Cushing's Support & Research Foundation
Winter/Spring 2018

cushing's newsletter

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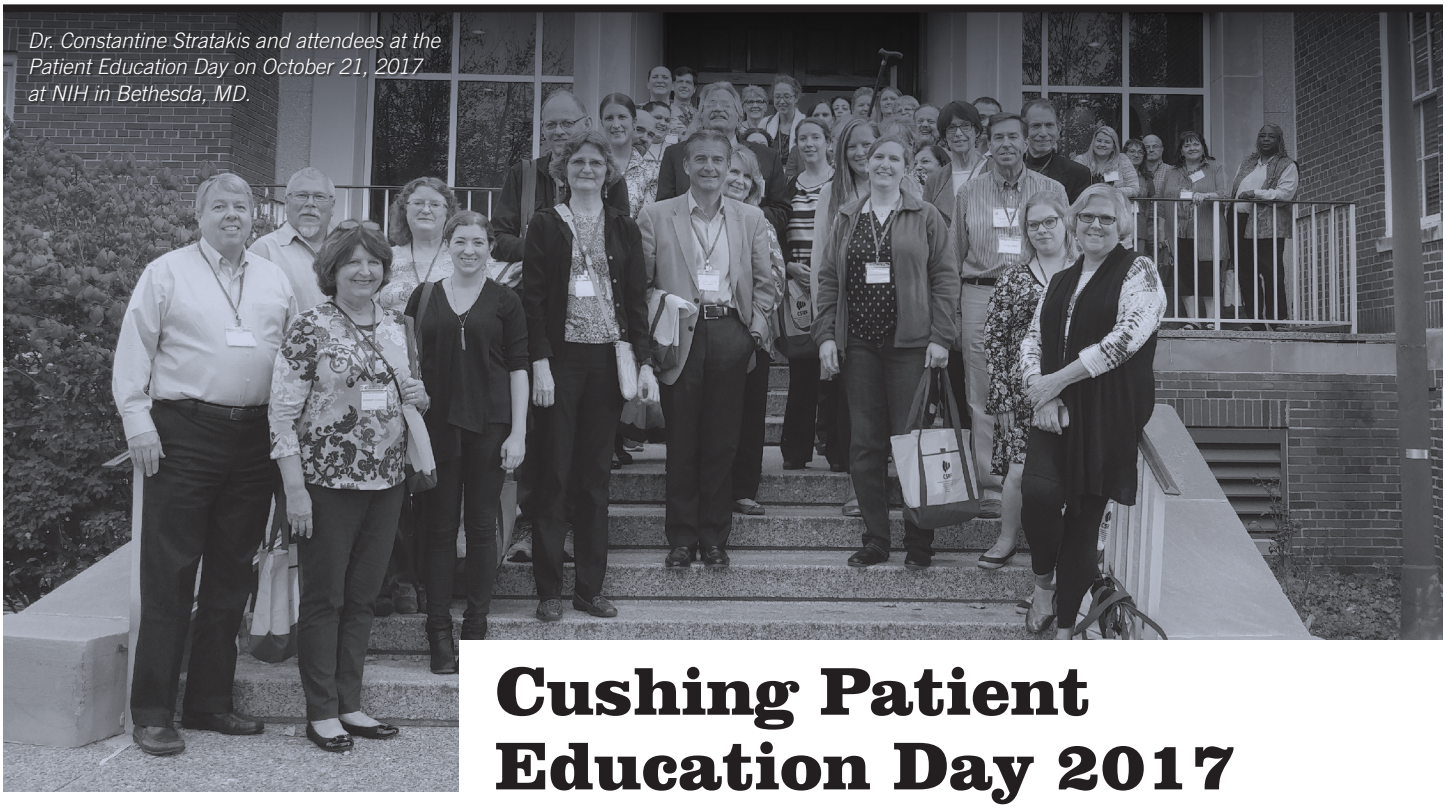

Dr. Constantine Stratakis and attendees at the Patient Education Day on October 21, 2017 at NIH in Bethesda, MD.

Cushing Patient Education Day 2017

By Dr. Meg Keil

Over 100 attendees gathered on a gorgeous fall day in October 2017 for the CSRF Cushing's Patient Education Day at the National Institutes of Health in Bethesda, MD. This educational event was made possible by our generous sponsors: The Eunice Kennedy Shriver National Institute of Child Health and Human Development (NIH), Corcept Therapeutics, Strongbridge Biopharma, and HRA Pharma. We are grateful for the wonderful support of our sponsors that allowed the conference to be held in a beautiful facility, provided an evening reception, offered delicious food during the conference breaks, and provided travel stipends to attendees. In addition, our sponsors made it possible for the conference to be video cast! (We did not videotape the afternoon breakout sessions to allow privacy for discussion).

To view the conference, go to this link:
https://videocast.nih.gov/summary.asp?Live=26564&bhcp=1

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AFFILIATIONS

American Diabetes Association (ADA)
The Endocrine Society
European Society of Endocrinology
EURODIS (Rare Diseases Europe)
Global Genes
National Organization of Rare Disorders (NORD)
The Pituitary Society
World Association of Pituitary Organizations (WAPO)

FOLLOW THE CSRF ON TWITTER!

We will be tweeting whenever new content is added to our website or there is breaking news. To follow us, sign up at:

https://twitter.com/CSRFnet

Message from the (new) President

In 2011, I had no idea what Cushing's was and wouldn't have been able to point out a pituitary gland on a diagram of a brain if asked. A year later, Cushing's became the only thing on my mind as I went through the diagnosis and surgery process… for the first time. My story is similar to many I have heard over the years from patients who weren't fortunate to reach long-term remission after one surgery. As of the time of the printing of this newsletter, I am in remission for the second time and beginning to balance hormone levels again. The last six years have been a real roller coaster, to put it mildly!

I first found CSRF in 2013, ironically *after* my surgeries. I still wonder to this day how I didn't find it when I researched information at the time of diagnosis the previous year. I sent a message and soon was on a long phone call with Founder Louise Pace. After that call I realized that I wanted to be more involved than I was with just my local support group, so I started doing tasks for CSRF and catching glimpses into the methods of a surprisingly small, all-volunteer Board.

I am honored to carry this title, and I am excited to dedicate my time and skills to furthering the mission of helping patients navigate all aspects of Cushing's. I look forward to the personal growth I will experience on this new journey, and I hope that I can be a helpful resource to anyone who needs it.

We have several new initiatives in the planning stages that we are eager to roll out later this year, so stay tuned! We like a vocal membership and encourage you to share your questions, thoughts, suggestions, and recommendations with us for things like newsletter articles, social media content, webinars, and doctor's answers.

Thank you for your support of the CSRF!
Leslie Edwin
President

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The topics for the Cushing's Patient Education Day were selected based on the feedback from our members from a prior online survey and included presentations by a variety of experts. Prior to the start of the meeting, attendees enjoyed a wonderful buffet breakfast and then DMV group member Eugenia Tyner-Dawson welcomed everyone to the meeting. Dr. Constantine Stratakis from NICHD started off the day with an overview about the diagnosis of Cushing's syndrome, which was helpful to allow attendees to be familiar with the terminology that was discussed throughout the day. Dr. Stratakis addressed many interesting questions from the audience. Next, Dr. Roberto Salvatori from Johns Hopkins University provided a comprehensive update about treatments, medical therapies and clinical trials for Cushing's syndrome. Then, Dr. Mary Lee Vance from University of Virginia spoke about long-term physical effects of Cushing's syndrome, the impact on quality of life, and the importance of anticipatory guidance to encourage patients to have realistic expectations about the recovery period. Dr. Jitske Tiemensma gave a fascinating presentation about quality of life and illness perception and discussed various tests that are used to evaluate cognitive function, memory, and executive function. Next, we listened to a panel of six patients share their stories about Cushing's; it was very interesting to learn about the similarities and differences in their journeys to diagnose, treat, and recover from Cushing's syndrome and the audience had many questions for the panelists.

After a delicious buffet lunch, Lisa Felber (LGSW) and Dawn Herring (LMFT) presented "Coping with Recovery and Life Changes," and discussed the challenges patients with Cushing's syndrome face during their journey from diagnosis to recovery. They shared experiences that are familiar to Cushing's patients and the importance of self-empowerment, honoring your feelings, and self-care. Also, they addressed the importance of keeping a perspective and discussed ways to develop support and coping strategies.

In the afternoon, attendees could choose from a variety of breakout sessions. The topics included Sexual Intimacy and Fertility Concerns (Dr. Alan Decherney and Dawn Herring, LMFT), Family/Caregiver Issues and Concerns (Lisa Felber, LGSW), Coping with Chronic Illness (Dr. Pedro Martinez, Dr Tiemensma), Nutrition and Exercise (Megan Shoenfeld, RD), Managing Adrenal Insufficiency (Dr. Meg Keil, Raven McGlotten, MS, RN)., What's New in Cushing's Research (Dr. Constantine Stratakis), and Pheochromocytoma (Dr. Karel Pacak). Many thanks to the presenters of the breakout sessions.

Last, but not least, a big shout out to the conference organizers, especially Leslie Edwin, whose tireless efforts produced spectacular results. Also thanks to those who assisted Leslie to organize the conference: Karen Campbell, Sabrina Whitt, Stacy Hardy, Eugenia Tyner-Dawson, Meggan Monroe, Joanie Kilbride, Meg Keil, Shianne Lombard-Treman, and Renee Dorsey.

Many patients gave feedback that they would like to see more in-depth presentations of some of the topics covered that take into consideration more specific aspects of dealing with or recovering from Cushing's. The CSRF Board met shortly after the event to discuss these suggestions and requests, and we are very excited to begin some new programs and offerings in 2018.

News and Updates

CSRF Attends NORD Rare Diseases and Orphan Products Breakthrough Summit 2017 in Washington, D.C.

By Leslie Edwin

In October 2017 the National Organization for Rare Disorders (NORD) held their annual meeting to gather patient group representatives, the U.S. Food and Drug Administration (FDA), researchers, doctors, and others involved in the rare disease community. NORD has been advocating for those with rare diseases for almost 35 years now; the organization began as a coalition of patients and families living with rare conditions in the late 70s and early 80s. There are about 7000 rare diseases currently identified, and fewer than 500 — approximately 5% — have FDA-approved treatments.

NORD is a 100% patient-driven 501c3 with membership and volunteers totaling more than 3500 individuals. There are over 1200 patient groups for rare diseases, and NORD operates in all 50 U.S. states and 30 other countries. NORD is fully funded through donations and providing services and educational programs. CSRF is a member of NORD.

Cushing's is covered under NORD's Patient Assistance Program (PAP). We encourage anyone having difficulty paying for treatment or organizing logistics of care to check with them to see if resources might be available to help. For more information, visit www.rarediseases.org and follow the tab "for Patients and Families" to the PAP section.

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THE ORPHAN DRUG ACT AND RECENT PASSAGE OF THE TAX ACT

One of the first major accomplishments of NORD was their key role in the passage of the Orphan Drug Act of 1983. Listening to presentations from the FDA and NORD today makes you realize how huge the machine is that works to navigate laws, patient needs, clinical trials, research, and drug approvals. At the individual level, a Cushing's patient, for example, might experience "sticker shock" when they hear the unsubsidized price of a drug, then relief when they end up with just a $25 co-pay. There's a complicated story behind those two pieces of data though, starting back with the Orphan Drug Act. A medicine might be vital to help your body manage cortisol while you wait for surgery or radiation, but you are one of a very small pool of total rare disease patients who will need that medication. Compared to common drugs taken by millions of people like diabetes drug metformin or anxiety treatment alprazolam, the ratio of total product cost (research and development, marketing, etc.) to affected patients is staggeringly large for a rare disease. The median cost per year per patient for an orphan drug is $32,880, and the average approximate number of patients on an orphan drug is 750.

We Cushing's patients have a limited number of medications available to us now, but the future for drug development for all rare diseases took a hit when the final tax act was passed in December 2017. The House wanted to completely gut the Orphan Drug Tax Credit (ODTC) program, while the Senate wanted to slash it by half. The ODTC was part of the Affordable Care Act of 2010. Prior to the current administration's budget decisions, groups developing treatments for rare diseases could claim a tax credit for up to 50% of their qualified clinical testing expenses. The final bill reduced that by half to 25%. CSRF was one of more than 200 patient groups who signed on to a statement issued by NORD opposing this cut to a program that has led to a wave of new therapies and treatments for rare diseases over the last seven years. This is huge because about one third of all new drugs approved by the FDA now are for rare diseases; in 2015, a hefty 47% were so.

PUT A FACE ON IT!

NORD encourages individual patient organizations to get involved with researchers and clinicians who are working on aspects of their particular disease focus. Building relationships goes a long way for the benefit of all parties; we are the end users of their work, and information shared in both directions increases understanding and produces better long-term outcomes. Therefore, we try to "put a face on it" — to humanize the disease experience and effect of the clinical work. A medical student might be more meaningfully impacted during a 10 minute patient presentation than from an hour-long lecture on data and scientific method.

There has historically been a bit of a separation between those who are looking for a cure or fix and those who are looking at quality of life (QoL) issues. We patients have long known that both are vitally important! Ongoing improvements should involve a cohesive unit of medical and surgical options, QoL considerations, and the patient experience. CSRF strives to provide the latter as frequently as possible, exhibiting several times a year at endocrine and other related conferences. We are encouraged by the feedback we receive at these events, as it seems that more and more doctors are seeing Cushing's patients and following accepted guidelines for diagnosis.

PCORI AND THE FDA

Congress established the Patient-Centered Outcomes Research Trust Fund (PCOR Trust Fund) in 2010 as part of the Affordable Care Act. This fund receives income from three sources: appropriations from the general fund of the Treasury (currently $150 million annually), transfers from the Centers for Medicare and Medicaid trust funds, and a fee assessed on private insurance and self-insured health plans (the "PCOR fee"). The Patient-Centered Outcomes Research Institute (PCORI) was then formed to administer these funds as an independent, non-profit, non-governmental organization. PCORI directs money to research looking at the effectiveness of two or more proven treatments and measuring their benefits in the real-world population. All stakeholders, including patients and caregivers, are vital to the success of these research programs. As of October 2017, PCORI has awarded $80 million to fund 27 research studies related to rare diseases. Cushing's has not yet been the focus of a PCORI study, but who knows what the future holds.

The FDA makes a substantial effort to get input from patients by offering multiple "access routes" like Twitter (FDA_Patient_Net), Facebook, and their Patient Network and Newsletter sent out twice a month (subscribe at http://go.usa.gov/xKzwb). The FDA works closely with NORD and seems to have embraced the value of patient input on rare disease issues relatively early. Just as many doctors are beginning to address patients' neuropsychological needs as part of their standard of care, these "big hitters" in the rare disease community are beginning to focus on the vital role we patients must play in decisions that will affect our long-term health and possibly also our short-term survival.

RARE PATIENT JOURNEY DATA TELLS THE STORY

According to NORD, on average, the diagnosis of rare conditions requires two to three misdiagnoses and visits to seven or eight doctors. It takes an average of five years to get a diagnosis, and even then many are still not fully accurate.

There are more Americans living with a rare disease than HIV, heart disease, and stroke combined. One in 10 Americans has a rare disease which equates to approximately 25-30 million people in this country; more than half of those people are children. The official definition of a rare disease in the US is "any disease, disorder, illness or condition affecting fewer than 200,000 people".

NORD has the motto "Alone we are rare, together we are strong." Our combined voices are changing the landscape of treatments and policy, and we encourage participation in academic research, surveys, and other outreach from professionals working to find solutions to some of the problems we face with a rare disease.

HOW YOU CAN GET INVOLVED

Involvement starts with educating oneself on the issues that are important to them. Follow trails of articles and research studies on Cushing's, individual symptoms, or whatever else interests you. NORD and the FDA both have very detailed websites geared to patient access.

If you are interested in looking at the legislative process where you live, participating in events, and connecting with other rare disease patients and advocates, NORD has the Rare Action Network for your consideration in all 50 U.S. states. Most of these groups are fairly new so if you have organizational or other skills to offer, that might be a good place for them. You can find more information at www.rareaction.org.

From time to time CSRF will partner with researchers to share surveys or other questionnaires; as this newsletter goes to press, Dr. Jitske Tiemensma and her team at UC Merced are finishing up a survey on QoL that was conducted with CSRF membership. We look forward to sharing the results with you, knowing that all the data came from patients who recognize the importance of lending their input to those hoping to change the future of Cushing's treatment.

Author's Note:

I have been attending meetings related to legislation, policy, and non-profit organization on behalf of CSRF for several years now. I'm always blown away by the magnitude of information out there and the highly qualified and immensely intelligent people working at some of these agencies. What we hear about healthcare in the news, online, and in our communities is negative and frustrating most of the time, but there are a lot of people working to bring about change. This should lead to exponentially better outcomes for all patients in a much shorter time than it has taken to get to this point so far.

All references in this article came from websites and conference slides, notes, and handouts from NORD, FDA, and PCORI.

Federation of International Nurses in Endocrinology (F.I.N.E)

Chris Yedinak DNP,FNP,MN

www.EndocrineNursesFederation.com

F.I.N.E. is a worldwide collaboration of endocrine nurse clinicians and researchers that evolved out of the need to share experiences, information and data regarding best evidence-based practice for improving patient outcomes. Our organizational goal is to provide an accessible global network of nurses in endocrinology who promote optimal health outcomes for persons of all ages who are living with endocrine disorders. Membership is available to all nurses in any sub specialty of endocrinology who work with either adult or pediatric patient populations, worldwide. F.I.N.E. now has 21 member countries and continues to grow.

Nurses focus on the needs and perspectives of the patient and family experiencing a disease or disorder. This may be in the form of direct care for an immediate clinical care need or clinical research to answer questions about the common experiences that may lead to more or better treatments. These research questions may include: What it means to live with endocrine dysfunction; detailing the struggle to achieve an acceptable quality of life; identifying the barriers to accessing care (particularly long term care); identifying current gaps in clinical care; identifying the challenges of developing and adhering to a treatment regimen; defining the meaning of a 'successful' outcome for the person/family affected by specific diseases/disorders etc. Nurses strive to provide patients with updated knowledge regarding specific diseases/disorders and options in treatment. F.I.N.E is a forum for nurses and patients to partner in order to bring new insight and ideas to helping individuals negotiate a path to 'best outcome' throughout the changes occurring during the life cycle as they deal with such problems as pituitary diseases and disorders.

Many more people are diagnosed with an endocrine disorder yearly. Diabetes Mellitus has become a worldwide epidemic, and other endocrine disorders are either increasing in prevalence or are more frequently diagnosed based on better diagnostic tools and better access to information, such as via the internet and conferences. Through the efforts of research, knowledge is predicted to double every 73 days by 2020. Taken together, these factors, along with

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the escalating cost of care, apply pressure for effective utilization of resources, better collaboration and information sharing among all health care providers and a focus on what has the greatest impact on the patient's ability to engage in a productive, satisfying life.

The Federation of International Nurses in Endocrinology (F.I.N.E) aims to promote excellence in clinical care and advance the science and art of endocrine nursing throughout the world by: Providing a platform for interdisciplinary collaboration, research, education and distribution of emerging knowledge in endocrine disorders; raising global public awareness of the contribution of nurses practicing in endocrinology; supporting the development of organizations of endocrine nurses in all countries; developing. Standards of Practice for nurse management of endocrine disorders; laying the groundwork for international certification of nurses practicing in endocrinology; providing a forum for collaboration with those affected by endocrine disorders plus other clinicians and researchers in related fields of endocrinology.

Currently, there are numerous initiatives in progress by F.I.N.E. members, including: Improvements in dynamic testing (Julie Hetherington. NP, Australia), functional responses to disease (Raven McGlotten FNP, USA), responses to treatment- endocrine side effects (Cecelia Follin PhD, Sweden); quality of life (Sofia Llahana PhD, UK), treatment modalities for Adrenal Insufficiency, (Phillip Yeoh, NP, UK), Medication Adherence issues & hypogonadism (Andrew Dwyer PhD, Switzerland/USA), plus continuity & care transitions (Karen Liebert BSN, USA). Many more investigations are ongoing by other members.

The first scientific meeting of F.I.N.E. as an organization was held in Beijing China during September 2016, with the support of the International Congress of Endocrinology (ICE). This meeting included presentation of research by members from China, UK, Vietnam, Australia, USA and Belgium. The next collaborative meeting, the 18th International Conference of Endocrinology (ICE)/Nurse Meeting, will be held in Cape Town, South Africa, 1-4 December 2018, with proposed presentations from local World Alliance of Pituitary Organizations (WAPO) members, clinical practitioners and researchers.

Partnership with patient organizations in research and program development is essential to achieving meaningful investigations and, ultimately, more effective care that simultaneously expands the reach of care. We are fortunate to be able to partner with WAPO and individual member organizations worldwide. We anticipate working together to develop many more research initiatives for mutual goals in the future. (Note: CSRF is a member of WAPO)

References and Samples of Member Publications

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On-line Tools for Symptom Tracking

Symptom tracking can be useful for gauging your recovery or determining when to test if cyclic Cushing's is suspected. A symptom tracking application from Corcept, maker of Korlym, is available here: *www.cushingsconnection.com*

Doctor's Articles

What's New in Cushing's Research? A Conversation with Dr. Constantine Stratakis of the NICHD

Dr. Stratakis gave one of the most highly rated presentations at the October 2017 Patient Education Day at the NIH in Bethesda, MD, when he discussed new strategies, research, and outcomes for Cushing's in his breakout session. The positive feedback led to our desire to share a version of this presentation in the newsletter. What follows is a Q&A based on comments from some of the patients who attended.

Q: What kind of information is out there for long-term survivors of Cushing's Disease / Syndrome? There are patients now who have been post-surgery for 10, 20, 30, even 40 years. Is there any new research on quality of life, long-term effects of hypercortisolism, or living long-term without adrenals or with hydrocortisone dependence?

A: There is quite a bit of information about quality of life (QoL) in Cushing's syndrome (CS) after treatment. A lot of unknowns also remain. In brief, we know that life for patients with CS is not easy with or without cure. Children with CS (Keil M. J Clin Endocrinol Metab. 2013 Jul;98(7):2667-78) suffer through deterioration in academic performance after their cure from CS for the first few years post-operatively. Most children find it difficult to cope, even when they are young adults; we described a few of these patients that attempted suicide, despite being cured from CS for many years (Keil M et al. Pediatrics. 2016 Apr;137(4)). Adults with CS suffer similarly and there are several recent publications on this issue (Pivonello R et al.. Front Neurosci. 2015 Apr 20;9:129). Adults report fatigue and executive dysfunction even after cure (Papakokkinou E et al. Behav Neurol. 2015;2015:173653).

Although other patients with pituitary tumors experience similar problems, the issues of patients with CS stand out (Romijn JA. Nat Rev Endocrinol. 2016 Sep;12(9):547-56). There is a lot of new knowledge on what QoL is like for patients with CS which did not exist even a decade ago. Patients now do not have to deal with the ignorance and disbelief of previous years, as there is good documentation in the literature. But, of course, lots of questions remain: now that we know that QoL is impaired in patients with CS even after complete cure of their hypercortisolemia, we do not know why this is the case. Is it because cortisol caused some permanent damage in the neuronal circuits regulating specific brain functions? Or, is it all reversible albeit after many, many years? There are no studies looking at 30-40 years after cure for CS. We just started a long-term study at the NIH that will attempt to study patients as long as 10 years after cure.

Finally, it is unclear whether all the effects are neuronal. Does the body change in a way that is irreversible if one has CS? This is much harder to answer, as muscle mass, body strength, bone structure, and skeletal function all change with aging; it would require, therefore, the follow-up of many, many patients for a number of years and their respective age-adjusted controls to answer this question.

Q: What types of advances in genetics are you seeing for Cushing's? How prevalent are genetics in pituitary vs. adrenal?

A: I have been studying the genetics of all forms of CS for the last 25 years. I am happy to report that what I was saying in the 1990s and sounded almost crazy back then, is now universally accepted: CS is frequently a genetic disease - not necessarily inherited but genetic, in the sense that it is due to mutations of specific genes (see a review by Lacroix et al Lancet 2015;386:913-927). This is all very good news: knowing the genes that cause pituitary or adrenal tumors that lead to CS may assist in the development of new medical therapies that will be far more effective than the currently available ones. There is really no difference between pituitary and adrenal tumors in terms of how frequently they can be genetically caused: for example, 50% of all pituitary tumors causing CS harbor mutations in the USP8 gene and 50% of all adrenal adenomas leading to CS have mutations in the PRKACA gene. There are other genes involved in predisposition to CS in forms of the disease that are inherited. These are much rarer in both the pituitary and the adrenal gland. Multiple endocrine neoplasia-type 1 (MEN1), Carney complex (CNC) and related diseases are some examples of these inherited conditions that may also lead to CS among other conditions.

Q: What kinds of advancements are you seeing in imaging for this disease?

A: Currently, we rely on magnetic resonance imaging (MRI) and computed tomography (CT) for most lesions of the pituitary and the adrenal, respectively. The technology is clearly suboptimal, especially for pituitary tumors. MRI is simply not enough, knowing that it can miss as many as one third of all pituitary tumors causing CS. We hope that the developments in genetics will lead to the discovery of probes that may be used for imaging and allow for the earlier identification of these tumors. Likewise, in the adrenal gland, we need imaging modalities that are safe and allow for the differentiation of the various functions of adrenal lesions, distinguishing cortisol-, aldosterone- and androgen-producing lesions from non-secreting ones.

Q: There's lots of chatter about dental problems caused by Cushing's. What kind of research do you see happening in this area?

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A: CS affects immunity and almost certainly the microbes that we all host in our body. Thus, I would not be surprised if patients with CS experience dental issues and/or increased severity of periodontal disease. There is paucity of data on this issue.

Q: Is there anything specific being done to try to reduce the statistic that approximately 50% of tumors come back? How about the one where there is a 50% chance of getting a tumor somewhere else in the body after Cushing's?

A: The best way of preventing a recurrence is being seen early and at an experienced medical center. For example, patients with pituitary tumors seen at the NIH have a less than 10% recurrence rate. This is because we go to an extreme length in diagnosing the right cause of CS in our patients the first time we see them. And then, a very experienced surgeon operates on them. Most recurrences of pituitary tumors are due to incomplete excision by an inexperienced surgeon. A great number of cases are due to the wrong diagnosis: if a tumor is not in the pituitary gland, temporary remission may occur after pituitary surgery, but CS will come back because a corticotropin-producing tumor may be elsewhere in the body. Finally, most benign adrenocortical tumors should be completely excised by an experienced endocrine surgeon. However, ectopic adrenocortical tissue can be left behind by an inexperienced surgeon.

Q: One patient recalls that you said that cortisol is so vital to the body that going without it for a very short period of time can lead to death. This is obviously a huge deal for those of us who are adrenally insufficient or headed toward that because of radiation or other factors. Do you have additional details to expand on this point?

A: My point is that patients that do not have adequate cortisol secretion face a serious problem, one that is life-threatening. Fortunately, for patients with pituitary tumors this is temporary and adrenocortical insufficiency (AI) only lasts for up to 12-18 months. For patients with single adrenocortical tumors, AI after surgery also lasts for about 18 months, on average. However, patients that had bilateral adrenalectomy face a life-long need for cortisol replacement. AI also complicates the management of patients on adrenolytic medications or those on Korlym. Finally, permanent AI may develop in patients with pituitary tumors after complicated or multiple pituitary surgeries.

AI can lead to acute hypoglycemia, hypotension, or severe electrolyte abnormalities that, yes, can cause death in a few minutes. At the NIH, we spend a lot of time training our patients on signs of AI, recognizing and treating it, and preparing for its recognition by emergency medical services if the latter are called in the case of emergency. Fortunately, like patients with diabetes and on insulin, patients with AI also learn to live with their disease, prevent crises, and treat their needs, in a way that allows them to live full and healthy and productive lives.

Q: There are standard tests that most everyone is familiar with now — the standard physical symptoms, blood, urine, saliva, dexamethasone suppression test… but you mention a "squat test." What is that, and what are some other similar tests that may not be commonly listed but that are good tests for Cushing's?

A: There are a few physical signs of CS that the average physician is not familiar with. All physicians know the extreme central obesity and thinning of the extremities, the full face, the stretch marks, the easy bruising, fatigue, hair loss and the overall appearance of the "classic" patient with CS. However, there are some subtler signs that appear early in the course of hypercortisolemia: weakness of the central body muscles that becomes only detectable by asking the patient to squat and get up without support. Most healthy adults are capable of completing this simple exercise; however, patients with CS are unable to complete it successfully even if the disease is in its early phases. Other soft and early signs of hypercortisolemia include soft, non-terminal hair in the face and elsewhere that can be felt rather than seen, fullness in the supraclavicular and/or temporal areas of the neck and head, respectively, brought upon from local fat accumulation, and facial plethora (redness).

Q: Any final thoughts?

A: The good news is that CS is a disease that today can be cured. The goal should be to diagnose it early, correctly, and offer the proper treatment. As we find more and more about what causes CS including USP8 and PRKACA mutations in pituitary and adrenal tumors respectively, new diagnostic testing and therapies will be designed; in the future, every tumor causing CS will be genotyped and the information will be useful in defining treatment and prognosis.

In the meantime, we have to make sure that patients with CS do not suffer from the many side effects of hypercortisolemia that go beyond what we know as CS. For example, patients with CS can develop severe infections, sometimes lethal ones; these can be prevented by implementing measures in patients with CS such as those used in other diseases that compromise the immune system. Fractures and/or other effects on the myoskeletal system should also be prevented or dealt with early and with appropriate physical therapy. Hypertension should be treated; and so on…

All of the above can be taken care and offered at experienced centers where both diagnoses and therapies and multidisciplinary care are offered by experienced teams of doctors and other caretakers. The goal is to turn CS into yet another disorder that is totally treatable; our patients should be able to live full and healthy lives, as medicine and technology advance.

Thank you, Dr. Stratakis!

Why can Cortisol Make us Obese?

Professor Onno C. Meijer, PhD, Einthoven Laboratory, Leiden University Medical Center, Leiden, The Netherlands

Cushing's Syndrome is what happens when we are exposed to (way) too much cortisol, or its synthetic variants. Out of the many unwanted aspects of Cushing's, fat accumulation is the most visible one, and also one of the biggest risks for additional health problems. The medical jargon talks — perhaps rather callously — about 'truncal obesity', 'moon face', and 'buffalo hump'. The latter refers to the accumulation of fat in between the shoulders. This is such a typical feature of cortisol excess that, when scientists were able to generate an animal model for Cushing's Disease, they prominently displayed a picture of a mouse-with-buffalo hump in the article that described their work(1). Why does cortisol lead to the appearance of all this fat?

The answer is related to the normal role of cortisol as a stress hormone. When we encounter a stressful situation, we need to somehow cope with that situation — we get 'stressed'. Being stressed means being in a particular 'state', not only mentally, but also physically. Although stress has a bad reputation, the stress response is essential for survival, as all organisms will meet challenges in their lives that require this state of being stressed.

The body has very effective ways of bringing about physical states: hormones. Hormones are messenger molecules that come from one source — glands, such as the adrenal gland. The power of hormones lies in the fact that they are transported via the blood to reach each and every cell in the body. They are 'public announcements', and as such a perfect way to bring about a coordinated change in the activity of many organs. They can cause a different state.

Immediately at the start of the stress response, the hormone adrenaline (a.k.a. epinephrine) is released to support the 'fight or flight' response. In other words: adrenaline prepares the body for active coping with the stressful situation. Adrenaline for example 'tells' the liver to release sugar and fat into the blood, which can be burned by muscles, to actually make us able to fight or flight. But this effect of adrenaline is relatively short lasting. Enter: cortisol. Cortisol is our second major stress hormone, slower, but ever so powerful. Its levels in the blood typically will start to increase after a couple of minutes. Cortisol levels can remain elevated for days or weeks when stress is chronic. One of the functions of cortisol is to support a sustained stress response.

Cortisol acts slower than adrenaline but in a more profound manner. Like adrenaline, cortisol tells the liver to spit out its stored sugar and fat as food for muscles, but it also tells the liver to make new sugar and fat. While both hormones tell the liver to make energy available, cortisol is the one to replenish the liver energy stores. What happens under the influence of cortisol is that the liver actually uses all the resources it can find and converts these into sugar and fat for the other organs to use. In parallel, cortisol causes breakdown of muscles that are not being used, in order to generate building blocks for fat and sugar production in the liver(2). Cortisol is thus an effective coordinator.(3)

These cortisol effects can be really useful for different stress situations. For actual fight-flight we need the sugar and fat. If the stressor would be starvation, it is even useful to break down muscles to ensure activity of the vital organs. However, in case of Cushing's syndrome, the patient is normally at rest and does not need all this extra energy. Yet: it is there in the blood. What happens next?

The body cannot do many things with sugar and fat circulating in the blood. In fact, with fat it can do only two things. It can burn it (normally into calories, that is: fuel for our muscles and other organs). Or it can store it: in our fat depots. With sugar there is one more option: the body can turn it into fat. To burn, or to store. And so, what cortisol does here is to ensure that more than plenty of fat is present in the blood, even by breaking down muscles. That fat has nowhere to go other than into its depots: and here we arrive at obesity. And, for that matter, at thinning of arms and legs, as muscles are actively being told to shrink and return their building blocks to generate 'fuel'. A nasty consequence of what normally is a healthy response to adapt to stressors.

What we do not know is why fat accumulates in the face and in between the shoulders. It may be that the latter involves a sort of reactivation of a special fat organ: the brown fat (also known as Brown Adipose Tissue, or BAT). This is a special type of tissue that takes up fat, but does not store it. BAT rather burns fat into heat. BAT is

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present in large amounts in babies: they have a relatively large surface and therefore lose heat more than adults. The major location of BAT in babies is right in between the shoulders. The bad thing is that the buffalo hump may represent 'awakened' BAT tissue, but this BAT tissue only stores the fat and does not burn it (otherwise there would be no accumulation)(4).

And so, the unpleasant effect of cortisol on fat accumulation does make sense. The increase in glucose in parallel increases the risk to develop diabetes. The friend that cortisol normally is, turns into a foe: it is really too much of a good thing.

1 Sahut-Barnola I, de Joussineau C, Val P, Lambert-Langlais S, Damon C, Lefrançois-Martinez A-M, Pointud J-C, Marceau G, Sapin V, Tissier F, Ragazzon B, Bertherat J, Kirschner LS, Stratakis CA, Martinez A. Cushing's syndrome and fetal features resurgence in adrenal cortex-specific Prkar1a knockout mice. PLoS Genet 2010;6(6):e1000980. doi:10.1371/journal.pgen.1000980.

2 This breaking down of muscles is called a 'catabolic' action. Androgens, the male sex hormones, do the opposite and support muscle growth — these are then the anabolic steroids.

3 One of the other organs that is part of this same coordination is the brain, that starts to generate hunger signals under the influence of cortisol. Although in case of acute threats this effect is counteracted by other stress factors in the brain. This is why stressed people may either eat more or less than normally.

4 Adults do have some active brown fat tissue, located more inside the body. Activating this BAT is presently a popular subject in research on obesity. Activated BAT will simply take up fat from the blood and make it disappear as heat (and some CO2 and water). A sure (but limited) way to lose calories.

Editor's Note: Dr. Meijer was a featured speaker at the 2017 Annual Summit of WAPO, the World Alliance of Pituitary Organizations, of which CSRF is a founding member. This article originally appeared in the December 2017 Global Pituitary Voice, the WAPO newsletter.

Doctor's Answers

All questions answered by:
Adriana G. Ioachimescu, MD, PhD, FACE
Associate Professor of Medicine and Neurosurgery
Co-Director, The Emory Pituitary Center
Emory University School of Medicine, Atlanta, GA

Q: I've heard people talk about having testing for 17-OH and DHEA in their pursuit for a diagnosis. I had my first round with Cushing's five years ago and only remember cortisol tests. What are these and how do they help diagnose Cushing's?

A: DHEA is a low-potency sex steroid produced by the adrenal gland. The best way to evaluate its production is by measurement of dehydroepiandrosterone-sulfate (DHEA-S). Measurement of dehydroepiandrosterone-sulfate (DHEA-S) may assist in diagnosis of Cushing's syndrome in patients with lower than normal or low-normal ACTH levels. DHEA-S is usually lower than normal in patients with adrenal Cushing's caused by a unilateral adrenal adenoma. DHEA-S can be elevated in patients with adrenal lesions that secrete both cortisol and androgens. Finally, most patients with pituitary Cushing's (also called Cushing's disease) have normal DHEA-S levels.

Q: There seems to be some controversy about Cyclic Cushing's — some doctors don't even believe in it. Many patients get frustrated because they "chase highs" hoping to test during an active period and, in a worst case scenario, spend many months or even years getting worse while trying to test only during periods when they don't feel good and associate that with high cortisol. Do you have any advice for these patients?

A: Cyclic Cushing's syndrome is very rare. Patients have episodic cushingoid manifestations (weight gain, easy bruising, increased blood pressure or glucose, muscle weakness or fatigue) associated with abnormal cortisol testing. The episodes usually last for several weeks. In between episodes, patients do not have these manifestations and cortisol tests are normal. Making the diagnosis is difficult and requires multiple measurements of 24-hour urine cortisol and bedtime salivary cortisol. If patients do not feel good or feel their health deteriorates despite normal cortisol testing, other causes should be investigated.

Q: I've heard that some doctors are starting to do neuropsychological testing during Cushing's and then during remission to help the patient understand how cognitive, memory, and other brain-related operations are affected by excess cortisol. Can any psychology department conduct this testing or would I need to seek out a special-

ist if my endocrinologist doesn't have a partnership with a neuropsychology department? How are doctors using the results of this type of testing to help patients?

A: Cortisol receptors are present in the brain and cortisol excess can cause a wide array of mood and cognitive changes. Neuropsychological testing is important in patients with Cushing's syndrome who experience these problems especially since some problems may persist to some extent after cortisol normalizes. The neuropsychological evaluation is done by trained neuropsychologists. Some pituitary centers have collaborations with neuropsychologists from the same institution. If this is not available, patients may see a neurologist or psychiatrist who can provide further guidance and referrals.

Neuropsychological results are useful in several ways: 1. accurate diagnosis of the mood and cognitive problems and their severity, 2. guidance regarding management (which may include observation, counseling, seeing a psychiatrist, etc) and 3. tracking the progress by repeat evaluations.

Q: Do you have any statistics on incidentalomas? I've heard that as many as one in five people have one, and I often wonder if some patients with several of the typical Cushing's symptoms get scanned and find a tumor on the pituitary or adrenal – how could they determine conclusively that the tumor is non-active / non-secreting when it presents on an MRI and they have physical symptoms of Cushing's?

A: Pituitary incidentalomas are found in approximately 15% of the general population upon undergoing MRI imaging. Most of these lesions are small cysts or adenomas and do not associate a high cortisol problem. However, once a pituitary incidentaloma is detected, the patient must undergo a clinical and laboratory evaluation by an endocrinologist. If patient has manifestations of high cortisol, appropriate testing will be ordered. In addition, imaging surveillance of the pituitary lesion is necessary.

Adrenal incidentalomas are detected with increasing frequency over our life span (up to 30%). They can be nonfunctioning adenomas, adenomas that secrete cortisol, cysts, pheochromocytomas, primary adrenal cancers, metastases and other. Similar with pituitary incidentalomas, patients benefit from endocrinological evaluation that will include specific cortisol testing.

Q: I recently attended a patient education day and there were several people in the room who were slim. The majority of us are still carrying excess weight, usually in the mid-section, regardless of where we are with active Cushing's or in remission. I know I am more active than it seems possible to still be carrying this weight. Do you have any research that addresses this situation? Only one of the slender patients I spoke with has an extremely active lifestyle – the rest sound more or less on par with everyone else when it comes to physical activity and diet.

A: Losing weight after normalization of cortisol level is not always an easy process. While some patients lose a significant amount of weight in the first few months after remission, others do not experience the same course. A thorough evaluation of other causes is necessary which may include, among other, insulin resistance and thyroid dysfunction. In addition, most patients who achieve remission after surgery require glucocorticoid replacement, usually hydrocortisone. Taking the physiologic amount is important and should be discussed at each endocrine visit. It is always a good idea to increase physical exercise as tolerated and to have a healthy diet. If desired results are not achieved, consultation with a weight loss specialist is the next step.

Prescription Assistance

There are many prescription drug cards that can be found by searching the internet. The CSRF does not endorse any specific card. One that has recently come to our attention is UNA Rx card which can be used nationwide. More information is available at *http://www.unarxcard.com/index.php*

Rx Hope is another program with discounted prescription pricing: *https://www.rxhope.com/*

PAN has programs for some of the medications specific to Cushing's including Korlym and Signifor: *https://www.panfoundation.org/*

NORD also provides help with medication costs as well as travel and other medical expenses: *http://rarediseases.org/for-patients-and-families/help-access-medications/patient-assistance-programs/*

Call: 1-800-999-6673 x 326
Email: Cushings@rarediseases.org

Service Dogs for Cushing's and Adrenal Insufficiency Patients

By Amy Dahm

CSRF member and Cushing's survivor Amy Dahm, accompanied by her service dog, Sam, co-facilitated an informal discussion with renowned dog trainer Bill Creasy about service dogs for Cushing's and patients with adrenal insufficiency (AI) as a side meeting to the National Cushing's Patient Education Day in Bethesda, MD on Friday, October 20, 2017. Bill Creasy has over 40 years' experience training dogs and is the current Chair of the International Association of Canine Professionals (IACP) Service Dog Committee. The goals of the presentation were to inform the audience, examine the current landscape for Cushing's and AI service dogs, and to eventually create a replicable, reliable protocol for training adrenal insufficiency dogs.

Ms. Dahm defined an adrenal insufficiency dog and presented an overview of the Americans with Disabilities Act (ADA) and legal protections for service dogs and their owners. She defined an Adrenal Insufficiency Dog (AID) as a dog trained to perform specific tasks which can detect changes in cortisol in the body and alert a patient when the patient is heading into medical crisis. Under the ADA, a service animal is defined as a dog that has been individually trained to do work or perform tasks for an individual with a disability. Service providers are only allowed to ask two questions: 1) Does the owner have a disability?, and 2) Does the dog assist the owner with that disability? The tasks performed must relate to the disability. She compared service vs. therapy vs. emotional support dogs, and noted that the category of service animals is the only category that has legal protection under the ADA, although some airlines and housing boards do recognize emotional support animals (ESAs). State law also comes into play. For example, for service dogs in training, trainers must follow state law, not the ADA.

Although the interest is keen throughout the Cushing's and AI communities, at this point, efforts have been one-off and it is difficult to obtain a reliably trained AID. Traditionally, there are three ways to obtain a service dog: 1) from a professional trainer and/or specialty organization, 2) to train one yourself, or 3) to have the dog alert naturally. Although there is an online group dedicated to training service dogs for "Cushies" (Cushing's patients), at this point in time, there is no recognized, replicable training protocol for adrenal insufficiency dogs in the United States. There are at least a few vetted AI service dogs in Europe. According to experienced Diabetic Alert Dog (DAD) trainer Becky Causey, the standard she recommends for training is that the trainer has trained a service dog focusing on that specific disability that has successfully alerted over the course of one year. Mounting interest from the Cushing's and AI communities, coupled by a burgeoning awareness by experienced dog trainers, is leading to a new push for developing a reliable source of trained medical alert dogs.

BENEFITS OF AIDS

The potential benefits of developing a reliable, large pool of AIDs are many. Medical alert dogs serve as an additional disease management tool for those patients dealing with the day-to-day management of a very tricky disease where there are very few tools available. Until extremely recently, there was no way for patients to check their cortisol levels on an immediate basis – and the accuracy and authenticity of the iphone meter is still unknown long-term and it is expensive and not commonly available (it is also not FDA-approved). A reliable medical alert dog can provide an alert to a patient when heading into crisis, buying the patient more time to take more steroids, self-inject, or head to the ER for treatment. If a patient loses consciousness during crisis, a service dog can alert bystanders. A trained dog can fetch extra steroids if the owner becomes incapacitated, and has the extra benefit of making an "invisible" disability more visible, accepted, and accessible.

Renowned trainer Bill Creasy has been studying the mechanics of Cushing's and adrenal insufficiency and is planning on applying his refined training and scentwork methods to an attempt to train a new AI dog starting in Spring 2018. He estimates that, if successful, it will take 18 months to train the puppy to be a fully functional, public access AI service dog. He hopes to use this dog as a template for proof-of-concept and for developing a replicable protocol for training other AI dogs.

CHALLENGES

There are several challenges facing efforts to develop a replicable, reliable protocol. Cortisol is not easily measured, and every "Addie" (patient with Addison's) has a different cortisol "cliff" at which point they begin to nosedive into an adrenal crisis. This is cutting edge science. It's not clear that all Addies emit a "scent" when going into crisis.

THE SERVICE DOG INDUSTRY

Per renowned trainer Becky Causey, a potential service dog owner should practice "buyer beware." The service dog industry is not regulated, is highly fragmented, and there is no one standard. A trained service dog, for example a diabetic alert dog, can cost anywhere from $15,000 on the low end to $25,000-30,000 on the high end, depending on the skill level and seniority of the trainer. Many illnesses have organizations dedicated to funding and helping train service dogs geared towards the needs of patients with those particular illnesses; Cushies and Addies do not.

VETTING A SERVICE DOG TRAINER

Per Becky Causey, these are the questions you want to pose before spending thousands of dollars and hundreds of training hours on a service dog charged with assisting in life-threatening situations:

- How many Addison's or Cushing's dogs have you successfully trained? And how long have they successfully been working with their owners?
- What is the motivation for training a dog with this skill? Personal first-hand knowledge of disease, personal interest, or paycheck?
- Does the trainer understand the mechanics of the disease?
- Is the trainer training towards the dog providing helpful tasks or preventing a life-threatening emergency?

Amy and Sam's Story

After developing post-operative adrenal insufficiency after my ULA in 2014, my beloved dog, Sam, spontaneously alerted on me one night when I was home alone on the couch with a raging cortisol headache. Sam is a Cyprus poodle, and he was a hunter with a strong sense of smell. I didn't feel well, and he alerted — he issued a piercing shriek, he jumped on the couch, and he started pawing at my hair and nudging me. I took steroids, and he calmed down. Puzzled, I lay back down and rested. The next day, after googling and finding a story for an Addison's dog in the UK and realizing that he might have been alerting on me, I wanted to learn more about service dogs and make Sam legit. I lived in mortal fear of having an adrenal crisis, lived by myself, and had problems with my balance and frequently dropped things.

Despite my best efforts, including researching online, consulting my vet, and asking the online groups, I hit a brick wall. I couldn't tell what my local service dog laws were, where I could get Sam properly trained, or what the standards were. One night I was home watching TV and a news segment came on about a renowned service dog trainer. I called her the next day and attended a service dog workshop geared towards service dog owners an hour outside DC. At this workshop, she tested Sam and other dogs, educated us on the basic tenets of the ADA, and introduced me to other trainers, including Becky Causey. The final day of the workshop the dogs rode public transportation and did a check-out in a nearby town, and Sam passed with flying colors. Both Sam and I were exhausted, and would take two hour naps when we got back to the hotel. By the end, he would not even look at me — his journey from a pampered pet to a full-on service dog was not an easy one for him.

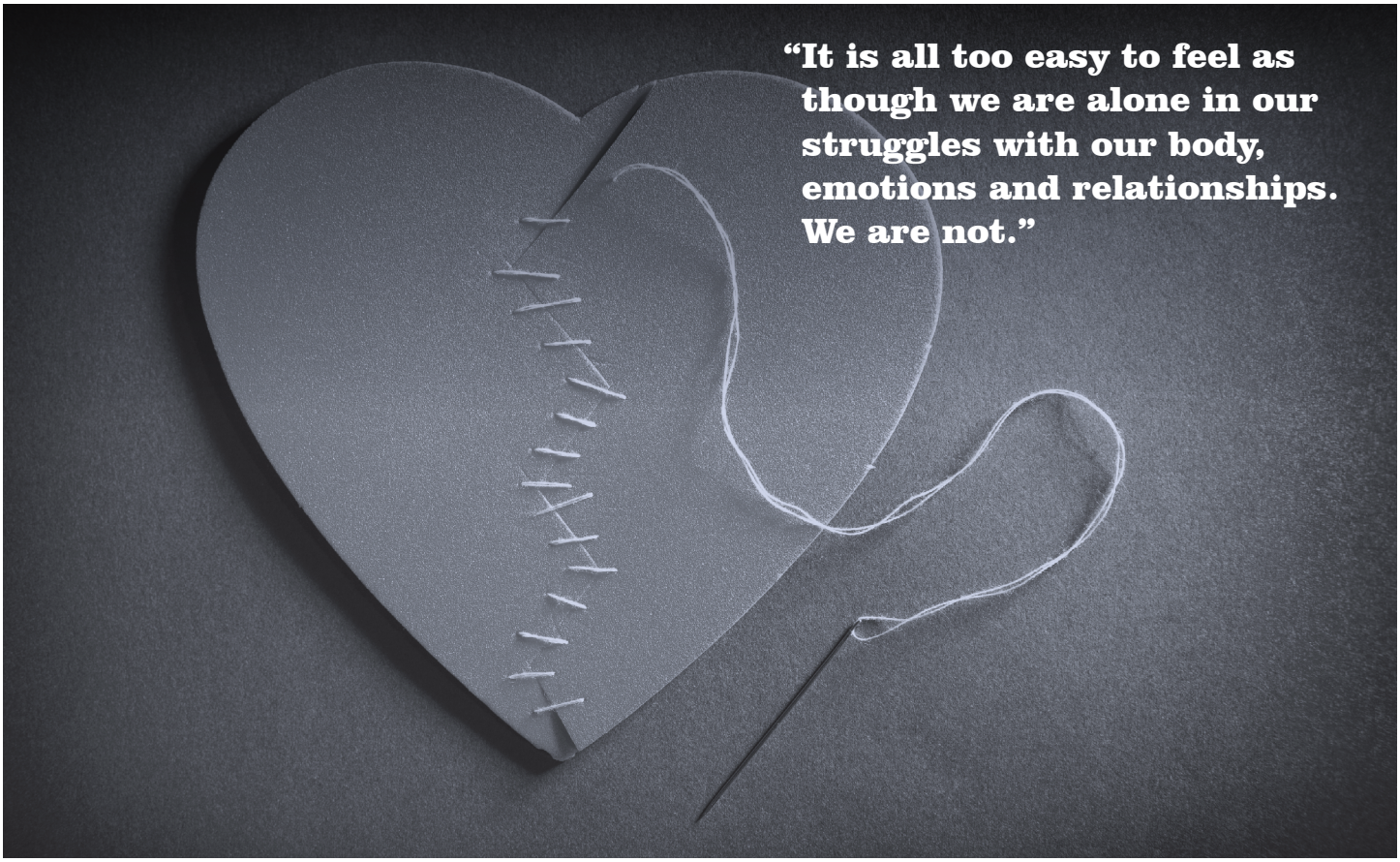
Over the next few months, my boyfriend and I would take Sam and practice what we had been trained to do. We slowly exposed him to an increasing amount of stimuli in varying controlled environments, and gave him attention when he behaved well (and cut off attention when he didn't). He hated the service vest, and chafed at wearing it. Sam, who was very social, was upset that his meet and greets were sharply curtailed. We visited restaurants, attended lectures, and took him to other people's houses in an effort to expose him to additional stimuli. I sought out empty daytime movie theaters to see how he would react to the big screen.

The question remained whether his initial alert was a fluke or if he could repeat it. Eventually, we received the answer when he alerted on me again. Although I didn't feel in crisis, per se, I headed to the ER, where the doctors discovered I had a serious infection that was impacting my cortisol.

My boyfriend and I took Sam to our first meeting of the DC Chapter of the CSRF Support Group in 2015 and he became the unofficial mascot. Although many people recognized he was a social and polite dog, many people were suspicious because he was not the standard issue Labrador service dog. I became tired of explaining over and over again how he was a service dog and educating various business owners about what they could legally ask us. To be sure I was operating within the law, I checked in with the service dog coordinator for DC, who assured us we were. Over time, Sam and I fused as a unit. We became more comfortable walking in stride together, obeying commands, and working as a team.

Sam and I went together to Capitol Hill for Rare Disease Week. While preparing to medically retire, Sam accompanied me to my seminars at the Foreign Service Institute, where U.S. diplomats go for training.

Sam continues to support me, and we hope to use our relationship as a framework for developing a protocol for medical dogs for Cushing's and AI patients.



Sex and Cushings

For many, having a sexual relationship seems critical to life satisfaction. From a purely pragmatic viewpoint, sex *is* critical for life itself. Over the centuries, people have ended and began relationships for sex, married to have sex, killed over sex and taken remarkable risks where sex is concerned. Sex has long inspired the creation of music and poetry. Even the Bible has the Song of Solomon. Cushing's Disease appears to hijack this very life force, leaving us to feel anger, loss and/or apathy toward sex. Many Cushing's patients have incurred relational losses because of the challenges faced sexually and intimately. We often experience self-loathing, anxiety and depression due to physical and psychological changes. Research points to the complicated variables that exist when it comes to attraction, libido and sexual relationships. We know a complex interplay exists among various biochemical, psychological, relational, religious, familial and cultural components. This article will focus on the Three C's of Sex: Compassion, Communication and Collaboration.

This article by no means will crack the code on such a complex subject, however, the objective is to normalize the complexity of our feelings toward sex, our bodies, and relationships, and offer tangible tips to support our recovery in this area. This article will not address sex from a spiritual stand point as I believe that is better left to clergy. That being said, nothing in this article is meant to offend anyone's specific values. Please feel free to take what is useful and leave the rest.

COMPASSION

The type of compassion I am referring to is self-compassion. It is all too easy to feel as though we are alone in our struggles with our body, emotions and relationships. We are not. As other patients have shared their various challenges, it has become clear that we often suffer from a sense of isolation, self-loathing and frustration. Some have lost relationships due to physical and mental changes. Some still want to have sex until they are having it. Some don't want to until they are. Some feel too weak or are in too much pain. Some have libido and can enjoy sex but are not able to orgasm. Many report that their body and mind can't quite agree. Some desire sex but can't stop the racing thoughts. Others feel too tired to even consider the act. And then there is the chronic pain and joint and muscle deterioration. Sex is, after all, a physical act. Can we be patient with ourselves, curious rather than condemning, observing our feelings, thoughts and sensations without judgment — just noticing? From this place, our self-compassion may grow.

Tip #1: Listen to our internal thoughts. Would we talk to a friend as we talk to ourselves about our bodies? Try to talk more gently to

yourself. We are healing, changing, little by little, daily, weekly. We have control over how we talk to ourselves and what we communicate to our bodies at a cellular level.

Tip #2: Listen to Kristen Neff's self-compassion meditations at www.self-compassion.org.

Tip #3: Explore various forms of touch. Try regular massage. Most of all, try in little ways to embrace your body, note its resilience and its attempts to balance despite the cortisol levels. Some people report pain or a tickle-ish sensitivity upon touch, sexual or otherwise. Experiment with various forms of touch from firm to light to see if your body has a preference. Perhaps only a hand massage or foot massage feels good. Consider a pedicure. Visualize the healing taking place at a cellular level. Bathe in epsom salts with a favorite essential oil. Hug a friend. Cuddle a pet. Remember to daily nurture your body in little ways to fight the self-loathing, disgust and physical pain. The goal is to embrace rather than ostracize our bodies while they fight for our wellness with everything they have got. The self-compassion we find will fuel our ability to have conversations on our behalf with partners and the medical community. Your cells are always listening to everything you say, even in your head. Speak to them as you would a close friend or family member.

COMMUNICATION

Many of us come from families in which sex was not talked about. I remember as a pre-teen hearing the word "masturbation" and asking my mother what it meant. Her reply? "Look it up in your Funk & Wagnalls". So I did. Needless to say, I didn't understand the definition I read but I somehow knew not to return to her for greater clarification! Therapists have long urged couples to engage in a more open dialogue regarding sexual needs, preferences and boundaries. This is challenging for a number of reasons. We may not want to disappoint or offend our partner, may not feel entitled to having our sexual needs met, may not believe pleasure is OK, and frankly may not want to say we have no interest. Once we work on our self-compassion, we will be in a better place to imagine having any conversation with our partner.

Much of sexual desire begins in the brain. Did you know that quadriplegics can orgasm? True. Clearly, that's primarily a brain induced experience. It is the perception of the experience that got them there. Both men and women have reported waking up in the middle of an orgasm. Clearly, this is without help from anything but the mind at play, on its own. The communication happening inside our head can help or harm us. Women are also known for multitasking mentally during sex. For instance, many women with no chronic illness report that they struggle to focus only on sex, during sex. They can be creating the grocery list, contemplating the next day's agenda, all while trying to be present during sex. Men have their own pressures to perform and the inability to fake an orgasm (not that faking orgasm is the goal here). So, once we infuse our self talk with compassion, we can now begin imagining what it is we would like to say to our partner, our doctor, a friend, about our needs or feelings toward sex, our bodies, our losses, our hopes and dreams. Cushing's seems to affect everyone a little differently so your experience is your own. Only you can communicate that to another. No one else will have had quite the same symptoms and challenges.

In our breakout session on sex at the 2017 Patient Education Day at NIH, I referred to sex as feeling like a fiery torch entering my vagina. Because I was comfortable saying this out loud, I was able to share this with my partner as well as my doctor and dozens of you! Doing so brought relief and the ability to problem solve this so that I was no longer frustrated and incapacitated in this way. I also had to communicate about the limitations of having two total hip replacements. Unfortunately, my orthopedic surgeon was unable to give me the same flexibility I once had. Now, not everyone is going to be comfortable conveying these things to a partner, yet, the power in doing so is in the potential for the partner to have a better understanding and the opportunity to not take our changes, ambivalence or disinterest in sex personally. This gives the couple the freedom to be more creative, exploring what works and what doesn't. For some, just being able to have sex with the lights off is enough. But to be able to convey that need can be challenging. I encourage you to do so. In addition, it is important to acknowledge that to make ourselves this vulnerable, we must believe we will feel heard and respected, received compassionately. Sex is at its best when it is light and playful, connecting, not a chore. The oxytocin released from any kind of safe and wanted touch, including sex and orgasm, is bonding. It somehow makes the sky bluer and the grass greener, at least for a day. It can make a couple feel closer immediately.

To partners out there, I invite you to open up a dialogue with your loved one. Ask openly what they need, want, don't want. Be available to hear a sense of hopelessness and grief. You don't have to fix it. It's not about you. Yes, it affects you. Remember that the emotions in the moment are not permanent. The anxiety and depression are most likely a result of the disease and are also treatable. Ask if there is anything you can do that does feel good to your partner, sexual or otherwise. Maybe a foot massage, cuddling on the couch. Not personalizing the lack of interest and the emotional ups and downs is key to preventing your own burn out and sense of hopelessness or powerlessness. Seek professional support if needed. When partners stand by helplessly, asking what they can do to help, I remind them, the prescription is in the listening. It is not your job to fix, but to listen, non-judgmentally. In so doing, you are making your partner the expert in his/her own experience. For many Cushing's patients, being heard accurately has been a challenge. This is your area in

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which to shine. Nobody knows your partner like you do. No one else can listen quite like you do. It may feel like you are doing very little but, after all, to be seen and heard accurately is essential to all.

Tip#1: Armed with self-compassion, begin to talk with your partner, using "I" statements. Follow the "I" with a feeling word. The reason for the "I" is that people hear us better when our sentences start with it. When we start a sentence with "you", for example, people tend to feel immediately defensive or put off. Our goal is to create a safe space in which to share our thoughts, feelings and needs. Use the exercise below as an example if it is difficult to imagine talking about this. There is no right or wrong. You are entitled to your feelings, your thoughts, your preferences and your needs. If you have not had any discussion about this, then it may feel quite awkward. To introduce the conversation, you may say something like:

"I've been thinking about my feelings toward sex and want to talk with you. Is now a good time?"

Any sentence with the word "sex" in it has a good chance of grabbing your partner's attention! If they agree that now is a good time to talk, you can continue with the below. Some people like to write out and contemplate first what they are wanting to say. Some couples keep a journal. Feel free to do this if it helps you form your thoughts and clarify your feelings. If single, I encourage journaling of this sort to give you an opportunity to explore further your own thoughts and feelings in a non-judgmental space.

I feel_____ when I think about sex.
i.e. "I feel confused and sad when I think about sex."

I wish_____.
i.e. "I wish I felt clearer, excited."

I miss_____.
i.e. "I miss wanting sex, how connected it used to make us."

I still like (or appreciate)_____.
i.e. "I still like and appreciate cuddling with you, your hugs, the way you look at me."

If you could _____, I'd feel_____.
i.e. "If you could continue to be patient with me, I'd feel less anxious and guilty."

For a free list of emotions, see the website www.cnvc.org. They give a list of feeling words to use when our needs are met and when they are not.

Tip #2: Watch the following:

1. Jason Headley's video on YouTube, "It's Not About the Nail."
2. Mark Gungor's video on YouTube "A Tale of Two Brains".

COLLABORATION

Collaboration in the healing of our body is best done with a team. I had the listening ear of my Endocrinologist, Acupuncturist, Naturopath and Ob-gyn. Find your team. Once we can identify our goal as it relates to sex and sexual intimacy, we will know with whom to talk. The "fiery torch" feeling? My Ob-gyn. The lack of libido? My Naturopath and Acupuncturist. The intimacy issues? A couples' therapist. Any hormone replacement I had, I cleared through my Endocrinologist. More times than I can count, I have wanted all of these professionals sitting around a table to discuss my care. In the absence of that, I formed my own team, apprising all of them of any changes made to my care. My Acupuncturist was there from the beginning of my symptoms, through diagnosis, an ovary removal, two hip replacements and transsphenoidal surgery. His focus was constantly two-fold: assisting my body in finding its own balance (while believing it wants that), and challenging me to challenge my perceptions and self-talk. He would say, "I can balance your body, but I am not in charge of your perceptions". He assisted me in not allowing the disease to become my identity. He taught me greater self compassion by example. He challenged me when I would say, "I have Cushing's". He would say, "Do you? Be careful how closely you bring this to you". He offered up instead that I was experiencing an imbalance that was treatable. It was not allowed to "move in" and become my identity. He also challenged me to consider masturbation if I couldn't imagine sex. His justification? To remind the body what it is capable of, its natural birth right, and to gently nudge the hormones to remember this dance. He referred to this as "yoga" and told me my homework was to do my "yoga" 2-3 times a week. The comfort I found in being able to have candid conversations was

priceless. I could say to him, "I don't feel like it." He would reply, with a slight smile and a raised eyebrow, "That's not the point." So, I encourage all of us to collaborate with providers that we trust, with whom we feel seen and heard accurately and who listen to us and allow us to be the experts of our own experience.

Tip #1: Define your goals as they relate to sex and intimacy. This will inform the collaborative team you choose. Do you want to want to have sex? No libido? Body image issues? Feeling disconnected with your partner and wanting to feel reconnected? Considering dating again? Vaginal atrophy? Unable to maintain an erection? Define goals and find a team.

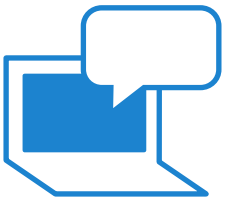
Tip #2: Seek referrals. Ask your Endocrinologist, friends and family. Sometimes, word of mouth referrals feel the best.

Tip #3: Stay connected with other Cushing's patients and survivors and the CSRF newsletter and website and through a support group.

In sum, Sex & Cushing's is a complex topic. I have attempted to normalize the complexity of our feelings toward sex and experiences with sex while giving tips to continue the conversation and aid in our recovery and the reclamation of our full selves. Yes, this is a process that takes time, years in fact.

I encourage everyone's feedback on this article, requests for further topics, conversations, information. Maybe in the future we can ask various components of our healing team to weigh in on these issues. In the meantime, with all the love I have, I invite everyone to continue to focus on the three C's of sex: Self-Compassion, Communication and Collaboration.

My Best to you all as we continue this healing journey together,
Dawn Herring, LMFT
Cushing's Survivor



From the CSRF website:

Coping With Cushing's: Tips for Caretakers

If you are helping a loved one to deal with Cushing's, you have taken on a big job. Cushing's is complicated, difficult, and frightening. Your loved one may be unable to do things that were once easy for her. He may be physically weak and exhausted. She may be anxious, sometimes to the point of panic, or too depressed to get out of bed. He may be forgetful, or burst into tears over something that seems minor to you. She may be self-conscious about her appearance, and reluctant to participate in social activities.

You may have more responsibilities at home as your loved one is unable to keep up with normal household tasks. Your time may be filled with endless doctor's appointments and the stress of wondering what is going to happen next. Medical expenses, traveling to out-of-town doctors and taking time off work create financial hardships. Most of all, you may be afraid that something terrible is happening to someone you love.

It may sound counter-intuitive, but one of your first responsibilities is to take good care of yourself. Your loved one may not be able to support you as she did before, and if you fall apart, you're no use to either of you. Be sure to eat well and get plenty of rest.

Offer to attend medical appointments with him. Confusion is a common symptom of Cushing's, and the diagnosis and testing are complicated. Many patients appreciate having someone with them who can take notes or ask questions, and most doctors will welcome your being there to support their patient.

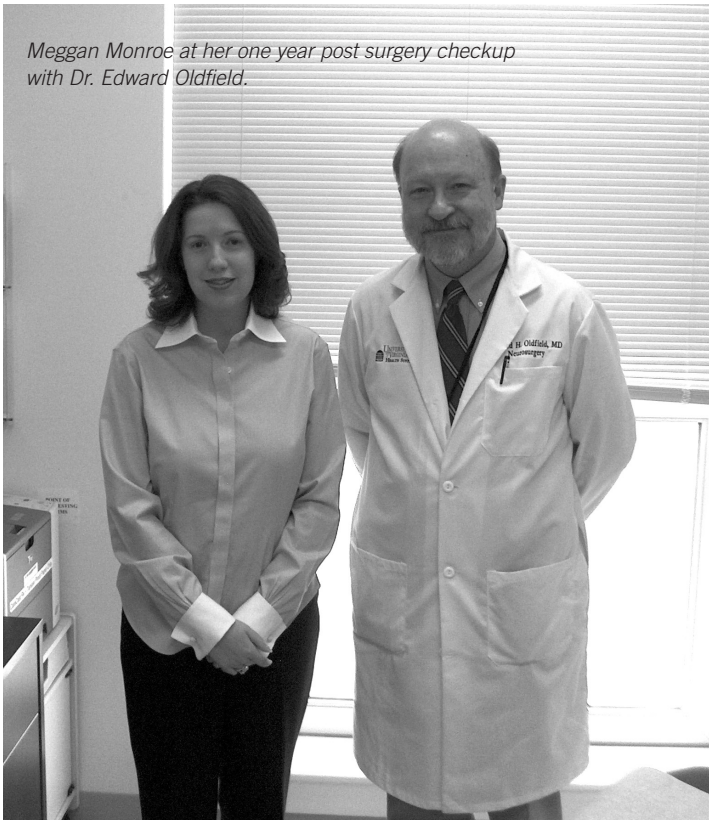
Get informed! Read about Cushing's, ask questions, and talk to other patients and caregivers. The more you understand about Cushing's, the less alarmed you will be by changes in your loved one's appearance and behavior. Be familiar with her medications, learn to recognize any emergencies such as an adrenal crisis, and know what to do if one happens.

Ask for help. Often friends and family want to lend a hand, but don't know what to do. Ask someone to bring dinner, run some errands, take the kids to the movies, mow the lawn — whatever tasks you are finding overwhelming.

Talk to someone about how you are feeling. It may be a friend, a therapist, clergy, or anyone else who will listen and understand.

And most of all, keep your sense of humor and perspective, and don't lose sight of the fact that although it can take years, with proper treatment most Cushing's patients are able to return to a normal, healthy life.

This article is published on the CSRF website:
https://csrf.net/coping-with-cushings/tips-for-caretakers/



Meggan Monroe at her one year post surgery checkup with Dr. Edward Oldfield.

By Meggan Monroe

When we say someone is a "rock star," they are a legend, a person that people idolize and want to emulate. The "rock star" label can often imply that the person also has a boisterous spirit and a big ego.

I had just a few encounters with the "rock star" of my life. He was undoubtedly an expert in his field, idolized for his skills and knowledge, and a person that people wanted to emulate. But his ego was nonexistent. He was humble, unassuming and empathetic to me, a desperate 29-year-old seeking someone who could find (and remove) my pituitary tumor.

My rock star is Dr. Ed Oldfield.

Dr. Oldfield had been at the University of Virginia for less than a year when I met him. His partner in treating me was Dr. Mary Lee Vance, a lover of bulldogs and quite a "bulldog" herself in her demand for excellence.

I was at the end of my rope in my Cushing's journey. While the lab results clearly pointed to Cushing's, my MRI (at a very prestigious academic medical center) showed a "normal" pituitary. I had spent hours on PubMed looking at the most published authors on Cushing's. Both Dr. Oldfield and Dr. Vance were on that list, and Charlottesville was just 4 hours away from my home. So I sent an email, then sent labs and scans, and waited to see if anyone believed me.

Drs. Oldfield and Vance believed me, and the wonderful Monte Parsons scheduled an MRI and consult. Dr. Oldfield knew I needed another MRI. The team at UVA had explained that his MRI was "different" and that it had a higher likelihood of finding my tumor. What they didn't tell me then, and I learned only later, was that my insurance company denied the prior authorization for the MRI since I had one a few months earlier. Dr. Oldfield had personally called the medical director, explained the differences, and reminded them that an inpatient petrosal sinus sampling (IPSS) would be more costly than a second MRI (the IPSS has not always been done on an outpatient basis). Without any worries to me, he made sure my MRI was covered.

While I waited the long six weeks for an appointment, I of course stalked both doctors on Google. I learned that Dr. Oldfield had dedicated more than 20 years of his life to research at the NIH and was one of the most respected neurosurgeons in the world. On the day of my appointment, I had an MRI, then waited in the pituitary clinic for my appointment with Drs. Oldfield and Vance. Dr. Vance said, "I'll let Dr. Oldfield tell you the good news," and my eyes filled with tears.

"Did you find my tumor?" I asked cautiously.

"We found it," he affirmed. Dr. Oldfield pulled up the scans on a monitor and showed us the 6-7 mm tumor. He told us that my tumor likely had "carotid involvement," meaning that it was wrapped around the carotid artery. He explained that it was a serious complication, but that he still felt confident that surgery was the best option.

I couldn't hold back the tears of worry, and Dr. Oldfield showed me immense empathy without even saying a word. He simply stopped talking, and graciously gave me time to absorb the news, to cry, and to gather my emotions so that I could listen and ask questions.

My husband and I both worked in health research at the time, so we asked, "Have you seen this before? What is the success rate with carotid involvement?"

Dr. Oldfield, in his quiet manner, calmly shared that he had operated on about 200 cases with carotid involvement, with a 98% success rate. Needless to say, he confirmed in that moment that we were at the right place with the right care team.

My surgery was scheduled for two weeks later, and I asked if I could be the first patient of the day. Demonstrating his dry sense of humor, Dr. Oldfield said, "I think so, but are you sure you don't want me to practice on someone else first?" He never masked the seriousness of Cushing's, but he made us smile and reassured us that I was not

an anomaly for his team, but the kind of patient he operated on regularly.

Two weeks later, Dr. Oldfield successfully removed my tumor using his expert technique that meticulously kept the tumor encapsulated in its tiny membrane and removed the tumor in one intact piece, reducing the likelihood of recurrence. In explaining that my surgery was successful, he noted that my tumor was "unremarkable" — meaning that it was exactly what he had expected to find. I went home three days post-op with fluid restriction for diabetes insipidus and replacement hydrocortisone, but no other complications.

At my two month follow-up, I came with gift baskets of treats for Dr. Oldfield, Dr. Vance, the wonderful pituitary clinic team and the hospital's neuroendocrine ward. Of course, no gift basket could show the deep gratitude to a team who literally gave me my life back, but it was a small token to show my thankfulness.

Of course, Dr. Oldfield not only thanked me then, but mailed a handwritten note to me. It was not signed "Dr. Oldfield," but "Ed" as though we were long-term friends or colleagues. Needless to say, I still treasure that note as I treasure the gift of restored health that was bestowed upon me.

At my one year follow-up, I shared with Drs. Oldfield and Vance that I was eight weeks pregnant. Dr. Oldfield told me that my life would find new meaning in ways that I could only imagine. My husband and I were blessed with both a daughter (now eight) and a son (four). We feel confident that this was possible due to the skilled hands of Dr. Oldfield who successfully removed my tumor and left my pituitary intact.

As I approach my 10-year "surgiversary," my gratitude has only grown. Dr. Oldfield gave the gift of health not only to me, but to thousands of others. He also spent thousands of hours in research to further the knowledge and best practices of Cushing's and neurosurgery in general.

I met one of his more recent patients at the CSRF patient education day this past fall. She shared that she asked Dr. Oldfield how he knew it was Cushing's and how he knew he could remove her tumor. He compared it to bird watching, a hobby of his. Dr. Oldfield said it required patience and dedication to hone the skills that enable you to see the rare birds.

Dr. Oldfield was both a rare bird and a rock star. He was a humble leader who was far beyond "practicing" medicine; he was perfecting it. His contributions to the Cushing's community and his gentle spirit are missed greatly.

Thank you for your support!

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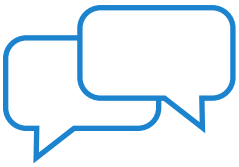

Need to talk?

Turn to page 27 for CSRF local support groups and contacts.

Patient Stories

Battle with a Brain Tumor / Cushing's Disease


With Cushing's


In recovery

My name is Teri Hudson. I am one-year postop and in recovery from endoscopic transsphenoidal resection of pituitary adenoma. Here's my story…

My long journey with Cushing's began almost 13 years ago before I knew I had the disease. I was a 39-year-old female, living life in the fast lane. I was a wife, a mother of 3 teenagers, and worked full time. I was generally healthy. I only weighed 130 lbs and was constantly on the go, being involved in school, sports, and church activities. But one Saturday in 2005, it seemed everything changed. I began experiencing some tingling in the left side of my face and arm. I became very tired. I brushed off the feelings as just being tired from a busy week and a day of running errands. I went home to rest, but when I awoke, I had no feeling in the left side of my face or my arm, and my leg was heavy. My speech was slurred, and I was very confused. My husband knew immediately that something was wrong. I was taken to the hospital to seek help and given blood thinners and told I was possibly having a stroke. I was admitted for a series of tests; after the tests were completed, I was diagnosed with having Bell's Palsy (a mild stroke affecting the face) and carpal tunnel syndrome (damage of nerves in the wrist.) Physical therapy was ordered to repair the damaged muscles in my face and strengthen my arm. I was also given steroids as part of the treatment. I was in hopes that this was just a one-time thing, but little did I know, over the next few years, life as I knew it would be turned upside down.

After the first neurological episode, my health seemed to decline for the next 12 years. I had kidneys stones several times that had to be surgically removed, developed Crohn's Disease, fell often and had several broken bones. I had several cases of strep throat that turned into abscesses that had to be lanced. In most of these cases I was given steroids as part of my treatments. After these treatments, I started gaining weight and was unsuccessful when trying to lose the weight. I asked several doctors and they explained that sometimes the extra weight could be from the steroid use and I would just have to work extra hard with diet and exercise.

Despite the health problems, I did not have any other neurological episodes until 2009. I began having tingling in the left side of my face and my arm, experienced confusion and slurred speech. I was admitted to the hospital, and after several tests I was diagnosed with having a mild transient ischemic attack, or TIA ("mini stroke"), high blood pressure, high cholesterol, and was considered obese at 160 lbs. I was prescribed blood pressure and cholesterol medication. I had been under a lot of stress during this time. Doctors told me that stress could have possibly been the cause of my latest episode but were unsure, so I was referred to a specialty Neurological Clinic for further testing. The physician I saw was very concerned about the swelling in my face and my blood pressure that was still spiking. He changed my blood pressure medication, and after reviewing my test results, diagnosed me with having a lacunar ("common") stroke. He said I should diet and exercise and take an aspirin daily. I followed his recommendations, but the same thing happened again in the Fall of 2010 with numbness in the left side of my face and arm. More CT scans, MRI, MRA, ultrasound of the arteries, heart tests, and lab work. All the tests came back negative. After several days of observation, I was diagnosed with having another TIA. I was once again referred to the neurological specialist. My previous physician had left the practice, and the physician that replaced him told me I just needed to lose weight and quit babying myself because my tests were negative. I consider myself a strong and determined person, so her comment that I would need to just learn how to better deal with my stress was frustrating for me and my family. Despite my illness, I worked full-time, took care of household chores daily, and attended every event possible. My family would have to force me to rest in fear I would get sick again. I was so determined not to be sick, but I knew deep down inside something was very wrong.

I returned to my local neurologist for more tests in 2011 after experiencing spells of dizziness. I was tested for Lyme disease, Lupus, MS, Myasthenia Gravis, Parkinson's disease, and many others. After all tests came back normal, it seemed the only thing left was to just accept that my illness was "undetermined" at this point.

My next "attack," as I had begun to call them, and a new symptom came in 2012 when I fell out of my chair at work and began having a seizure and stroke-like symptoms. I did not have any warning signs before this happened. I was taken to the hospital where I was admitted for several days and was told I had another TIA and multiple seizures. The seizures were unexplained, as it did not show any results on the EEG test. I was referred by my local neurologist to the Mayo Clinic for extensive testing. I spent a week being tested and

left with a diagnosis of complicated migraines. I didn't understand this diagnosis, as I had never had a headache as a symptom. It was explained to me that I could have a complicated migraine and not feel the pain of a headache. I was prescribed migraine and seizure medications.

At this point, I was willing to try anything because the label of "seizure patient" restricted my life to the point of adding stress to family members who had to do things for and with me for safety. I had several hospital admittances after seizures, and at one point I was referred to a sleep lab and a new diagnosis: sleep apnea. They thought my seizures might stop once I began using a CPAP and increased my medicine dose, but they continued periodically for four years. I was referred to a stroke specialist in Louisiana for a medical record review, and he concurred on my migraine and apnea diagnoses after confirming that signs of older strokes were visible on my scans. He suggested more rest and that I lose weight.

2016 was my worst year. It seemed that I stayed sick the entire year. In February, I had several seizures and was hospitalized, and it took weeks for me to recover. In May of that year, I was hospitalized again with kidney stones. At my follow-up visit with my primary care doctor after my hospitalization, she was very concerned about my blood pressure and my weight. I seemed to have so much swelling in my face and stomach. I was referred to the gastroenterologist for testing; all tests ruled out anything gastro-related. She added additional blood pressure medication and put me on a high protein diet. I began losing weight rapidity; however, I began having more symptoms of confusion, dizziness, chest pains, and was falling a lot.

My physician added a fifth blood pressure medicine when it did not seem to be controlled on the four I was already taking. At this point I was on 17 different medications daily. My health was declining and I spent time in ICU and had every scan and test in the book. Heart tests came back normal but seizures, high blood pressure, and stroke-like afflictions continued. I was newly diagnosed with low potassium and diabetes. My vison was changing daily, my hair was falling out, I was having memory loss and frequent panic attacks, chest pains, and tremors. I had a lot of left-sided weakness from the recent stroke, difficulty walking, and trouble swallowing. I began physical therapy for these side effects, but I was so physically weak that I was unable to hold up. I was unable to return to work as well. I felt like I was crumbling and falling apart.

A nutritionist reviewed my case and recommended a low carb diabetic diet. I immediately began gaining weight again and was still fatigued. I was then referred to an endocrinologist for my diabetes. My confidence was low after years of incomplete answers, but this new doctor took a real interest in my case. In November 2016 I was hospitalized after two more strokes, and while in hospital she called and said she thought she'd found my problem: I had a tumor on my pituitary gland. She said she felt I had Cushing's Disease, which was ironic because much earlier while trying to figure out what was happening with my body, my husband had done some research and came across Cushing's. It seemed like I had every symptom, but at the time my doctor said my lab work would have shown it and so ruled it out.

My new endocrinologist explained that she had done enough repeat testing to be sure and all we had left to do was an MRI to confirm the pituitary source, which it did. She gave me hope that my symptoms had been caused by this and should end once I had surgery. We speculated that my health had gotten so bad so quickly recently because something had put the tumor into overdrive producing high levels of cortisol. I was so thankful I had a correct diagnosis. I was immediately referred to a neurosurgeon at a larger hospital as an urgent patient and within three weeks I was scheduled for surgery to remove the tumor.

Wow, I finally had an answer. The surgery was performed in January 2017. I received an endoscopic transsphenoidal resection of pituitary adenoma. When I was admitted to the hospital, the lab work indicated that my cortisol level was a 29. The day after surgery, my lab work indicated that my level was a 7. This was a great sign that the surgery was a success. I can't describe how it felt to me and my family to hear that. The doctor explained that she had removed the tumor, but I had to understand that the recovery process was a long and difficult road. Recovery is not instant! She told me I did not get sick overnight and I will not get well overnight. The tumor had controlled the pituitary gland for so long that it was no longer working. It was in sleep mode, and it would take several months to wake up and function on its own. During this time I was cortisol-dependent and had to take the prescribed dosages for several months, then wean off the medication slowly. This was a very trying time. As the recovery process began, my body had to go through a lot of emotional and physical changes. My hormone levels were up and down. I would have sad dark depression days… but also some very bright days. The joint pain was unbearable at times, and the fatigue problems got worse. Every time my steroid dosage would be adjusted I would go into withdrawals; nausea, vomiting, joint pain, and severe depressions that would last for days.

Over the next year I slowly became myself again. The weight gain and puffiness went away, the fatigue and depression became less each day, my hair began re-growing, and my overall skin appearance looked better. I felt better than I had in a long time. One by one, the medications were stopped as symptoms went away. I had seven months of physical therapy to rebuild my muscle strength from the damage the Cushing's had caused and a few side effects from the strokes. Therapy was very difficult, but very helpful in the recovery process.

Continued on page 22

At my last follow-up visit with my endocrinologist, she was amazed at my results and the fact that I was no longer a seizure, stroke, and diabetes patient, that my potassium and Vitamin D levels were normal, and that the hypertension was gone. She did, however, make me aware that I will need follow-up visits to have MRI and lab work done because there is a 20% chance that the tumor could return, along with the symptoms. I realize that there is a chance that this terrible disease could return, but I am staying positive and looking at the 80% chance that it will not.

This long journey with Cushing's has been the worst experience in my life; the symptoms and side effects of Cushing's were very tough on me. The seizures and strokes, potassium loss, chest pains and dizziness, high blood pressure and diabetes, and the constant joint pain all over, as if you feel you have been hit by a train, was a daily struggle. I always had a "foggy head" and confusion. The weight gain when I was dieting and exercising regularly was frustrating. The fact that I just never felt well, had trouble walking, memory loss, and not able to do daily tasks was discouraging. As I look back and realize all that Cushing's has put me through and everything it has taken away from me, I am overwhelmed. All my symptoms did go away, but I still have a few side effects from the strokes; thankfully these are minor. I will also say after having gone through all of this that my faith is stronger and I realize what I can overcome. I am thankful for this clarity. I certainly have more respect for people with disabilities and illnesses.

As encouragement, I remind myself that a year ago, I was on 17 medications and now I only take three and I am being weaned from them. I wish that this disease was easier to diagnose. I wish people were made aware of the dangers of excessive use of steroids for treatment. I certainly will strive to educate those around me and perhaps my story could help someone get diagnosed.

I am thankful for the endocrinologist who finally diagnosed me, but also for the doctors along the way who did the best they could to treat my severe symptoms. I am most thankful for my entire family for all their love, support, and encouragement daily, especially my husband. I am the one that had Cushing's Disease, but it has truly affected all of us.

It's the beginning of 2018. I am about 90% recovered. It's like I have my freedom back. I would like to say to anyone who is battling any kind of illness, cling to hope, faith, love, and always stay positive that a cure or diagnosis may be right around the corner.

Teri Hudson
teri324@bellsouth.net
Alabama

With Cushing's


In recovery

Skin Deep

My story begins about three years ago, two years before I was diagnosed with Cushing's Disease. The difficult thing about Cushing's for me is that the symptoms are mostly based on physical appearance. I first noticed acne, weight gain, and irregular periods. When you try to explain these symptoms, they're written off as things that "just happen when your body changes". My mom agreed that something was off and encouraged me to keep going to the doctor. I got medication for acne but everything else was disregarded.

Thinking I must just be imagining these problems, I ignored them and tried to push them aside. I studied abroad in France for four months and didn't notice anything extremely out of place. There was so much going on in my life and everything was so different, I wasn't sure if my symptoms were just from being in a new environment. But when I got home, over the course of about a month, I lost half of my hair. I had always had thick, wavy hair. I complained about the frizz and having to manage it but all of a sudden, when it was gone, I missed it. I couldn't put it in a ponytail without bald spots and it was too thin to put in braids. It wasn't just my hair that changed. I didn't identify with this image of myself. My face swelled, my skin reddened, my blood pressure increased. I was gaining weight, regardless of any dieting or exercising. I felt like I was running in circles, spiraling downward. This was the body of my disease.

It was clear that something was wrong. I could feel it. Yes, women gain weight, and yes people lose hair during stressful events. But I was an active, 21-year old female. I know my body and I knew that something wasn't right. But Cushing's is difficult to diagnose and requires precise testing. Unfortunately, the first doctors I saw dismissed me with comments like "It's stress", "It's because you traveled", and "It's because you're getting older". Finally, after six months of appointments and worsening symptoms, my mom and I did our own research and suspected Cushing's Disease, and I found

"It was clear that something was wrong. I could feel it. Yes, women gain weight, and yes people lose hair during stressful events. But I was an active, 21-year old female. I know my body and I knew that something wasn't right."

an endocrinologist willing to test me. I took the dexamethasone suppression test at 11pm, got my blood drawn at 8am the next day as instructed, and I waited.

"Lila, I have your results and I'm a little surprised. Did you take the dexamethasone the night before your blood work?"

"Yes", I responded.

"Were you feeling at all sick or was anything wrong?"

"No", I responded again.

"Your results show abnormally high cortisol levels. We're going to do a few more tests."

Three weeks, another round of blood work, a saliva test, a urine test, and an MRI later, we found the source of my disease; I had a tumor on my pituitary gland. It sounds strange to be excited about a tumor but at that moment, I was ecstatic. I finally had an answer. I finally understood what had been happening the past two years.

Getting the diagnosis was only the beginning of the journey, however. After that came rounds of tests, appointments, follow-ups. I had to be careful with my body, being under constant surveillance and scrutiny. Well-meaning advice ate away at me — get more sleep, eat less sugar, exercise more, rest more. It was exhausting fighting against my own body and my growing insecurities. And for a while it was hard to feel beautiful. It was hard to be a 21-year old female, feeling like my appearance was slipping away, entirely out of my control. These changes might have seemed small or even unnoticeable to other people but this was my body. With these aesthetic changes came deeper emotional struggles. I had consistently high blood pressure and anxiety. I knew it would all go away after the surgery but I had been living with these problems for two years and the end still seemed so far.

One day, as I was rushing from yet another doctor's appointment to a blood test, trying to make it back to work on time for the third time in two weeks, I broke. On the sidewalk outside Mass General Hospital, the anxiety, nervousness, and frustrations that had been building and building finally overwhelmed me. I always try to be a positive, independent person. I want to find the silver lining and solve my own problems. But this was too much. I needed help. At that moment, a kind stranger saw me crying and changed my life. She hesitated at first, unsure if she should approach me while I was so upset. She came over and told me she worked right next door and that I was welcome to come in and talk if I wanted to. I wasn't sure at first — I didn't want to burden this stranger with all of my feelings. But I needed a helping hand and here it was. I went in with her and we talked for hours. She helped me make appointments, figure out insurance, and just vent about everything that was happening. At that moment I realized how far one act of kindness can go. It really is true that you never know how much of an impact one action can have.

After that point, I still got frustrated by what was happening but I felt so much relief and encouragement knowing that people cared. My friends and family were beside me every step of the way. I couldn't have gotten through it without my roommates, sympathetic listeners whenever I needed it. My mom was in NJ while I was in Boston but she did everything she could to support me. I started to realize that hard times bring out the best in people and make you see what is really important.

Ironically, as my physical appearance declined, my confidence soared. I saw how shallow and fleeting appearances are and really took notice of what makes a person beautiful. It's not their hair or their makeup. It's their passion. It's their personality. It's their love. And while I didn't exactly enjoy having bald spots, I did enjoy looking deeper into myself and finding my true beauty.

On December 7, I arrived at the hospital at 5:30 am with my mom. 7 hours later, I woke up, tumor free. A few ups and downs in the following weeks, but overall my medical team was incredible and I recovered amazingly. Now, about a year after my surgery, I am back to my old self. I feel healthy and active. I am training for my second half-marathon and loving my senior year of college. To any other Cushing's patients out there, please know that it will get better. There are highs and lows and it seems like the journey will never end but know that it will and you are strong. I will never forget what I learned from this experience. Through this disease, I was able to realize so much about myself and what really matters. I was able to see that my friends and family are so dedicated and loving and will do anything for me. And I was able to truly understand that beauty is only skin deep.

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In 2004, I was experiencing extreme pain in both knees. Swelling and pure agony. I was referred to an orthopedic surgeon. He started injecting me with cortisone to help the joints. The injections were really painful and caused more swelling than helping. Without knowing what was going on, I started to gain weight. My doctor chalked it up to my thyroid, but this was only part of the problem. About the same time, my gynecologist and an endometric surgeon were injecting a steroid-based medication into my uterus to try to help with the terrible spasms I was having with my menstrual cycle. I was not sure if they even knew that I was already receiving a steroid treatment for the knees. Unlike the knees though, the injections into the uterus were actually helpful. Still weight layered on.

Early in 2005, I awoke one morning to find my arms from my elbows to my fingers swollen like two giant clown hands. My skin took on a waxy, then crepey, consistency and I had HUGE blisters on all of my fingerpads. My doctor had no idea what was going on. Neither did the hospitals, but I was assured it wasn't fatal. I was prescribed even MORE steroids, my wounds were dressed and I was given a pat on my head and sent away.

Suddenly one night, I blew up like the Incredible Hulk. What follows sounds very horrific, and for me it was. I have not yet heard of any others who experienced such severe symptoms at such a rapid onset. The trunk of my body seemed to swell by the minute and stretch marks appeared from my elbow, up my arm and down to my waist on both sides. My torso turned red and I sought emergency help. I tried to explain what was happening but in my panicked state it was difficult to communicate well and the hospital staff didn't seem to know what to do with me. They told me to pull my shirt down when I tried to show them the stretch marks, which were bleeding. To me, the blood seemed never ending and I thought I might need stitches, but the hospital didn't feel that way. I ended up using maxi pads as bandages because they were large and flexible enough to curve around my underarms. Exposing any part of these marks at the pool or gym attracted unwanted attention. I still have two large scars.

I also began to sweat excessively. At worst, I could soak through my clothes in less than 10 minutes. Some days I would go through several outfits. My doctors were stymied; they had never seen something like this before, and they had no idea how to treat me. Adding insult to injury, the sweat had a very unpleasant smell — to me it was like dead skunk meets dead fish wrapped in cabbage! One insensitive hospital worker suggested I should have bathed before coming in. Psychiatric was called at one point and inpatient treatment was considered when they thought maybe I had caused the scars and was neglecting basic hygiene. I was depressed, exhausted, and constantly soaking wet. I could never get warm or dry. The best I could do was keep hydrated, keep clothing minimal, and wrap myself in wool blankets.

One morning I woke up in extreme pain, like a hive of bees had stung me between my shoulder blades. I am allergic to spiders and thought this might be caused by a bite. I rushed to the ER only to be dismissed yet again when doctors could not determine what was wrong. I got no help, not even for the pain. Near the end of 2015 I attended a Halloween Gala as a volunteer and feel that my physical condition went well with my costume: a zombie. People thought my "makeup" was great. I showed a friend my dark, angry stretch marks and she commented that it looked like I'd been rolling around with the Wolverine!

I was nearing the end of my rope when my doctor suggested I see the resident Dermatologist. This new doctor gasped when he saw my physical condition and asked if I'd ever been tested for Cushing's because I showed many of the symptoms — weight gain, oily skin and hair, profuse sweating, lowered body temperature, and the "Moon Face". And the scars. I was referred to an outside Dermatologist for another consult.

I had high hopes for this referral because what I had learned about Cushing's made me feel that I was headed for a diagnosis after so much suffering. This second Dermatologist asked me to take my clothes off so he could get the full picture of my physical condition. In fact, he wanted to literally take pictures; I agreed, thinking they might help speed up my diagnosis. Fast forward a month and that doctor was no longer with the practice but my photos were suddenly up on his website. I felt exposed and violated. I filed a complaint of sexual misconduct with the Ontario College of Physicians and Surgeons and requested my photos back, but I never got a response. The photos were eventually removed from the website.

My doctor helped me continue to pursue a Cushing's diagnosis, and next up was a "specialist" who was supposedly a leading authority on Cushing's. I couldn't get an appointment for 10 months, and when I did finally have my appointment I was dismissed in minutes and told "see you in a year." He never examined me, and I never went back.

I wish I could end my story with an understanding doctor who put me through a string of tests and gave me an official diagnosis. The truth of the matter is that I remain undiagnosed. I have struggled for almost 15 years now with a variety of debilitating physical and emotional conditions. I don't know if I have Cushing's but from all the research I have done based on what's going on with my body, I'm fairly certain I was experiencing Cushing's syndrome from the heavy use of cortisone injections. My relatives have not been kind and often make snarky comments. Former friends have dropped out of my life, but not before they have found it necessary to accuse me of living in unsanitary conditions, amongst other things. I am no longer asked to present Drama Sessions at a local children's organization (I've been a member since 1975) because people complained that I smell and don't wash my uniform. I now use a scooter for long walks. I can't camp anymore because I can't kneel or carry a backpack; this is something I used to love to do. I have been called Quasimodo and Igor.

The one thing my doctor has been able to help me with is the excessive perspiration — we found that with a prescription for clonazepam, a mild anti-anxiety medication, my panic attacks are controlled. My family is starting to understand that this isn't "all in my head". I've named my scars, which for me has taken some of the hurtfulness away from having to look at them every day. I try to live as healthily as possible, all things considered, and do not do drugs, smoke, or drink. I did lose my temper over the holidays when my aunt started to taunt me when I had one cookie with a cup of tea. I didn't feel too terrible when that cup of tea found its way onto her lap!

I have not given up on the chance that I might find a doctor who can help me regain some of the health I lost throughout this ordeal. The city of Toronto is the largest in Canada and home to some of the best hospitals. It seems unfathomable to me at this point that I am still without a diagnosis, especially because I have been seen by several doctors who know about Cushing's. I am in constant pursuit of answers but in the meantime I have found adaptive behaviors to lessen the physical and emotional impact of my condition. I am thankful to have found the CSRF website and online communities of people battling for a diagnosis and living with painful conditions. This fight has made me stronger. Thanks to everyone who bravely shares their story; they really help others facing this rare disease find ways to cope. Be proud; you are beautiful! Don't let people take that from you with their words. If necessary, dump tea in their laps.

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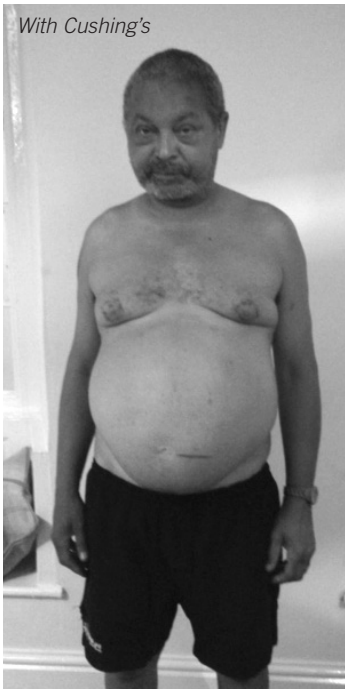
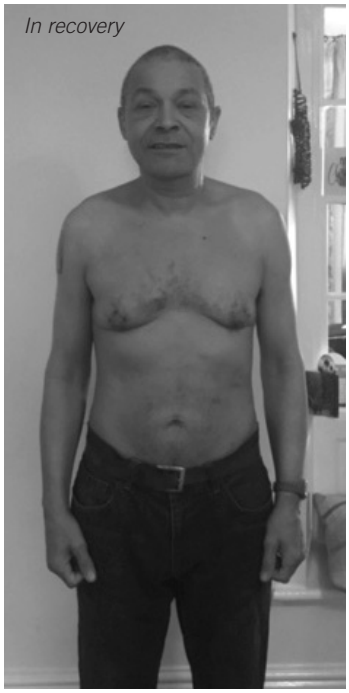

With Cushing's


In recovery

Prior to being ill I was healthy and fit, hard working, and living life to the fullest.To this day I still don't know how long I had Cushing's, but between my doctors and consultants we think about seven years — six before diagnosis! When I first found out that I had Cushing's, I found the stories of others really helped me to understand the disease and what to expect after treatment — which in my case was the removal of an adrenal gland and tumor.

At various times I had almost every single symptom of Cushing's, yet despite numerous doctor visits, hospital scans and a multitude of ailments, I went undiagnosed for more than half a decade. Eventually I was referred to an endocrinologist who was very intrigued to hear that I only pursued a referral due to my sister-in-law and brother's constant nagging for me to be tested for Cushing's. My sister-in-law is a vet and she was convinced that I had it; she said that if presented with the symptoms I had, that would have been the first thing she would have tested for! After diagnosis I was told that I would need surgery; after being cancelled three times, this finally took place in July 2017. I was told what to expect and an approximate length of recovery, but I was still unprepared for the recovery stage.

On the positive side, most of my ailments went away quickly — I lost the big belly, the round face and shoulders, my diabetes has gone, and my blood pressure is back to normal and I am no longer insomniac. The last six months have been very tough, however — I have been in constant pain and had an adrenal crisis so was back in the hospital for a bit — my body felt like I'd been run over by a

Continued on page 26

steam roller. We had a family holiday booked for Spain in August and I managed to go on that, however I missed a few days as I was ill and bed ridden, but it was nice to be in the sun. I had to keep adjusting my steroid levels and finally got it correct and still take it now with the hope that I can reduce/come off it very soon. I have had numerous doctor and hospital visits since, as I was still in pain and don't feel right. This is despite the surgery being a success.

My most recent hospital visit was to a consultant rheumatologist after a few doctors determined that the increasing pain in my joints might be a form of arthritis. So back to hospital I went. After more tests, a very confused consultant informed me that they concluded that I have Lupus, which is odd as again it's very rare in men and for people of my age. Lupus and Cushing's seem to be rare in men and hormonally related — we even joked that perhaps I was turning into a woman!! The consultant was a bit confused and unsure if this had anything to do with the Cushing's or its treatments, so I was referred back to my endocrinologist. She confirmed my remission from Cushing's and that we're just waiting for my adrenal gland to kick in so I can come off the steroids. The Cushing's could have been masking either Lupus or rheumatoid arthritis, and although I am still poorly recovering from Cushing's and in a lot of pain, I am now in another loop of testing to determine if my new symptoms are indeed Lupus or arthritis in hopes of finding a treatment for whatever it is. All in all it's been a long journey and it's not over yet but I do feel I am nearing the end and will be on the mend soon.

My journey with Cushing's contains a lot more details, but my main goal is to let fellow suffers know that yes it's a difficult road to recovery but unless you're unlucky like me your body and life will get back to some form of normality and you too can start living it to the fullest again. To those of you awaiting treatment I wish you all the best and those recovering good luck and God bless.

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We need your patient and recovery stories!

If you are interested in having your story in the CSRF newsletter, please email it to cushinfo@csrf.net or use Share Your Story under the Quick Links on our homepage.

Cushing's Patient Pamphlets

Pamphlets include *Understanding Cushing's, Diagnostic Testing, Recovery, Prevention and Treatment of Adrenal Insufficiency,* and *Why Replacement Medications are Needed.*

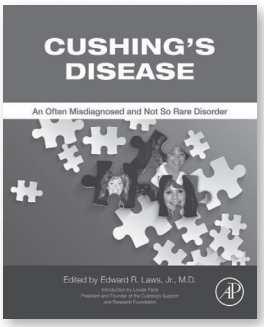
ENGLISH TO SPANISH TRANSLATION

Thanks to the efforts of Dr. Marta Araujo Castro, an endocrinologist at the Puerta de Hierro's Hospital, in Madrid, Spain, the Cushing's Patient Pamphlets have been translated into Spanish and are posted to our website!

ENGLISH TO FRENCH TRANSLATION

Thanks to the joint efforts of Anne Marie Bergevin and Dr. Andre Lacroix, French versions of some pamphlets have been posted to our website.

Cushing's Disease: An Often Misdiagnosed and Not So Rare Disorder



Edited by E. Laws, Jr., M.D., F.A.C.S., Professor of Neurosurgery, Harvard Medical School, Director — Neuro-Endocrine / Pituitary Program, Brigham and Women's Hospital, Boston, with an introduction by Louise Pace, Founder and President of the Cushing's Support and Research Foundation.

This comprehensive guide to Cushing's disease describes the functions of pituitary, adrenal and other hormones, lists the typical and atypical symptoms of Cushing's disease and its subtypes, outlines the causes of elevated cortisol and explains how clinicians can test for, diagnose and treat Cushing's disease. Physicians will find this concise book detailed, thorough and well-referenced. Patients will also find clear and helpful information in this important book. *Published in 2017 by Elsevier Inc — an excellent resource for physicians (and patients)!*

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If you aren't already on our mailing list, you can join through our web site at www.CSRF.net — Member Services, or just return this form to: CSRF, 4155 Lawrenceville Highway #8130 Lilburn GA 30047.

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Suggested donation is $30.00 or whatever is best for you. All donations are tax deductible. Please make checks payable to CSRF.

Are you a Cushing's patient?
☐ YES ☐ NO

Did you have:
☐ pituitary tumor ☐ adrenal tumor ☐ other

Would you like to be listed as a New Member in the next newsletter?
☐ YES ☐ NO

Can CSRF provide your email to others that contact us?
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Can CSRF provide your phone number to others that contact us?
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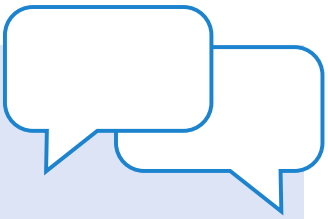

Need to talk? Local support groups and contacts

Many of you have expressed interest in local support groups and contacts. If you don't live in one of the following areas, consider starting a group in your area! Email the CSRF if you are interested. In particular, we have an existing group in the Los Angeles / Orange County area without an organizer.

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