

Sascha Groh

ONE OF



300

My Life with
Susac Syndrome



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Thank you

Dear readers,

I want to tell you my story. I fell ill with Susac syndrome in June 2012. The final diagnosis was not made until February of the following year. In the meantime I went through an odyssey involving various hospital stays, rehabilitation clinics and misdiagnosis. In those long eight months it was unclear whether I would survive, and if so, how. This book has been written for other sufferers, their families and friends, and also for doctors. I want to show which drugs helped in combating the rare disease, as well as the obstacles my family and I had to overcome. This book could be seen as an advice, guidance and information booklet. It is not about attributing blame because of one misdiagnosis but more about creating awareness of this and other rare diseases. Christine Scholz considers a disease “rare”, if “less than 1 in 2000 people on average contract it. More than 8000 diseases are currently considered rare. In Germany approx. 3 million patients live with rare diseases.”¹

This text was created in the form of collages, from my personal memories, those of my family and friends, of my boss at the time as well as my treating physician. For those interested, there are also reports from my doctors. Some of my memories were expanded by the narratives of others or corrected.

Sascha Groh
Cologne, March 2020

¹ Scholz, Christine: Liste der Zentren für Seltene Erkrankungen [List of Centres for Rare Diseases]. Current status. In: medizinische Genetik volume 29, 2017, pp. 13-20. online at: <https://link.springer.com/article/10.1007/s11825-017-0128-4>; accessed on February 29, 2020.

Susac syndrome, information from the European Susac Consortium

According to information from the European Susac Consortium, Susac syndrome is one of the rarest diseases. Based on the current research status, its cause and origin are not yet known in detail; nor have any genetic or other risk factors for the syndrome so far been detected.² An autoimmune cause may be assumed. According to the current state of medical knowledge significantly more women fall ill than men between the ages of 16 and 40. With reference to the progression of the disease, the website of the European Susac Consortium states: "There are two types of progression: a monophasic course of development (after a few years self-limiting in most cases) and a polyphasic and progressive one [...]. Considering the likely vasculopathy in the relevant three organs, Susac syndrome manifests itself symptomatically with a triad of (1) central nervous symptoms (predominantly encephalopathy and / or focal neurological symptoms), (2) hearing disorders (inner ear hearing loss, tinnitus) and (3) visual disturbances (visual impairments, visual field defects); the three components are rarely found simultaneously. In some people they appear over time. [...]. There is a narrowing and occlusion of very small vessels in the brain, ear and eye. Recent research results suspect misdirected immune cells, so-called cytotoxic CD8 + cells which become dysfunctional. Initially common are episodic headaches that can occur several months in advance of other symptoms. The combination of the clinical symptoms of visual disturbances and CNS symptoms in patients in the second to fourth decade of life leads the polycyclic form in

particular to be frequently misdiagnosed as multiple sclerosis [...].”³

At the time of my diagnosis in 2012 a total of 304 patients had been evaluated worldwide.⁴

² see <https://eusac.net/ueber-Susac-syndrom-krankheitsbild.html>, accessed on March 17, 2019.

³ Presentation by Ilka Kleffner for the Eusac network.

⁴ Online at: Nat Rev Neurol. 2013 Jun; 9 (6): 307-16. doi: 10.1038 / nrneuro1.2013.82. Epub 2013 Apr 30. Characteristics of Susac syndrome: a review of all reported cases, accessed on October 21, 2019.

**Preface by Prof. M. Krämer, Senior
Consultant in Neurology, Alfried
Krupp Hospital, Essen⁵**

Dear readers - anyone, that is, who is interested in Susac syndrome - In this preface I would like to recommend that you read these autobiographical memories by a Susac patient.

For rare diseases, information in easily comprehensible language is important. This book, however, can do more than just provide knowledge about a rare disease. It is not a factual book, but gives hope and courage to other sick patients with subjective impressions. I recommend this book to healthy readers and healthcare workers because the multifaceted description of the many dimensions of this disease is an invitation to change one's perspective. From smelly sick rooms to confused bed neighbors and the yearning for cigarettes, every human aspect is nicely covered.

Susac syndrome - the name sounds mysterious.

Many sufferers have been through a real odyssey before receiving a correct diagnosis and finding a targeted therapy. This book makes you understand the biology behind this disease better. As with many rare diseases, it is not just hazardous if you miss out on proper treatment at an early stage; in addition, the risk of dangerous and expensive mistreatment is immense. Rare diseases are in their entirety not "rare" at all. In Germany more than 4.8 million citizens are affected, the EU has around 40 million people who suffer from one of the approximately 8,000 rare diseases. Since no doctor can know all rare diseases, but humans tend only to

see what they know, humility is the greatest virtue in a doctor; so when dealing with patients who present with unclear conditions, you should listen, question and ask for advice and a second opinion.

In recent years in Germany a lot has been done, with federal government initiatives, academies for rare diseases, and especially advances in the field of medical education, advising health care professionals to listen to patients without bias and without passing judgment.

These efforts come up against economic constraints, the limited time resources in healthcare and the all-inclusive price in medical practices and hospitals.

By reading this book I hope you can learn not only from Sascha's experiences but have fun reading it despite the underlying tragedy.

Some episodes are weird to bizarre, some just touching and sad. Family cohesion and the persistent support of just a few friends, along with Sascha Groh's relative recovery, make for what is definitely a happy ending.

I wish you ample courage and strength with rare diseases. In not letting himself be got down, Sascha Groh is unquestionably a model worthy of imitation!

Yours,
Markus Krämer
Professor of Neurology at the Medical Faculty of
Düsseldorf

⁵ Prof. Dr. M. Krärner has been my treating neurologist since 2013.

Prologue

My name is Sascha. I was born in Koblenz, Germany in 1976 as the second child of my parents Horst and Marlies Groh. My sister Nicole, three years my senior, and I grew up on the outskirts of the city where the Rhine and the Mosel meet. Our parents separated when I was nine. From then on I lived with my mother while Nicole stayed in the family home with our father. I was a mediocre student and preferred to stay inconspicuous and not be at the center of attention. I enjoyed hanging out with older people from my neighborhood. They were so wonderfully calm and had a lot of stories to tell. They also had ice cream in all varieties, which was not something to complain of.

In March 1997 I moved to North Rhine-Westphalia for professional reasons, after having successfully completed an apprenticeship as a hotelier. I stayed at various locations, both professional and private. When it all started, I was 35 years old and employed at a large well-known company in Bonn as an assistant in project procurement. My job offered me new challenges every day. I lived in a fancy single apartment in the heart of the city of Cologne. As befits an immigrant to the city, I was actively involved as a carnivalist in an up and coming carnival society. My circle of acquaintances was large, I was often on the go and enjoyed the cultural opportunities the cathedral city offered. Fear of the future didn't exist for me - until a certain day in June 2012.



This photo shows me before I got ill.

June 2012

Sascha

June 26, 2012

It all started on a Monday morning late in June 2012. When I woke up, I was trembling all over. Tears were running down my face. Any attempt to form a thought seemed to fail. I had no idea what had just happened to me, only that I needed help as soon as possible. So I tried to contact my family doctor by phone. There, however, only the answering machine answered and announced the phone number of the stand-in physician. I can't remember today how I managed to make an appointment with him. The receptionist realized my panic and gave me an appointment the same morning. Unable to drive myself, I asked my sister Nicole to accompany me to the doctor.

When the doctor treating me asked a little later what was wrong with me, I immediately started to cry. I don't know why I reacted like that. So I searched frantically for a logical explanation for my present state. I explained my current state of emergency to myself and the doctor treating me as follows: The new work processes in my job have increasingly been stressing me out for quite some time. The longer I stayed with the company, the more activities in my area of responsibility had increased.

At the time of my breakdown I was working as an assistant in project procurement. Recently, department reporting had also been part of my work. In the past week the associated tasks had been increasingly demanding. Used to managing problems on my own, I had not been able to switch off for some time now, which inevitably resulted in

my becoming more and more nervous and tense. This was explanation enough for me, and also for the doctor. Apparently it needed no further investigation. The doctor prescribed me an antidepressant and rest. I panicked and asked him to inform my boss about my sick leave, and tell him to please not contact me during this time. I was uncomfortable about showing weakness on the one hand, but on the other I wanted nothing more than to be able to switch off again. I didn't listen to the call between my doctor and my boss.

Torsten Stannieder, Sascha Groh's superior 2008-2012

In July 2008 I changed employers; at the new job I was counting on the support of an assistant. On my first working day I found out that my assistant was currently on vacation. This was my first encounter with Mr. Sascha Groh.

After his vacation we got to know each other. I noticed Mr. Groh's caution towards me very early on, it took some time to wear off. It was apparent that Mr. Groh had already experienced a lot in the company and was nonetheless still highly motivated. Our workflows became increasingly familiar. Each of us knew what to do and to expect from the other. As a team (boss and employee) we got better at working together month by month and over the years until 2012, when I observed a change in our professional relations. Mr. Groh was more thoughtful, his lightness seemed to have been lost and he was reacting more sensitively than usual.

The first thing I always do is check myself: Did I change, have I done something wrong or did I demand too much of Mr. Groh jobwise? I concluded on reflection, however, we were communicating as we had done in previous years. So I asked Mr. Groh directly and he just said, "Everything is fine,

everything is OK.” His job also involved responsibility for the weekly departmental report. Once he had got used to this it went smoothly, though it wasn’t exactly rocket science. Just a matter of playing around with a few Excel tables to make it look good. Mr. Groh started to find this work more stressful in 2012, he did (in my view) slightly panic, and apparently he was also having stomach and bowel problems. Following consultation I decided to split the reporting and another colleague took over half of it. Some team members took exception to my decision, which I for my part found hard to understand.

Then in June 2012, I was sitting in my office and received a call. The caller introduced himself as the doctor of Mr. Sascha Groh. The first thing he said was, “I can’t tell you everything for reasons of medical confidentiality and protection of privacy.”

This call shocked me. Never before had the doctor of a colleague or employee called me. The doctor wanted information about the past few months of our working together and about Mr. Groh’s activities. I confirmed to him that the reporting, in particular, wasn’t exactly rocket science, but at the moment Mr. Groh was finding it too much. Then he described his experience with Mr. Groh superficially and I thought: BURNOUT! at once. I asked the doctor if it was a case of BURNOUT. He didn’t confirm it, but left me with that impression. At the end of the approximately 20-minute conversation he informed me that he would have to take Mr. Groh out of circulation for quite some time. After the phone conversation I immediately informed my superior and the colleagues from my department. Again I was struck by the rather curt reaction of colleagues and employees, who seemed to make light of the absence of Mr. Groh, whereas my then superior responded superbly and also offered to help - quite like me in fact.

Sascha
June 26, 2012

With the diagnosis of burnout and a prescription for an antidepressant I was on sick leave for three weeks. Relieved, and in the firm belief that my condition would improve soon, I left the practice.

Today I know that I would no longer give any doctor an initial self-assessment.

The way home turned out to be arduous in my condition, as my sister had gone back to work. Even though my head ached massively and I myself had a hard time concentrating on the drive home, which I see in retrospect was irresponsible as I was a threat to myself and my fellow human beings in the condition I was in. At home I immediately crawled into bed. Still my head seemed completely confused. Inwardly I was agitated even though I was lying calmly in my bed. Fear rose and seemed take the breath out of me. An irrational fear seized me. Fear that someone could ask me something I did not want to and could not do and that I didn't feel able to do. My head didn't seem to be able to filter anything anymore, all thoughts and stimuli were immediate. Was I losing my mind? For the first time in my life I turned off all of my communication devices. Now I couldn't be reached either on my two cell phones (one company, one private), or on my landline.

This calm around me gave me security because now nobody could make any demands. I withdrew from everything and everyone. I do not know how long I was lying there, but at some point I must have gotten tired and fallen asleep.

The next few days I spent alternating between the couch and lying in bed. Eventually I noticed that I got calmer thanks to the tablets I had been prescribed. I finally succeeded in what I hadn't done in a long time: just relaxing! I blossomed. After 14 days I was high spirited and