Q & A
By Paul Margulies, M.D., FACE, FACP

Q. My father has been on some form of steroid everyday for the past 20+ years for rheumatoid arthritis. Is it possible to get completely off this drug after so many years?

A. Tapering off long term steroids used to treat chronic illness like rheumatoid arthritis is usually difficult, and is harder with very long term use like this. The first issue is what is the dose of steroid used. If prednisone over 5 mg or Medrol over 4 mg is used for many years, the adrenals have been totally suppressed all that time and are less likely to work again. Another issue is the state of the disease being treated. There is no point in reducing the dose of steroid if the disease is still very active, unless another drug can be substituted. Once the taper is started, if the disease symptoms flare, then the steroid dose must be increased again. Even if tapering is successful, it will take about a year for the adrenal responsiveness to be normal and during that year, steroids will be needed to handle stress. Most of the time, after 20 years of steroid use, a stable replacement regimen like prednisone 5 mg is the easiest solution.

Q. Dear Dr. M, I was diagnosed with secondary pituitary adrenal insufficiency in 2000 after being treated with high doses of prednisone for sudden adult onset asthma. I went to Philadelphia this summer for a second opinion, as my endocrinologist is a very optimistic person and says he can get me off the steroids. He orders blood work every 6-8 weeks and gets a serum 8 a.m. cortisol level. Mine has been under 1 for most of the 9 years. The other endocrinologist ran the ACTH test. I take 7.5 mg of prednisone and he changed me to dexamethone for 5 days for the tests. The results came back as: Beginning cortisol level .4 - after an hour 3.9 - ACTH was <2. He concluded...1. I had Addison’s. 2. I will be steroid dependent for the rest of my life. Do you agree? I want to stop weaning. I have been down to 6 mg of Prednisone a day and .2 of Florinef and felt half decent. The second opinion said my adrenals are shot and weaning now is not an option. Thank you for your time.

A. You do appear to have secondary adrenal insufficiency. The term Addison's disease is reserved for primary adrenal insufficiency, so do not use that term. Since you are unable to taper off steroids over the past 7 years, and the ACTH stimulation test shows an inadequate reserve, I would agree that you should stop trying to taper. It is interesting that you are on Florinef. Most people with secondary adrenal insufficiency have adequate aldosterone production and need only prednisone. If
your pattern included a high potassium level and/or low blood pressure, then you would need to stay on the Florinef.

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A MISUNDERSTANDING AND WEANING SUCCESS STORY

NADF received the following e-mail in reference to a Q & A, originally printed in NADF News®, VOL. XXII, No. 3 • 2007.

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I am writing in response to the Q/A in Dr. Margulies’ column in the last issue of the NADF News. I am extremely disappointed in Dr. M’s response, and feel that he is doing a disservice to those members with secondary insufficiency by providing at least incomplete information, if not information that is just plain inaccurate and not current. I know this from my son Andrew’s experience.

Unfortunately, my son was initially a victim of two endocrinologists that shared Dr. M’s philosophies. They both felt that the results of stim tests proved that he could never recover from secondary adrenal insufficiency (they even went so far as to say that his AI was not caused by asthma meds but must be “idiopathic”). He had three separate stim tests, all with lousy results, ranging from .2-2.

Due to some continuing growth concerns that I was unable to get addressed with our local endocrinologist, I contacted an endocrinologist at Johns Hopkins, who referred me to an endocrinologist at Children’s Hospital in Boston. We took Andrew there for a consultation and it seemed obvious to Dr. Breault that Andrew (age 10 at the time) was a candidate for weaning, despite being on steroids for over half of his life.

I questioned him due to Andrew’s prior poor stim test results, even after being on dex prior to being stim tested. Dr. Breault pointed out that it was absolutely illogical to believe that a patient’s adrenals would begin working within days of being on dex, if they had been “asleep” for years before that. It takes significant time for the HPA axis to “wake up” after not having to do any work for so many years.

Dr. Breault had success weaning other patients, and said that it would take at least 6-12 mos. after being completely off of steroids before a person’s stim test results would be normal. So, we began to wean Andrew very, very slowly.

He remained healthy throughout the entire weaning process.

After 3 mos. of weaning, Andrew’s baseline cortisol was still very disappointing, at under 5. Still, we continued and dropped his dose further, and then did an ACTH stim test after being on only 2.5mg/day for 2mos.

At that time, while his cortisol levels were still below normal, he actually stimmed - his scores on both high dose and low dose more than doubled. Again, not to normal levels, but for the first time, it showed a response from the HPA axis.

We took Andrew off of Cortef® entirely, and then did a repeat stim test 6 months later and guess
what? He had a completely normal low dose and high dose stim test and is no longer an AI patient! If I had believed those first stim tests, Andrew would still be on Cortef today. Lest you think that Andrew is an anomaly, he is not. I am aware of several others like him that have regained adrenal function and endocrinologists at major teaching hospitals (ones in Boston and Philadelphia immediately come to mind) have begun to attempt to wean patients with secondary adrenal insufficiency that has been caused by asthma medication (when asthma is stabilized).

Much of the medical community has come to recognize that a stim test while being on steroids is close to useless. You must be off of steroids for a significant period of time before you can get accurate stim test results, due to the sluggishness of the HPA axis.

I would think that someone like Dr. Margulies, who essentially promotes himself as an expert in matters of adrenal insufficiency, would be aware of that. This is not the first time that I have seen him provide questionable information, and I would appreciate it if he would post a clarification/correction in the next issue of the newsletter for the benefit of your readers with secondary adrenal insufficiency who need to know accurate facts about stim testing, as it pertains to them. Perhaps he could also provide readers with some information about his current research in adrenal insufficiency (I was only able to find his writings for various adrenal insufficiency groups).

RESPONSE FROM DR. MARGULIES

I am delighted that your son was able to successfully wean off his steroids and get back to normal pituitary/adrenal function. I am biased toward achieving that goal in all individuals with a history of adrenal suppression from steroid use. I completely agree that the use of ACTH stimulation testing in this situation is very limited. Unfortunately, there is no useful test to predict who will be able to taper off steroids and get back to normal.

The management of secondary adrenal insufficiency from prolonged steroid use is one the most challenging tasks for a clinical endocrinologist. The ACTH stimulation test is most useful if it does show some stimulation at the onset, and, as in your son’s case, as an indication during the taper that there is some progress in restoring adrenal responsiveness.

The ACTH stimulation test should not be the major factor in making a clinical decision about whether to try to taper steroids. The choice of trying to taper a person off steroids or to find a comfortable maintenance dose for chronic replacement must be individualized.

Generally, the longer use of high dose glucocorticoids, the longer it takes to restore pituitary-adrenal function. A simple guide is that it takes one month for each month of suppression, and about 9-12 months for suppression over one year.

Unfortunately, many people on steroids still have the underlying disease that required high dose steroids in the first place. If the asthma, rheumatoid arthritis, Crohn’s disease or ulcerative colitis is still present, it may flare during any attempt to taper. This is a major limiting factor in a large percentage of patients on steroids. The ideal patient for taper is one who no longer has the underlying disease, especially one who is young and otherwise healthy.

I wish this situation was more common! Most people with prolonged steroid use will experience significant symptoms from their relative adrenal insufficiency as they are tapered and waiting for their adrenal responsiveness to return. These symptoms may include severe fatigue, weakness, loss of
appetite, weight loss, dizziness, depression, anxiety and muscle cramps. Obviously, one’s tolerance for these symptoms will be affected by the person’s age, other medical conditions, psychological state and attitude. If the symptoms are severe, it can often be unbearable, and the attempt to taper must be aborted. Again, young people and those with few complicating medical conditions are more likely to succeed.

Each patient must be an active participant in any decisions about treatment. There is no magic formula for everyone. I believe everyone with adrenal suppression should be evaluated with successful taper as an ideal goal, but not everyone can succeed.

I must point out that you may have been misled about my opinion in the Q and A in the newsletter because the original question was edited. The questioner made it clear that she had been trying to taper off steroids for 7 years!

The fact that she failed to stimulate on ACTH was not the central factor in my advice. I also tried to clarify the misuse of the term Addison’s disease in this situation.

As to my credentials: I am a clinical endocrinologist in practice for 31 years. I do not currently conduct any basic research, but I do teach medical students, residents and fellows.

My responses to questions brought to the NADF are voluntary and meant to give general information about adrenal diseases. My answers are never meant to be definitive consultations. Everyone should discuss any response with their own doctor.

The latest word, dated 9/20/09, from Dr. Margulies:

"There is no good formula for weaning that applies to everyone. It is such an individual experience. The most important aspects are discussed in the Q and A answers in NADF’s Weaning Compilation hand-out. Dialog with the endocrinologist is the key."

The National Adrenal Diseases Foundation is a non-profit organization providing information, education and support to all persons affected by adrenal disease. For more information on joining NADF, or to find a support group in your area, contact:

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