines, like rules, are made to be broken. The late Ronald A. Asherson and others began to notice certain patients with APS had significant symptoms and complications occurring in a relatively short period of time involving several organ systems at once, and associated with an infection, trauma, or stopping medications. These patients sometimes had smaller vessels involved, and sometimes larger vessels. They often did not respond to usual blood-thinning medications used for most people with APS. That the blood clotting was hard to control, and involved multiple vessels quickly led to the name “catastrophic APS or CAPS”, because these patients were in dire straits.



MAPS: Microangiopathic APS. Then there are patients whose features are not covered by APS or CAPS. Syndromes where there are elevated liver enzymes and low platelets in the setting of pregnancy (called HELLP syndrome) or red cells being destroyed (hemolysis) with loss of kidney function (uremia) occur (called hemolytic-uremic syndrome or HUS). These forms don't always fit into the other previously mentioned categories. These patients don't have big blood clots in large arteries or large veins, but more diffuse problems and findings in smaller vessels. They have symptoms related to the effects of aPLs on platelets and the endothelial cells that line blood vessels, effecting changes in them over time that are not always related to one localized blood clot.

These different forms also explain why a patient might have their diagnosis of APS, CAPS, or MAPS made by a variety of different physicians or healthcare providers from differing specialties who consult with each other. Since blood vessels are everywhere in the body, this explains why APS can have signs and symptoms in every organ in the body, and why family practitioners, internists, surgeons, and specialists in all areas of medicine should keep learning about these types of involvement.

References:

- Hoppenstadt D et al: Laboratory Evaluation of aPLs. *Hem Onc Clin N Am* 22: 19-32 '08
- Asherson RA, Pierangeli SA, Cervera R: Editorial: Microangiopathic Syndromes Revisited-New Concepts Related to APS and aPL Antibodies. *J Rheum* 34: 1792-4 '07
- Espinosa G et al: Catastrophic APS (CAPS) and sepsis-a common link? *J Rheum* 34: 923-6 '07
- Wilson WG et al: Int. Consensus Committee Report on Prelim. Criteria for the Diagnosis of APS (The Sapporo Criteria). *Arthritis Rheum* 42: 1309-11 '99
- Miyakis S et al: Int. consensus statement on an update of the classification criteria for definite APS (The Sydney modifications of the Sapporo Criteria). *J Thromb Haemo* 3:1-12 '05



Are There Natural Products That Can Replace Aspirin, Plavix®, and Warfarin (Coumadin®)?

Written by: Elyse Attwood, Pharm D Candidate

Reviewed by: Al Lodwick, RPh, MA

Antiphospholipid syndrome (APS) is the most common acquired clotting disorder. Clots can form in both arteries and veins. They may also form in the placenta causing a miscarriage. There can also be clots in unusual places such as the brain, eyes and kidneys. APS causes changes in platelets, the clotting cascade and the lining of the blood vessels.

When a blood vessel is injured, platelets are activated. They stick to the site of the injury forming a plug that prevents blood from leaking out. Many more steps in the clotting cascade happen that lead to the formation of fibrin. Fibrin strengthens the platelet plug. In APS there can be an over-reaction causing more clots than are necessary.

Aspirin and Plavix® can be used to slow the start of the process by making it harder for platelets to activate. Platelets are most important in starting clots in the arteries where blood flow is fast and under higher pressure. These clots usually result in chest pain, heart attacks, transient ischemic attacks (TIAs) and strokes.

Warfarin (Coumadin®) causes less fibrin to be made. Fibrin starts clots where blood flow is sluggish and under lower pressure than in the arteries. In someone with APS this usually means the veins in the legs.

Sometimes people with APS may wish to use a natural product to replace prescription medicines and aspirin. Ginkgo can slow the action of platelets in starting clots in the arteries. Garlic is another product that is often sold for this effect. In tests in a laboratory

these seem to have some effect. However, they have never been tested against aspirin and Plavix® in people with APS. If you choose to use ginkgo or garlic you may be giving up some of the protection from clots that you get from aspirin or Plavix®.

There are no natural alternatives to warfarin. No herbal product works against the formation of fibrin. You will certainly be giving up almost all protection from clots in the leg veins if you attempt to use a "natural" warfarin replacement.



References

1. Sherer Y, Blank M, and Shoenfeld Y. Antiphospholipid syndrome (APS): Where does it come from? Best Practice and Research Clinical Rheumatology [serial online] 2007 [cited 2008 July 22]; 21(6): [1 screen]. Available from: URL: <http://www.sciencedirect.com>.
2. Current Critical Care Diagnosis & Treatment – 2nd Ed. (2003). [book on CD-Rom]. Jackson: Teton Data Systems; 2003. Based on: Bongard FS, editor. Current Critical Care Diagnosis & Treatment – 2nd Ed. (2003). New York: The McGraw-Hill Companies, Inc.; 2003. STAT!-RefElectronic Medical Library.
3. Klasco RK (Ed): DrugDex® System (electronic version). Thomson Micromedex, Greenwood Village, Colorado, USA. Available at: <http://0-www.thomsonhc.com.library.uchsc.edu:80> (cited: 07/22/2008).
4. Klasco RK (Ed): AltMedDex® System (electronic version). Thomson Micromedex, Greenwood Village, Colorado, USA. Available at: <http://0-www.thomsonhc.com.library.uchsc.edu:80> (cited: 07/22/2008).
5. Burket's Oral Medicine – 11th Ed. (2008). [book on CD-Rom]. Jackson: Teton Data Systems; 2003. Based on: Greenberg MS, Glick M, and Ship JA editors. Burket's Oral Medicine – 11th Ed. (2008). Ontario: BC Decker, Inc.; 2008. STAT!-RefElectronic Medical Library.

Cameron's APS Story

Written by: Cameron Scott

My name is Cameron; I'm 48 years old and was just diagnosed with Primary APS. I am married to a wonderful, helpful and understanding woman and we have a beautiful 13 year old daughter, a 27 year old daughter, an 18 year old son and a 7 year old grandson. We also have 2 cute doggies, a 3 year old Puggle named Blue and a 1 year old Pug named Peanut. We all live in Michigan and are hating the cold weather since I'm originally from California and my wife is from Arizona.



and had 2 different EEGs (the one with the glue then the painful one with the pins), that showed nothing. I have also suffered from aura migraines since childhood.

I have been having problems since I was a kid. I had horrible nose bleeds and had to eventually have the vessels in my nose cauterized. I had small seizure like activity

I believe at the age of 8 I had a stroke because I couldn't speak and my arms and legs were very weak and heavy. I couldn't even lift a hot dog to my mouth. At the time we were on Welfare and therefore didn't go to the doctor that often. My mother's boyfriend who was an alcoholic asked his drinking buddy who was also his doctor about it and he said I was probably just doing it for attention. My attention seeking episode cost me 2 open

heart surgeries to replace my damaged Mitral Valve, which now they are saying was probably an outcome of the undiagnosed APS.

Since then, I've have numerous problems through my life including a stroke at age 26 which was right after my first heart surgery, numerous TIAs and asthma. Three times now I was told I may have Lupus and was always told later I didn't. I guess I have the type of APS with the Lupus-like flares; I even get the butterfly rash across my face. About 4 years ago, I was in a flare and couldn't get out of bed, not even to use the bathroom. I was told I had Mono, however, the test results didn't show Mono. I've had Pneumonia and was coughing up blood. I've had numerous episodes of excruciating chest pain radiating into

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INRs that Fluctuate & Possible Reasons Why

Written by: Stephan Moll, MD

Question: "I have been taking 15 mg Coumadin® a day for nearly a year and my INR was staying relatively o.k. 'till about 2 weeks ago, when it was found to be 1.0. My Coumadin® was increased to 17.5 mg a day and Lovenox® shots added. My next INR had dropped to 0.8. What could be causing this sudden problem?"

Answer: Whenever an INR value is significantly higher or lower than usual in a patient whose INRs have been relatively stable and well controlled, the following reasons should be considered by patient and physician:

1. **Lab error:** Was the out-of-line INR a lab error (significant trouble at the time of blood draw with tissue trauma before blood could be obtained; the blood tube was not filled appropriately)? It may be indicated to repeat the test to confirm that the INR is out of line.
2. **New prescription medication:** Has any new prescription medication been started or has any old medication been discontinued?
3. **Over-the-counter medications:** Is the patient taking any new types of over-the-counter medications, vitamins, herbs, homeopathic medications, weight control pills?
4. **Time of medication intake:** Is the patient taking his/her various medications at the same times as always or are any medications taken closer to the time when the Coumadin® is taken? Some drugs interfere with the absorption of Coumadin® and should therefore not be taken at the same time.
5. **Diet:** Have there been dietary changes that would change the patient's vitamin K intake? In my opinion, a patient should be familiar with the approximate vitamin K content of the foods that he/she eats.
6. **Infection:** Has the patient recently had an infection or diarrhea? Both can increase the INR.
7. **Compliance:** Has the patient really taken his/her medication or has he/she taken too much warfarin? Since various generic warfarin preparations and Coumadin® all look different, the switch from one drug to the other can lead to incorrect medication intake.
8. **Lupus anticoagulant:** Does the patient have a lupus anticoagulant? In some patients the lupus anticoagulant can have an influence on the INR. Since lupus anticoagulant levels can fluctuate over time, the INR can fluctuate as well. Furthermore, if the lab changes its reagents or the INR is tested in different labs, discrepant INR results are possible in some patients with lupus anticoagulants.
9. **Shelf life:** Was the Coumadin® outdated? Efficacy of the drug is only guaranteed for the time printed on the package. The drug may lose efficacy thereafter.
10. **Stress, physical activity:** Has there been an unusual amount of stress, sleep deprivation, or physical activity in the days preceding the INR test? While I am not aware of any published data on this issue, it is possible that in some patients there may be an influence on the INR (increase or decrease), possibly through an influence on the metabolism of Coumadin®.
11. **Generic warfarin:** Could (a) taking generic warfarin, or (b) switching from brand Coumadin® to generic warfarin or vice versa, or (c) switching from one type of generic warfarin to another generic warfarin explain INR fluctuations? Unlikely. Studies indicate that generic warfarin and brand Coumadin® are equally effective and bioequivalent, i.e. for example 5 mg Coumadin® leads to the same INR as 5 mg generic warfarin¹. However, an individual patient assessment is needed, with correlation of INR values to the time of use of generic warfarin or brand Coumadin®, to help clarify whether the fact that a patient is taking generic warfarin may play a role in the INR fluctuations.

Reference:

1. Southern Medical Journal 2001;94:16-21

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my back over and over again that made it unbearable to breath. At the time, my doctor called it Costochondritis, which now I believe were either small numerous PEs or pleurisy.

I have been on warfarin since 1987 for my prosthetic valve and believe that it has saved me from having a severe PE. My current flare has lasted 7 months now and has resulted in another test for Lupus.

In July of 2008 I had another TIA which landed me in the hospital. While in the hospital, a Cardiologist (not mine), asked why I hadn't ever been tested for a clotting disorder since I was having TIAs at a 3.0 INR? He did the necessary tests and that is when I learned that I have a positive Lupus Anticoagulant. I have since been placed on

Aspirin and Plaquenil along with the Warfarin for the clots and Lupus like symptoms and I take other medication for nausea and dizziness. I am now also on Plaquenil, although it was hard to get them to give me this medication. I took an article entitled "Hydroxychloroquine – Everything Old is New Again!" from the APSFA Spring 2007 Newsletter to my doctor. They changed their mind after reading it and prescribed me the much needed medication. I believe this medication has really helped alleviate some, if not all, of my Lupus like symptoms.

Unfortunately my story does not end there. I have just recently been diagnosed with Sjogren's syndrome and have been put on Restasis drops and had to have plugs placed into my tear ducts. I have also been diagnosed with other autoimmune diseases. They are now saying I have mild Lupus and may

also have Autoimmune Hepatitis. However, I have never had a positive ANA, so they are going on my symptoms and other lab work, like low lymphocytes and haptoglobin. I am being considered as one who has ANA Negative Lupus, but then again I have read that people with APS will have a negative ANA.

Even though I have finally been diagnosed it was difficult to get that diagnosis which in turn enabled the disease to then affect my organs permanently. Along with the heart disease I also have Hemolytic Anemia and a damaged liver. I believe that had they diagnosed me properly when all of this started, I would not have heart or liver disease and who knows what else is damaged, since they are really still trying to figure this all out.

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APS Foundation of America, Inc.

Our Mission Statement

Founded in June 2005, the APS Foundation of America, Inc. is dedicated to fostering and facilitating joint efforts in the areas of education, support, research, patient services and public awareness of Antiphospholipid Antibody Syndrome in an effective and ethical manner.

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CafePress—What's In Store For 2009?

If you haven't seen our CafePress store, be sure to check it out! We have many one of a kind Awareness items available for APS, Lupus, Infant Loss, MS, and many other related syndromes. We have new designs and lines in the works for 2009 and there are even a few new items such as travel mugs, pet bowls, and dark colored shirts and sweatshirts! Our CafePress items are high quality and the clothing comes in a variety of sizes from infant to many different adult sizes, including plus sizes and maternity. Many items also come in a variety

of colors. The APSFA gets to keep a small % of each sale from our store when you buy from it, so not only will you get a quality item, but you also make a donation to a worthy cause! Check out our store at the address below and be sure to check back often!!



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