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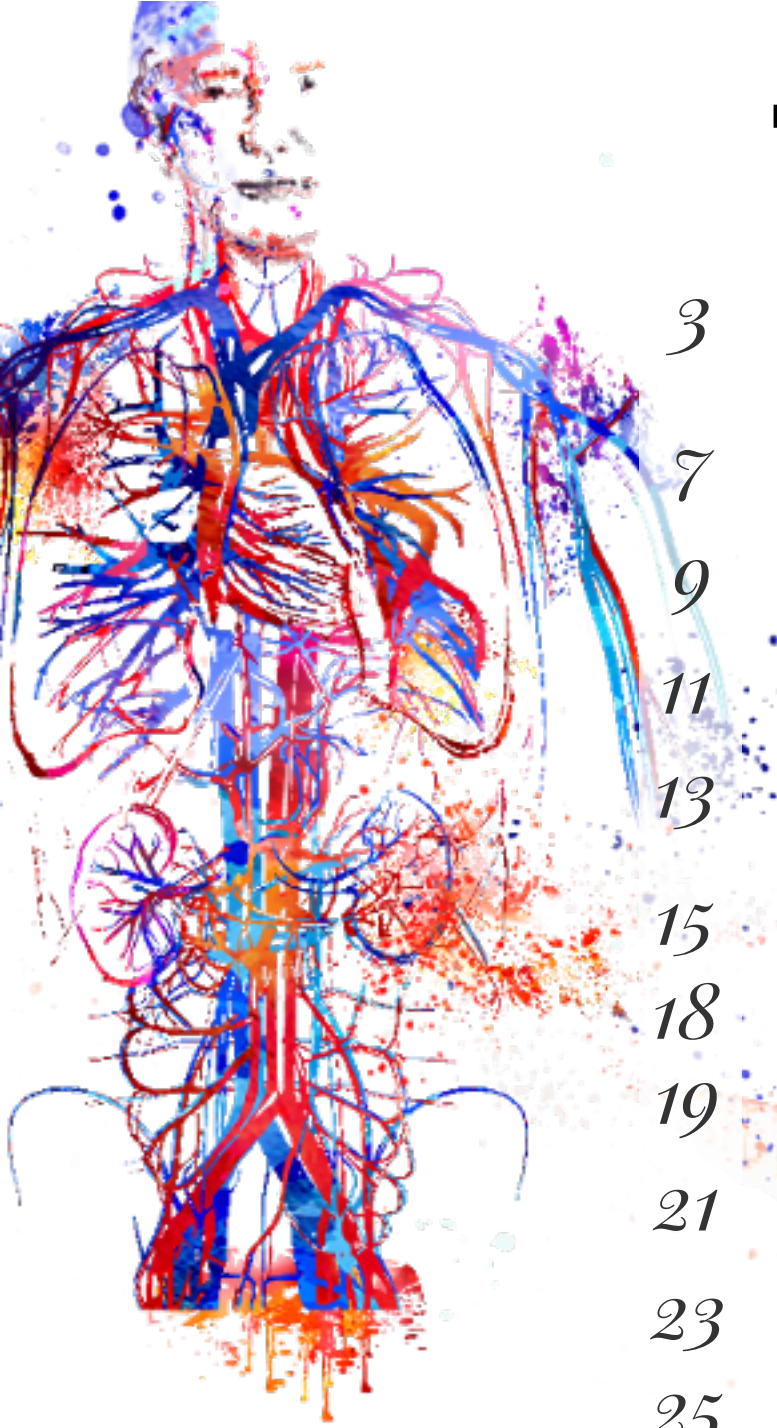
RARE REVOLUTION™ Magazine

Special Edition

ANCA-associated Vasculitis

World Vasculitis Day
15 May 2020



During the present Covid-19 pandemic, we encourage all vasculitis patients to follow the advice of their medical professionals and their country's public health guidance.

Front Cover: **Martina (left) and Maresa (right) pictured, both from Germany, photograph courtesy of SEE ME. HEAR ME.**

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Meet *our Team*

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Editor's *Welcome*

Join the #RARERevolution

Raising awareness is a pivotal part of the RARE Revolution movement and at the centre of all of our activity. Whether it's through our RARE story-telling, our social media #tuesdaytakeover initiative or our day-to-day work in building communities and facilitating powerful collaborations in the rare disease space, awareness raising is what we do—it's what we are passionate about.

In every edition of our quarterly RARE Revolution Magazine we like to hone in on a theme and spotlight particular fields or topics within the wider rare disease space which allows us to cover a vast range of diseases and disease groups and highlight topical and emergent issues.

But throughout the year, between core editions, we are always very excited when we have the opportunity to work on special supplements which allow us to really drill down further into specific diseases. This allows us to consider the whole picture—from scientific and medical challenges and breakthroughs, to clinical care delivery, patient diagnostics, and, most significantly the varied and unique experiences of individuals, families and care-givers—in one disease group.

It is such a privilege for us to work closely with the professionals, charities and families to create these special editions and we are immensely proud to create content for these communities that provide lasting and comprehensive awareness materials which are highly shareable for the whole global disease community.

To this end, we are incredibly proud to have partnered with the team at Vifor Pharma enabling us to work closely with a range of international contributors to create this wonderful and engaging special edition, shining a spotlight on the rare disease of ANCA-associated vasculitis (AAV). This special edition supplement provides powerful insights into the impact of living with AAV and showcases the latest developments, and initiatives that are promoting disease education in the field.

It has been our honour to work with the AAV community on this informative, colourful and thought-provoking edition to mark **World Vasculitis Day 2020**, and we hope you join us in helping to raise awareness on their special day this May 15.

Nicola Miller
Editor-in-chief
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This issue has been proudly sponsored by Vifor Pharma Group

VIFOR PHARMA

VIFOR FRESENIUS MEDICAL CARE RENAL PHARMA

RARE Revolution: To bring about a dramatic and wide reaching change in conditions and attitudes for the **rare disease** community

It's time to **turn the tide!**

Spread the word - the **RARE Revolution** has begun

RARE Revolution
Turning the **tide** for **rare disease**

Shining a spotlight on ANCA-associated vasculitis: A unique journey for every patient

AAV is a group of rare autoimmune diseases,[1–4] meaning that the body's immune system attacks healthy cells.[5] AAV is a long-term condition, that results in the inflammation of small blood vessels throughout the body leading to damage to organs such as the kidneys, lungs, ENT (ear, nose and throat), skin, nervous system, gastrointestinal system, eyes and heart.[4]

- **Each year, AAV affects up to 20 new people per million in Europe[1]**
- **AAV can affect both younger and older people, but is especially rare in children and young people[1,6]**
- **AAV is slightly more common in men than women[1,6]**
- **The most common subtypes are Wegener's granulomatosis with polyangiitis (GPA), microscopic polyangiitis (MPA)[1] and eosinophilic granulomatosis with polyangiitis (EGPA)**

What causes AAV?

In autoimmune diseases such as AAV, an antibody, which usually protects the body from anything seen as foreign such as bacteria and germs, develops to antigens (proteins) on the body's own cells. These antibodies are known as autoantibodies.[5]

In AAV the autoantibody called ANCA (anti-neutrophil cytoplasmic antibodies) develops to an antigen (protein) expressed on the surface of neutrophils, a type of white blood cell.[2] Neutrophils are activated when ANCA binds to them, leading to inflammation—the body is not meant to respond like this, as the inflammatory process is designed to fight infections.[2,7]

C5a is at the heart of vasculitic damage in AAV[2,7]

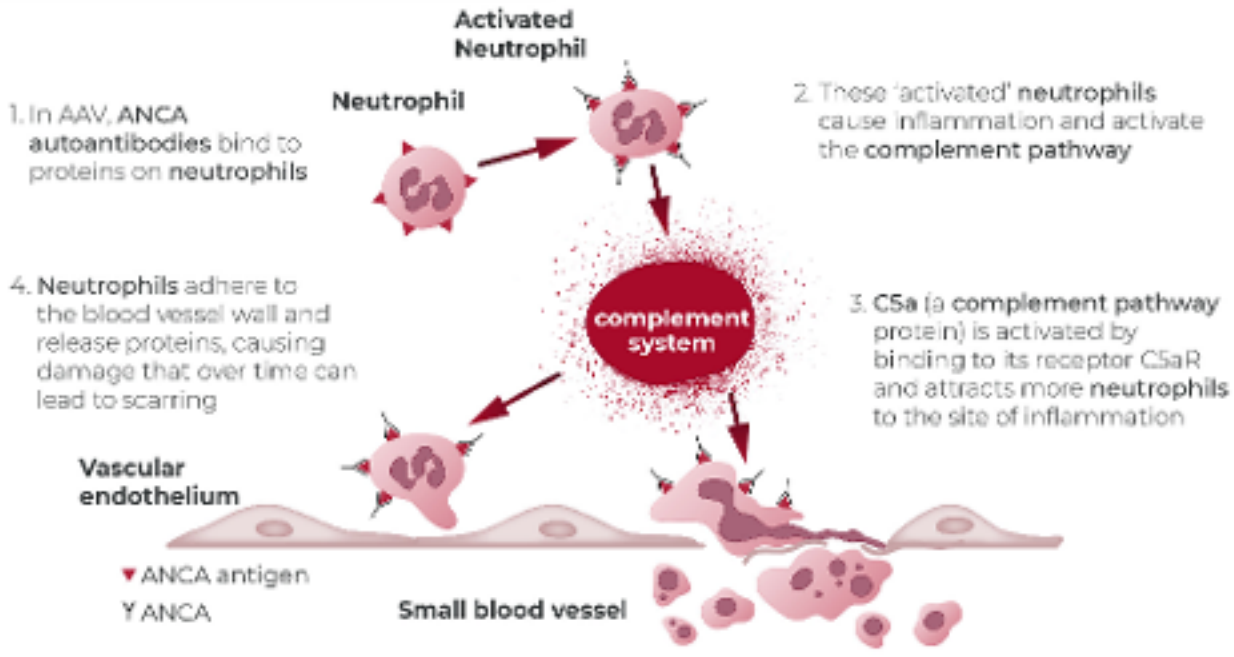


Diagram adapted from Jennette JC, Nachman PH. *Clin J Am Soc Nephrol* 2017

ANCA-ASSOCIATED VASCULITIS

Activated neutrophils then activate the complement system, made up of a series of proteins that interact with each other. Activation of C5a, a complement system protein, attracts more neutrophils to the site of inflammation and readies them for activation.[2,7] The activated neutrophils stick to and penetrate the blood vessel's walls releasing proteins that would normally aim to fight infection, but in AAV they cause inflammation and damage in and around the vessel wall.[2,7] The interaction of the complement system and neutrophils drives a cycle of inflammation that results in damage to blood vessels. The body's immune system over time can regain control of the inflammation, but there is typically scarring of the tissue in the area where the blood supply has been affected.[2,7]

C5a is at the heart of vasculitic damage in AAV[2,7]

The initial causes of AAV and the development of ANCA are unclear, but various factors may increase the likelihood of developing AAV, these include:[2,4]

- Microbial infection
- Genetics
- Environmental causes e.g. chemicals and construction materials
- Medication or recreational drugs

The signs and symptoms

AAV can affect any part of the body, as the small blood vessels affected are located throughout the body.

The impact AAV has on individuals can vary greatly, with a range of symptoms depending on the organ(s) primarily affected.[8]

The main types of AAV, GPA, MPA and EGPA, have different signs that can help to distinguish between them, but people can experience characteristics associated with **GPA, MPA and EGPA.**

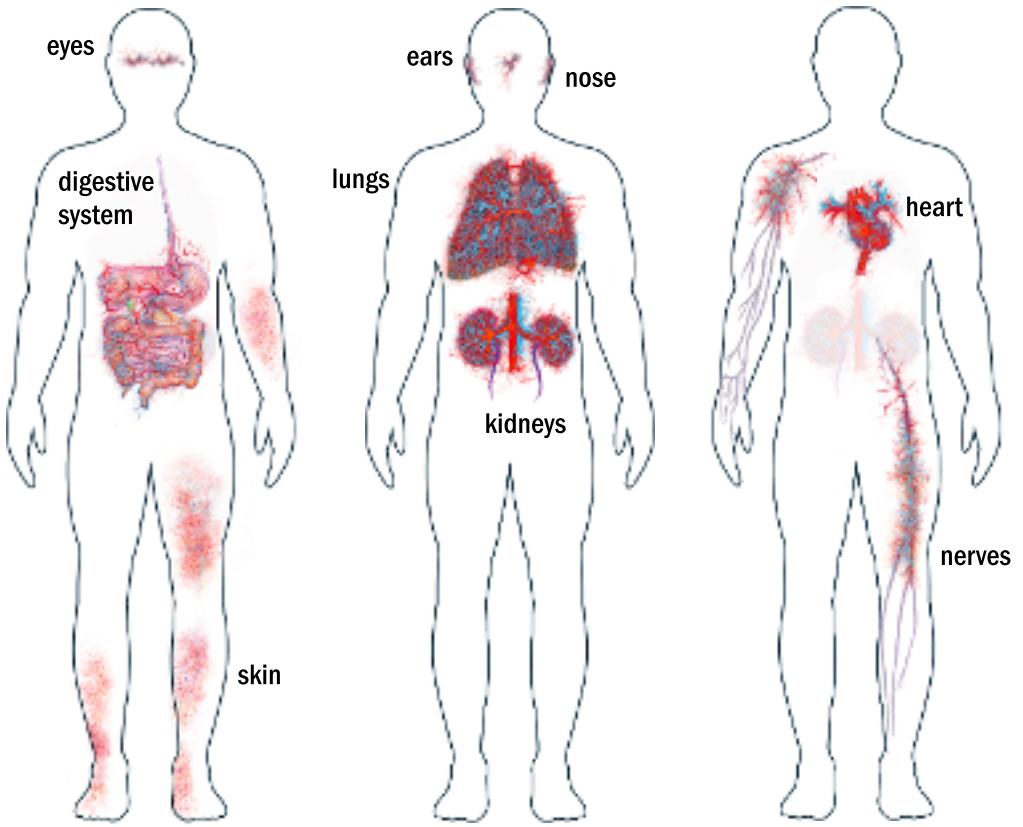
Subtypes of AAV

GPA often begins in the nose, ears, eyes or mouth; a common sign is chronic blocked nose accompanied by crusting of blood around the nostrils. Other common signs include redness in the eyes, ear infection and hearing loss, as well as effects in the kidneys and lungs[7–9]

MPA typically affects the kidneys; common symptoms include having a bloody cough, a purple rash and joint pain[7,8]

EGPA - the most prominent symptoms are those related to pulmonary, cardiac, dermatological, renal and peripheral nerve involvement

The figures show how multi-systemic vasculitis can be



ANCA-ASSOCIATED VASCULITIS

The essential principles of AAV management[10]

The patient experience

The rarity and complexity of AAV, with its broad range of symptoms, means that often patients have to see multiple doctors and it can take months, sometimes years, before they are diagnosed and begin treatment.[11,12] Patients typically present with a multi-organ disease. At diagnosis the most commonly involved organs are kidneys (78%), lungs (41%), skin (35%), sinuses (27%), nose (22%) and nerves (15%).[12] Often to confirm a diagnosis multiple tests may be required, including: blood tests, urine tests, imaging procedures and biopsies.

For one in three patients it takes more than six months to receive a diagnosis[11]

AAV is a long-term, relapsing-remitting condition, but with treatment it can be controlled and patients feel well; however, there can be other periods when symptoms worsen and damage occurs. At these times patients require more support. The variability of AAV means it is hard to predict how it will progress and how one individual experiences the disease can be very different to another.[13]

Treatment received is dependent upon the type of AAV and the severity of the inflammation. Patients are prescribed a combination of therapies that reduce the activity of the immune system.[13] The majority of new-onset patients have evidence of severe AAV, also known as organ-threatening or life-threatening disease.[11]

Referral, diagnosis and management of GPA and MPA patients[7]

of patients are referred by another physician

of patients are diagnosed as an inpatient

of patients are managed by more than one physician for AAV

ANCA-ASSOCIATED VASCULITIS

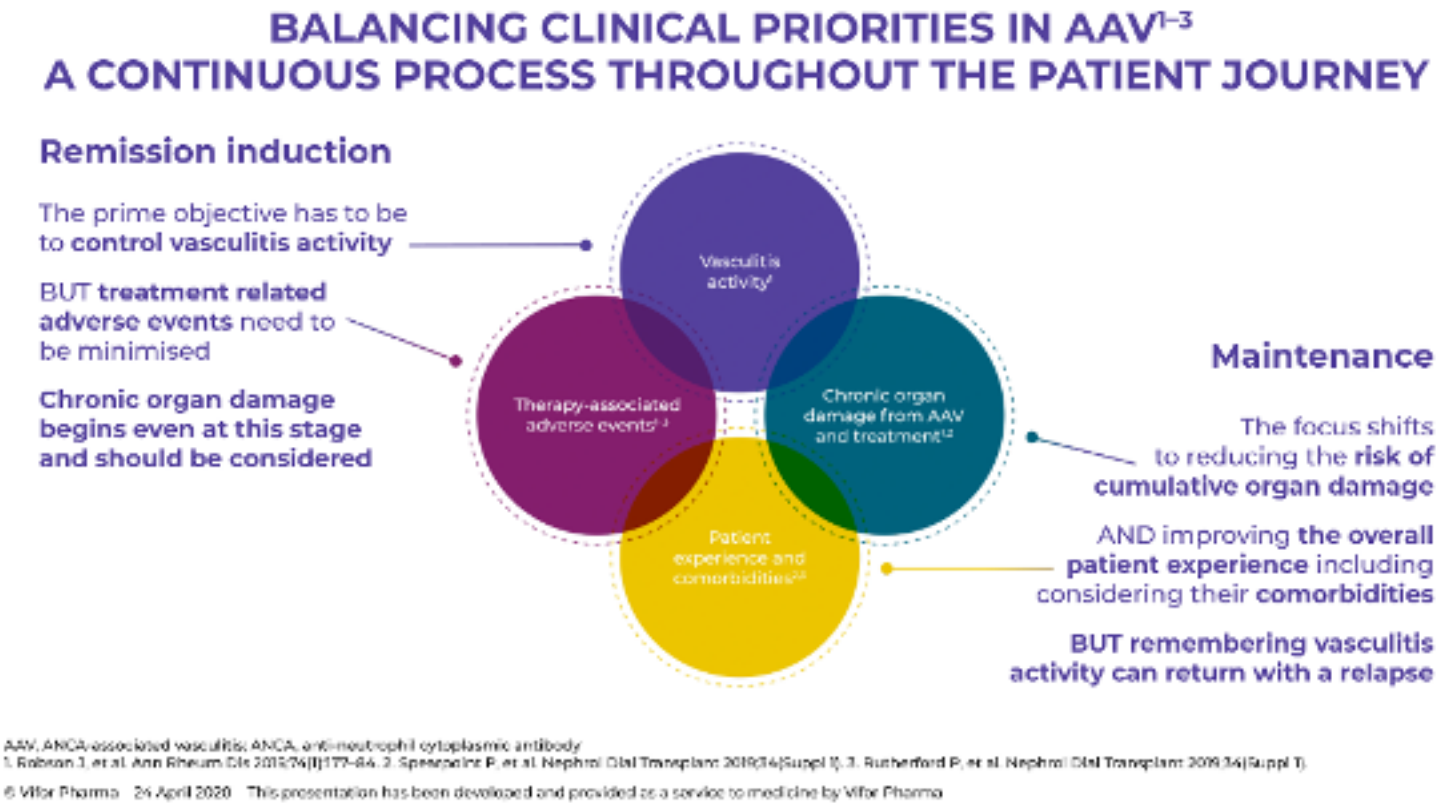
The primary goal of therapy is to achieve remission, the absence of disease activity. The current standard of care for remission induction in patients with organ- or life-threatening AAV, is a combination of high-dose glucocorticoids (GCs) plus cyclophosphamide or rituximab.[13] Whilst full drug-free remission can be achieved, disease relapse is common,[13] and in real-world practice patients still receive GCs throughout their treatment journey.[13–15]

Due to the broad action of treatment, patients often suffer a variety of side effects.[11] Irreversible chronic organ damage accumulates over time both from the activity of the disease and GC-related adverse events. Long-term high-dose GC use increases the risk of infection and diabetes, hypertension, osteoporosis and other debilitating side effects.[16] Patients also experience emotional effects from the use of GCs such as depression, anxiety and irritability.[17]

For each patient their experience of AAV is unique, to symptoms, diagnosis, treatment, living with the disease, and the differing physical and emotional impact at each stage of the patient journey. The major challenges faced by AAV patients throughout their journey leads to significant emotional distress.[15] Patients commonly experience a number of different emotions such as; fear, uncertainty, anxiety, powerlessness and isolation.[17,18]

AAV is a rare disease[1–4] that from the initial disease onset impairs the quality of life of patients.[19]

More needs to be done to support these patients on their AAV journey.



AAV, ANCA-associated vasculitis; ANCA, anti-neutrophil cytoplasmic antibody
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The rare disease ANCA-associated vasculitis

Professor Dr Bernd Hohenstein, consultant for internal medicine and nephrology, provides his insight and expertise on ANCA-associated vasculitis, discussing current and promising new treatments and highlights the importance of early diagnosis

Professor Dr Bernd Hohenstein

Consultant for internal medicine and nephrology, Nephrologisches Zentrum Villingen-Schwenningen

Rare diseases are indeed rare; how many ANCA-associated vasculitis patients are you currently treating at your centre?

Here at the centre we don't see such diseases very often. However, due to the large catchment area of our nephrological centre and the involvement of seven hospitals, we look after a total of 60–100 chronic patients during any given year. Patients affected by ANCA-associated vasculitis (AAV) are one of our main focus areas as far as rare diseases are concerned. On average, we see one new AAV patient per month.

What are the particular characteristics of ANCA-associated vasculitis?

Symptoms: AAV is a rare, severe systemic disease (one that can affect multiple organs) that, if left untreated, is fatal in over fifty per cent of cases. However, even with treatment, mortality rates are two to three times higher for patients with AAV. It is a disease that requires long-term aftercare, because these patients may relapse. Despite the severity of the disease, in many cases the right therapy will result in complete remission. Even in the event of renal failure requiring temporary dialysis, it is possible to regain almost regular renal function.

Currently available therapies/treatment options: AAV therapy suppresses the immune system. In cases of substantial organ involvement, the standard therapy consists of the administration of high dosages of glucocorticoids as well as chemotherapy.

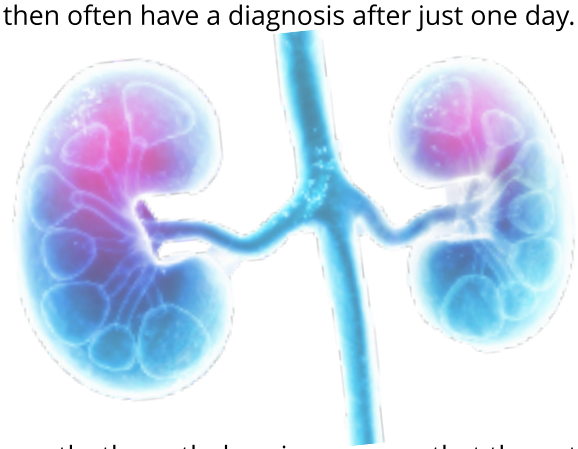
Among other components, cyclophosphamide is used as an active agent; this is also being utilised in cancer treatment—albeit in higher dosages. The antibody rituximab constitutes a good alternative, as its effect on vascular inflammation is achieved by removing the B cells. These therapeutic approaches are however also associated with risks for the patients so that additional medication has to be administered to protect the patients from complications.

Plasma exchange is currently a hot topic, as it constitutes a complementary therapeutic approach to treating renal and respiratory failure. This way, the pathogenic ANCA can be removed from the blood. However, according to recently published studies, the enhanced effect of the treatment is controversial. A plasma exchange should always occur early on, over a period of five to seven days and as an inpatient at the clinic.

Is the diagnosis difficult, and how long does the diagnostic process usually take?

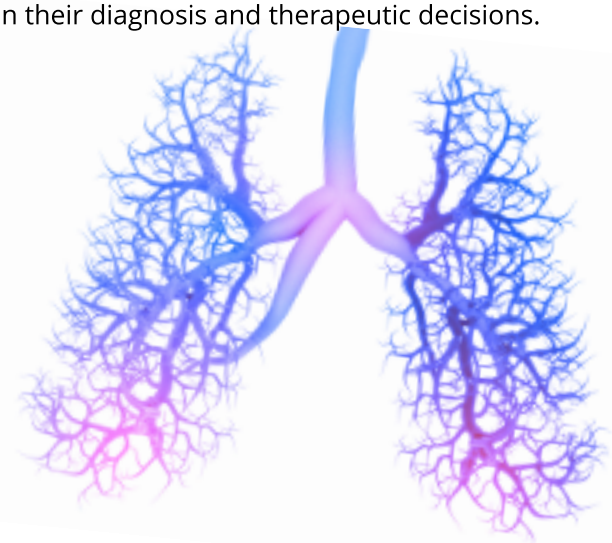
The diagnosis is definitely difficult because symptoms are so varied. There are patients with milder symptoms who often see a general practitioner repeatedly without being diagnosed properly for a long time. Frequently, AAV starts off with non-specific symptoms such as, losing sensation in one particular nerve branch, night sweats, loss of appetite, weight loss or joint pain. What comes to mind first are obvious illnesses such as the common cold or other infectious diseases. Hence this *mild presentation* is quite hard to diagnose. The patients will see a nephrologist once the kidneys are involved. In our specialist discipline, diagnosis actually forms part of a routine screening for systematic diseases with potential renal involvement, which is why we frequently detect new patients.

If the symptoms are mild, it can take weeks or even months to find the cause of the illness. If the symptoms are severe or even life-threatening, however, the patients can be diagnosed considerably faster, sometimes within just a few days. If the general practitioner determines serious changes in the blood count or signs of organ or renal failure, the patients are referred to a specialist within one or two days and will then often have a diagnosis after just one day.



Frequently, the pathology is so severe that the patients will require swift and extensive treatment, i.e. chemotherapy.

Apart from the kidneys, the lungs are also frequently involved. In these patients, the first suspected diagnosis is pneumonia (as the most obvious disease). Only once antibiotics fail to work and the symptoms do not start to abide other causes are taken into consideration. Sometimes, these patients present with symptoms suggesting pneumonia/involvement of the lungs and then start going into renal failure before it is finally realised that it is not the infection that is the root cause but rather a consequence of a severe autoimmune disease. In therapeutic terms this requires an about-face. For days, antibiotics are administered and the immune system is boosted, and then the therapy is switched to the opposite and everything is done to suppress the patient's immune defence. This requires experienced physicians who are sure in their diagnosis and therapeutic decisions.

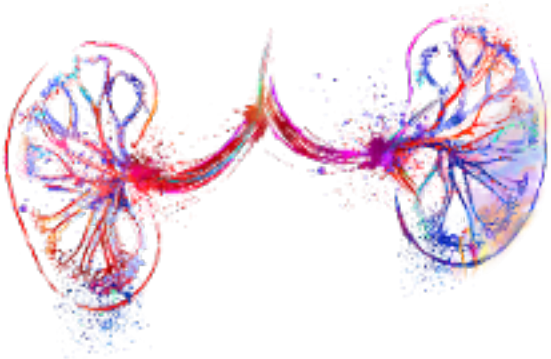


How frequently do you see these patients and how is collaboration with resident doctors and other specialists?

We see AAV patients every week, not necessarily always when they are newly diagnosed, but regularly during chronic disease management. After all, some patients require dialysis. In total there is a team of 16 specialist doctors at our centre and during our weekly conferences we discuss all complex pathologies such as our AAV patients. Our centre is directly linked to the clinic so that interdisciplinary cooperation with other specialist departments is straightforward and smooth.

During the early treatment stage, AAV patients are closely monitored. Immunosuppression must be managed very thoroughly and requires continuous adjustments. We are in charge of the treatment, and we keep the patient's general practitioner in the loop. We only relinquish a little more control once the patients are stable and no more severe complications are to be expected. Successful treatment and patient safety are our top priorities when it comes to AAV.

UNDERSTAND AAV



An educational initiative about ANCA-associated vasculitis for healthcare professionals

For most general physicians, to see a patient with ANCA-associated vasculitis (AAV) is rare. And since ANCA associated vasculitis affects each patient differently,[1] it can be challenging for the physician to immediately recognise the signs and symptoms of AAV.

When a patient is referred to a specialist for diagnosis, they will see a nephrologist, a rheumatologist, a gastroenterologist or an ENT (ear, nose and throat) specialist, depending on their symptoms and clinical signs.[1] The specialist needs to be aware of the current therapies and their effectiveness, since only 48% of patients sustain remission at one year after achieving full remission at six months.[2,3] And to appreciate that at a mean of seven years post diagnosis, two in three patients have potentially treatment-related damage.[4] Therefore, from diagnosis through to treatment, there remains a need to raise awareness and understanding of AAV within the clinical community.

The path to diagnosis can be difficult, lasting up to six months or more in one third of all patients.[5] And once diagnosed, challenges remain: **although current standard of care has increased survival rates, without continued use of glucocorticoids, only 48% of patients sustain remission at one year after achieving full remission at six months.[2,3]**

Bridging a gap in scientifically relevant information

Vifor Pharma, a leading player in nephrology, cardio-renal and iron deficiency is supporting the development of new therapies for rare diseases, and specifically AAV. Discussions with vasculitis experts across Europe confirmed the challenges around knowledge about the pathophysiology of AAV, diagnosis, current treatments and clinical outcomes.

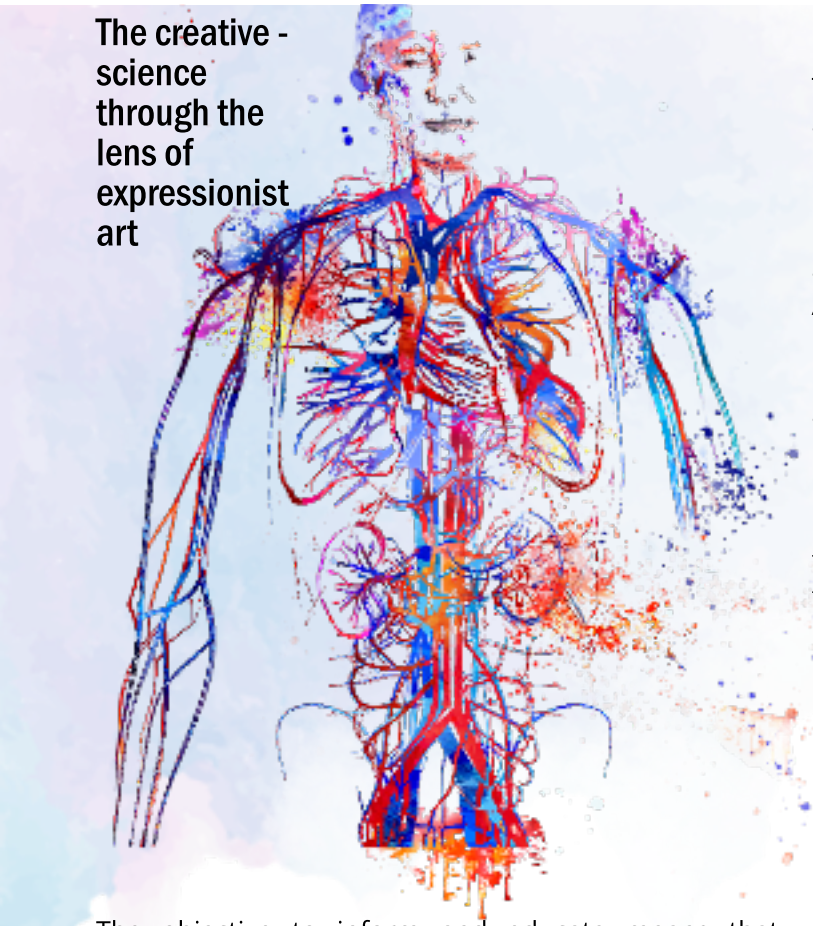
Vifor Pharma sought to bridge that gap and set out to provide EU-wide physicians with a multilingual online resource that would educate on AAV, providing a hub of comprehensive, evidence-based information: understandaav.com. The site went live at the beginning of 2019 and includes up to date information and interactive infographics, including an award-winning investigative tool to answer the key questions from physicians looking after AAV patients. These include;

- **Epidemiology** – what are the incidence and prevalence and how does this vary across Europe?
- **Disease classification** – what are the different subtypes and how do they fit in with the ANCA antibody results?
- **Pathophysiology** – what are the underlying causes of AAV? What drives the acute vasculitis process? How do complement and neutrophils interact in this disease?
- **Patient pathway** – what is the journey to diagnosis and who is caring for the patients? How do patients react to the diagnosis and what do they feel like over time?
- **Treatment guidelines** – what do the EULAR/EDTA treatment guidelines indicate about getting remission and then maintaining it? What are the definitions of the various stages of treatment – remission, relapse?
- **Clinical outcomes** – what is the evidence around remission rates? What is the impact of glucocorticoids in the long and short term? How much cumulative organ damage and renal disease occur?

The site also contains links to useful resources such as clinical scoring tools, professional societies, country patient associations and dates of upcoming congresses. In addition updates are made with relevant real-world data from Europe presented at

major international scientific congresses. The overarching objective of understandaav.com is to fill educational gaps around ANCA-associated vasculitis.

In order to get more physicians aware of the site, a multilingual online campaign (via paid search, social media and specialist journals) was launched in September 2019, and then rolled out to full implementation in early 2020.



The objective to inform and educate means that accurate and up-to-date scientific content is key but such an online tool needs to be interesting and attractive. To complement the content of the understandAAV online platform, an arresting core visual was needed to reflect the underlying science. The kernel of this creative idea was based on four key values that underpin understandAAV: driving innovation; showing leadership; transforming perceptions; and caring for others.

The core visual behind understandAAV is a marriage of science and art, combining the scientific insights with the digital painting skills of artist Sarah Govia. The result is an explosive Jackson Pollock-style representation of AAV patients and the pathophysiology of their disease. This image captures the underlying complexity of AAV, the interconnectivity of blood vessels and organs throughout the body, from the eyes and limbs, to the kidneys, lungs and heart, all potential *impact points*. As this disease plays out in the body's small blood vessels it was also imperative to *zoom in* and take a closer look at AAV disease activation at a microscopic level.

The result is a two-fold image: alongside the core abstract image of the patient, we can see the pathophysiology of the disease *emerging* from the kidney. This microscopic *side-bar* presents AAV pathophysiology with the development of ANCA antibody and subsequent activation of neutrophils by ANCA. The *zoom in* further shows that these activated neutrophils not only penetrate and damage the small blood vessel walls but also in turn activate the alternate complement pathway—an enzyme cascade critical to the vasculitis process—further amplifying inflammation and vascular damage.[6] The effect is a microscopic vicious cycle – the science is explained in the content of understandAAV with the art providing the visual reminder.

When this unusual and arresting visual was presented in market research to physicians against more traditional visualisations of a disease process, they overwhelmingly selected the understandAAV image as their favourite. This immediately became the centrepiece of the educational campaign. With understandAAV, physicians can gain knowledge of the complex pathophysiology of AAV, better understand existing treatments and their challenges, and gain insights into the experiences of real-world patients, all through the lens of a highly distinctive, impressionistic visual platform—a manifestation of science as art.

www.understandaav.com

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SEE ME HEAR ME

on myANCAvasculitis.com—a new initiative co-created with Pan-EU patient associations, patients and carers

From the initial diagnosis of ANCA-associated vasculitis (AAV) onwards, patients' experiences will vary, but as is often the case in rare diseases, confusion and questions about the disease are common

Based on extensive qualitative research into the patient experience and consultations with several vasculitis patient associations around Europe, Vifor Pharma set out to collaborate and *co-create* materials with patients, carers and patient association groups across Europe to support those living with, or affected by AAV. The aim of the initiative is to support the efforts of local patient groups and help fill any information gaps which may help patients and carers better understand and cope with the physical and emotional impact of AAV. These materials are housed on a dedicated digital platform—where visitors can also find links to local patient association groups: **myANCAvasculitis.com**.

Collaborating with Shanali Perera—physician, artist and vasculitis patient

As with the **understandAAV** information project aimed at physicians, this patient-focused initiative needed a visual *hook*. Thanks to an introduction by the Vasculitis UK patient association, the Vifor Pharma team met artist, physician and vasculitis patient Shanali Perera. Prior to her diagnosis, Shanali had been a practising rheumatologist. Suddenly, and first-hand, she was confronted with what she calls **"a truly humbling experience, both personally and professionally"** giving her valuable insights into what the patient's journey is really like. Forced to give up her medical career, through the creative process Shanali was able to regain control of a part of her life—a *turning point* she says.

A remarkable portfolio of patient portraits

Shanali, along with other AAV patients and carers took part in the inaugural co-creative patient workshop in Manchester, UK in May 2019, which is where the creative idea behind SEE ME. HEAR ME. began to evolve. A number of different concepts and names for the initiative were put to the panel of patients and carers: all felt strongly that this initiative should ***visualise*** and ***vocalise*** what lies behind such a complex and challenging condition. In the following months and in subsequent workshops, again in Manchester, then in Utrecht, Netherlands and Stuttgart, Germany, a further series of workshops took place where patients and carers came together to talk about their personal experiences of living with AAV and the gaps in information on AAV they could identify. The attendees also helped validate proposed new content for

development—for example, discussion guides for physicians to help them talk to newly diagnosed patients; a glossary of terminology to help clarify a complex disease area; infographics depicting what to expect along the journey from diagnosis to treatment.

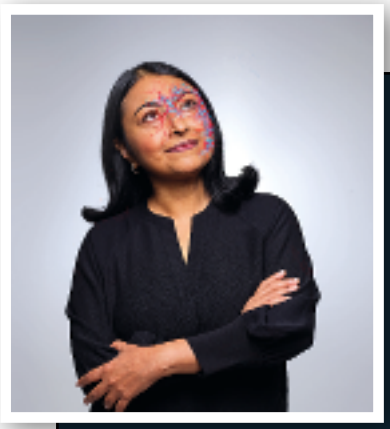
Each workshop also allowed for a photoshoot which has now turned into a truly distinctive portfolio—real AAV patients of all ages and different backgrounds. The beautifully realised photographic portraits present the patients as they are but overlaid on each image is one of Shanali's exquisite visualisations, depicting the inflammatory process that occurs beneath the surface.

At the centre of it all is the patient-focused information website, **myANCAvasculitis.com**, launched at the start of 2020 in English, German and Dutch, where alongside a wealth of information on AAV, you can find the personal case studies of each patient who has participated in this project, in PDF downloadable form and as a video interview.

Links to patient association groups are also provided so visitors can seek support in person and connect with others living with AAV in their local area.

This is only the beginning. The SEE ME. HEAR ME. initiative is continually evolving, and the aim is to reach out to other countries and other patient associations, all with the objective of helping empower patients living with AAV and their carers in feeling understood, seen and heard.

www.myancavasculitis.com



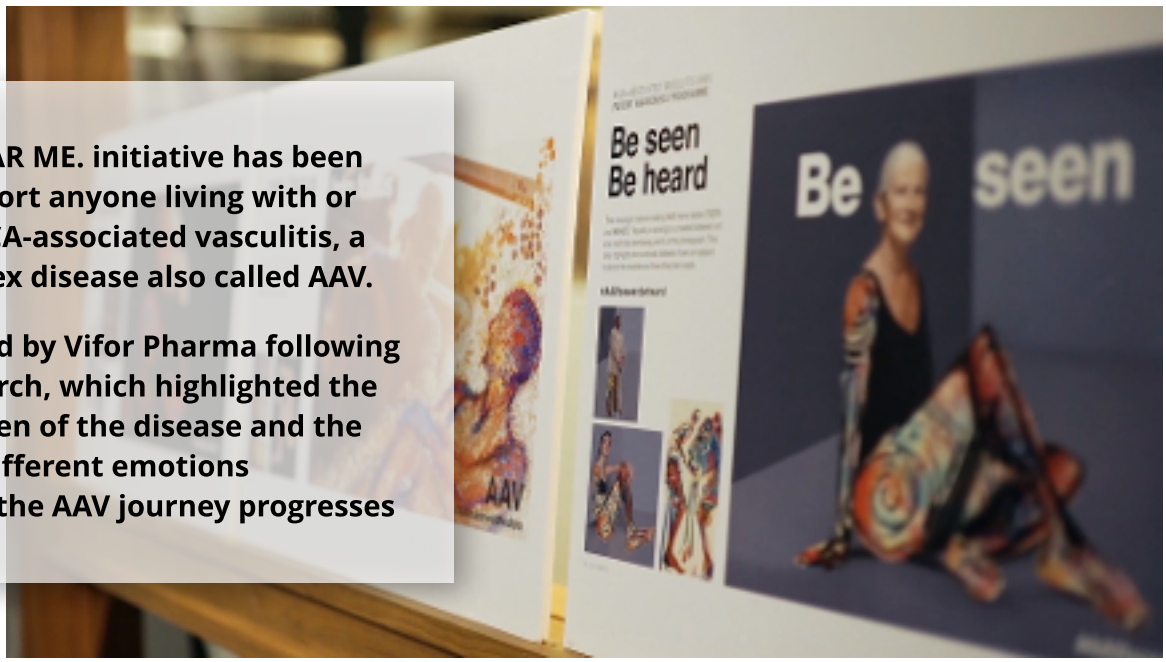
Shanali Perera's artistic talents and personal experience have combined to realise beautiful images illustrating the internal effects of vasculitis

Elisabeth NL Maresa DE Hermann DE Catherine UK John UK Hellen NL Rina NL Anne NL Wil NL Martina DE Iva NL Jeremy UK Klaus DE

SEE ME. HEAR ME.

a creative initiative collaboration

Shanali Perera



The SEE ME. HEAR ME. initiative has been created to support anyone living with or affected by ANCA-associated vasculitis, a rare and complex disease also called AAV.

It was developed by Vifor Pharma following extensive research, which highlighted the emotional burden of the disease and the wide range of different emotions experienced as the AAV journey progresses

"We are very grateful for the support of the patient associations across Europe. The patients and the carers that we had the opportunity to work with, but especially Shanali Perera who is not only an artist, but a rheumatologist by training and a vasculitis patient. She not only created the artwork for the SEE ME. HEAR ME. campaign but influenced the content so what we created is of meaning to the patients and the carers."

Dijana Krafcsik, director orphan diseases, Disease Awareness, Vifor Pharma

Shanali Perera—Art and Voice for AAV

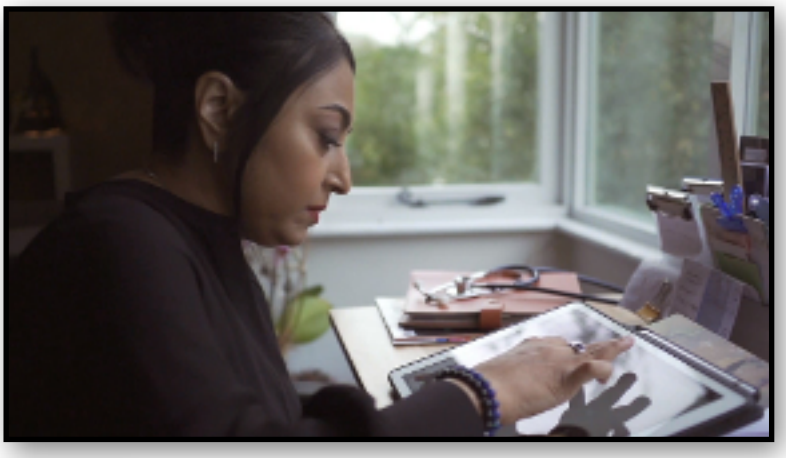
On being diagnosed with vasculitis

"Up until then, diagnosing, managing and treating conditions was more of a black and white reality for me. But going through the patient's journey was a reality with many shades of grey."

"I felt powerless. I could not control the direction my life was taking."

On giving up her career

"At that point I felt robbed. All my dreams and expectations had gone out the window and illness dominance really distorted my image, my identity, who I was as a person."



A new sense of purpose

"I could not control how my illness was progressing, nor my day, but I could control what I create which gave me a sense of purpose, a sense of achievement, helping me to regain some control and that certainly was a turning point for me."

thank you

"We would like to thank Shanali and the other patients, carers and patient association groups who have worked with us to make SEE ME. HEAR ME. a helpful resource for everyone affected by AAV." Vifor Pharma

***To find out more visit* www.myANCAvasculitis.com**

Martina's story

Martina is 29 years old and lives in Stuttgart with her two small children. She was diagnosed with ANCA-associated vasculitis, microscopic polyangiitis (MPA) with kidney involvement in 2019

Martina and her mother Lidia

This is Martina's story

Martina lost her brother six months before receiving her own diagnosis. Her motivation for taking part in this project is to share her story so that other patients with similar, unspecific symptoms might receive a diagnosis sooner.

"Nobody can tell us what my brother really died of, so I have to assume it was also a serious illness. It simply must not happen that so many people die of a disease just because the doctors react too late, somehow recognise things too late, diagnose too late."

The first symptoms Martina experienced were flu -like. She suffered with fevers, headaches and joint pain over a longer period of time.

"The time until the disease was recognised and until the diagnosis was very long. They underestimated all the flu symptoms."

Martina's mother Lidia was devastated by Martina's diagnosis.

"It was a shock for me—I just didn't believe it. I said right away, 'I lost my child ten months ago and now my other child needs help'."

When Martina began treatment it made her extremely bloated and her clothes wouldn't fit

"Me and my husband were shocked because Martina looked really bad and we didn't know what kind of disease it was. It was really hard."

"I just had to go in the waiting room and cry. Not in front of Martina."

Lidia

"I was honestly afraid of the diagnosis at first because I didn't know what it was. I was shocked. But I didn't have time to be shocked because the therapy started the very next day."

Right from the start Martina required a strong therapy to treat her condition. "The same night that I was diagnosed, I was given double steroids. The doctor warned me right away that I wouldn't sleep that night because steroids keep you awake. And I was awake all night. The next day, I looked really fat and bloated. My face was all swollen. I didn't even look like me anymore, nothing fit me anymore. I felt very uncomfortable and sick afterwards. Everything was totally messed up."

"In my everyday life I am affected by the disease over and over again. In the mornings everything works very well but over the course of the day it's like someone pulling a plug. I just feel totally tired, totally flat and I need at least an hour before I can somehow regain energy so that I can continue. In the past, I could just get dressed and go out, but it doesn't work like that anymore. I look for breaks; I make sure that I get to bed on time, because sleep is very important. I have completely changed my diet. I make sure I take my medication at the correct time every time.

"I pay much more attention to my body, to all the signs, and if I have a cough, then I have to go to bed all day and wait until it gets a little better."

"My professional life is affected by the disease. I was a kindergarten teacher for many years and really enjoyed working with children. But I'm not allowed to do that anymore, because under immune suppressants I simply don't have an immune system anymore and any infectious diseases from the children would simply be a too big a risk for me. That's why I have given up the job now."

"I connect with a lot of groups on the internet where people describe their own experiences, because it's real. I believe that nobody can give me more honest answers than the people themselves who go through the whole thing, who experience the disease.

They really understand what it is all about."

"It's important not to underestimate the symptoms. If you notice that something is different, you should go directly to the doctor and not let them turn you away. Unfortunately, it's often the case that the doctors say, 'This is a flu, why don't you go home and lie down,' but it's just not like that, you shouldn't underestimate it. "

"You explain it to somebody and often they say, 'Yes, but you don't look sick at all. You're in great shape. You look good. There is nothing wrong with you'. And the more you hear this, the worse it is for your own psyche."

"I support the project because I believe that more doctors need to be made aware of the disease, including different symptoms. Someone who comes down with flu symptoms doesn't necessarily suffer from a flu. It can also be something else."

Martina looking to the future

"It is very important to me that I see my children grow up, especially until they are 18.

...that I can prevent undergoing dialysis in regard to my kidneys, even in the next ten years.

...that I can lead a somewhat normal life, that I can do something with my friends, that I can go on vacation again with my family."

AAV information and resources—developed in collaboration with patient associations for patients

Three workshops in three countries and close collaboration with PAN-EU patient association leads led to a deeper understanding of how ANCA-associated vasculitis (AAV) affects patients' and carers' lives

Over the past year, Vifor Pharma has conducted three workshops in three different countries with the key objective of listening to patients and carers tell their side of the AAV story: how this rare disease affects their lives and their families, and what resources may help them better understand the condition. These workshops were only made possible through collaboration with local patient associations: Vasculitis UK; Vasculitis Stichting in the Netherlands; and two German patient organisations, Vaskulitis e.V. and Vaskulitis-Mainz.

The workshops took place in:

Manchester, England; Utrecht, Netherlands; and Stuttgart, Germany

Each group of patients and carers was shown visuals based on the concept of **SEE ME. HEAR ME.**—a theme embracing empowerment and the patient voice—and proposed content for the centrepiece of the initiative, a dedicated website to raise awareness, increase understanding and support AAV patients, their carers and families. The valuable feedback we gathered turned these workshops into truly co-creative efforts, with input on the visuals, the page content and downloadable assets. With the launch of **myancavasculitis.com** we now have a comprehensive platform in multiple languages with information on AAV, patient experiences, ideas to help daily self-management, as well as videos and personal patient stories available to view and download.

After initial introductions, the participants were invited to share their individual experiences. What came across in all groups was a desire for more information about AAV, whether from treating physicians or from online resources. Even though every patient's experience is different, it was broadly felt that giving guidance on what the journey ahead might hold was essential, especially for newly diagnosed patients.

A heartfelt thank you!

We would like to pass on our gratitude to all patient association groups, patients and carers who attended and contributed to these workshops, helping to create a website and suite of materials designed to inform and empower anyone living with or affected by AAV.

Vasculitis International

The international patient organisation joining forces to provide support, drive research and ultimately improve lives, by strengthening the voice of the vasculitis community, proving they are stronger together

Vasculitis International is the brainchild of Peter Verhoeven, chairman of Vasculitis Netherlands, and John Mills, chairman of Vasculitis UK. In 2002, Peter's wife was diagnosed with granulomatosis with polyangiitis (GPA)—an auto-immune disease formerly known as Wegener's granulomatosis. John too received a diagnosis of GPA in 2001

Firm friends John Mills and Peter Verhoeven

The pair met in 2013 at a vasculitis conference where the idea of joining forces and strengthening their collective voice was born.

"The motivation behind setting up Vasculitis International was realising that vasculitis, like any other disease, does not care about geographical borders," says Peter

Vasculitis Netherlands and Vasculitis UK are amongst the bigger, further established support groups which led to these two groups leading the combined initiative. "We realised that the more volume of data relating to patient experience you can jointly create, the more impact you can have on things such as research and quality of care," notes Peter. And by uniting they were quickly able to support research projects through their collaboration.

Promoting unity

Peter acknowledges that Vasculitis International is still a fairly informal group of vasculitis patient advocacy groups (VPAGs) and it is very much in its infancy. But it is an exciting time as the group begins to transform into a more professional organisation; it was recognised as an international organisation under Dutch law in 2019.

Vasculitis International started life as Vasculitis Europe, but as John states, "We didn't want to deny other countries and become an exclusive club." It is important to look beyond Europe. John notes that in some countries the condition is virtually unknown and about 100 years ago wasn't recognised in Europe.

"The most important part of the process of dealing with a rare disease is recognition," says John

In time Peter and John plan to reach out to other countries, especially those who are most in need of support, by providing patient-focused resources.

A challenge faced by the group is where countries do not have one distinct and comprehensive patient support group to reach out to. The language can also be a barrier to communicating and collaborating. Patients who have an understanding of English can access the UK or Ireland support groups, but it is those who do not speak the language that Peter and John want to reach out to in particular. And they feel it is paramount to provide materials in the local language to ensure inclusivity.

The group currently disseminate information to the individual VPAGs through the website and use social network and online communication tools to exchange information. They are yet to meet formally as Vasculitis International, but plans are underway to start conducting teleconferences along with hosting an annual meeting.

Stronger together

While both Peter and John admit the organisation is still finding its feet, by joining forces, Vasculitis International has already seen projects come to fruition through their collaborations. The VPAGs from Ireland, the UK, and the Netherlands, represented by Peter, John, and Julie Power from Vasculitis Ireland, took part in the FAIRVASC project, under the umbrella of Vasculitis Europe. The project aims to bring together leading scientists, clinicians, and patient organisations, and, using web technologies, will combine vasculitis registries across Europe into one single European dataset.

The VPAGs from Ireland, the UK, the Netherlands, France, Belgium, Finland, and Germany are currently working together to co-create and collaborate with pharmaceutical company Vifor Pharma in a joint effort to increase awareness for ANCA-associated Vasculitis (AAV). The digital and printed materials will be used for the digital platforms, **myancavasculitis.com**—primarily for patients and carers, and **understandavv.com**—primarily for healthcare professionals outside the US only. Due to the symbiotic nature of the collaboration, the VPAGs involved can also use all the material for their own organisations.

Another important European initiative is the European Reference Networks (ERNs). Vasculitis is a part of the ERN-RITA (European network on rare primary immunodeficiency, autoinflammatory and auto-immune diseases). Because of their collaboration, Vasculitis International/Europe were able to readily identify candidates to represent the vasculitis community on the board of the ERN-RITA patient community, and on the ERN-RITA board itself.

John and his wife, Susan

Peter and his wife, Elisabeth

Nurturing partnerships

Peter sees the benefit of coming together as the larger, more established, funded groups being able to support the smaller, less established organisations, most importantly as equal partners. By combining these organisations and collaborating, a greater volume of patient experiences, contacts, and expertise is collated, strengthening the impact and voice of the vasculitis community.

Hope for the future

Peter and John both want to see Vasculitis International continue to grow into an established, professional, internationally recognised organisation, providing a wealth of resources to the vasculitis community. Their aim is to continue collaborating and sharing information and experience with other VPAGs, and to further develop joint projects with all stakeholders in the field.

www.vasculitisint.com
p.verhoeven@vasculitisint.com

PAN-EU LOCAL VASCULITIS PATIENT ASSOCIATION GROUPS

Vasculitis UK is the national patient support group for those affected by all types of vasculitis and their familes by providing information, advice and support.

Vasculitis UK aims to raise awareness of vasculitis among medical professionals and the public and facilitates mutual contact and support by means of local support and online discussion groups. Vasculitis UK informs and educates people with vasculitis about their illness so that, working with their medical professionals, they are empowered to take greater control of it.

Vasculitis UK supports research into the causes and effects, diagnosis and treatment of vasculitis.

www.vasculitis.org.uk
info@vasculitis.org.uk

United Kingdom

The Dutch Vasculitis Stichting supports individuals with a primary vasculitis and their loved ones. Through a variety of initiatives, we make an ultimate effort to shorten their time to diagnosis, optimise their treatment and improve their quality of life.

www.vasculitis.nl
info@vasculitis.nl

Netherlands

We are an all of island **Ireland Support Group**, run by those affected by vasculitis, for people with any of the vasculitis diseases, their carers, and healthcare professionals.

Our aims are to support each other, assist research into improving current management and treatment, and increase awareness of vasculitis.

www.vasculitis-ia.org
julie@vasculitis-ia.org

Ireland

The patient association **France Vascularites** listens, supports, helps and guides patients and their relatives in France and French-speaking countries. It creates local links and develops tools to reduce isolation and fight against delays and diagnostic deadlock. It supports medical research by all the means at its disposal.

To know your illness better is to live with it better every day.

www.association-vascularites.org
association.vascularites@gmail.com

For Belgium contact details please visit;
www.association-vascularites.org

France/Belgium

PAN-EU LOCAL VASCULITIS PATIENT ASSOCIATION GROUPS

Vaskulitis e.V.
beraten · informieren · aufklären

We, the **Patient-Association Group Vaskulitis e.V.** located in Germany, provide a platform to exchange information and experiences and offer support for all social and health questions concerning the disease. We organise medical lectures and arrange contacts with expert healthcare professionals and clinics. We offer a wide range of information material about vasculitis.

Affected people helping affected people; ending isolation.

www.vaskulitisverein-rlp.de
info@vaskulitisverein-rlp.de

Germany

Gefäß VASKULITIS Entzündung

We are the self-help group called **Vaskulitis Mainz** in the area of Mainz and surroundings; we support people of different ages who suffer from different types of vasculitis.

vaskulitis-mainz@gmx.de

Germany

APACS
Associazione Pazienti Sindrome di Churg Strauss

APACS is an Italian organisation aiming to help patients affected by EGPA (formerly Churg-Strauss syndrome). They cooperate with practitioners to spread awareness, offer advice and improve healthcare assistance for patients and their families.

www.apacs-egpa.org
infoapacsegpa@gmail.com

Italy

Suomen Vaskuliittiyhdistys ry

Suomen Vaskuliittiyhdistys ry is a nationwide association in Finland that aims to be a connecting partner for people with vasculitis, improving the quality of life, promoting research and care, and supporting families. The association has active co-operation with other associations representing rare diseases, The Finnish Rheumatism Association and Eurordis.

As a small association we are very happy to have the opportunity to network with other associations and parties representing rare diseases and especially vasculitis.

www.vaskuliittiyhdistys.fi
vaskuliittiyhdistys@gmail.com

Finland

A RARE connection

Patient exchange through digital and social media to cope with ANCA-associated vasculitis

Martina is 29 years old, from Stuttgart, and was diagnosed with microscopic polyangiitis (MPA) with kidney involvement in January 2019.

Maresa is 19 years old, from Nordhorn, and was diagnosed with granulomatosis with polyangiitis with lung and kidney involvement in March 2019.

Martina and Maresa met through social media, first connecting on Facebook and then following each other on Instagram. They provide each other with invaluable support through their shared experience of ANCA-associated vasculitis.

"We basically wrote to each other every day. How we are, what we are doing, what our illness is doing, whether we have had any changes in medication, in therapy. It was always nice." *Martina*

"It is everything to have someone who always understands you and who knows how you feel when you are not doing so well, and shares in the good days. We are always there for each other and can talk about everything." *Maresa*

Friendship at first sight

"It was actually friendship at first sight. We actually understood each other immediately." *Maresa*

The two women offer such support and understanding, and the insight that can only come from having the condition.

"When we text at night or send each other voice messages, then we know immediately how the other one is doing, whether you're trying to hold back tears. That is when you notice just how much this whole thing affects your psyche. With Maresa I don't have to hide my feelings as I know that often she is feeling the same way." *Martina*

Martina and Maresa discuss the side-effects of steroids, both the psychological and physiological effects; the bloating, the mood swings, the binge eating. No explanation is needed between the two women because they just *get it,* something "many other people simply cannot understand because they do not have this condition". *Martina*

While the two may be separated by physical distance, they are united by the virtual world, and are in constant contact.

"We are both in the Facebook group and you can ask anything there. No one ever says anything stupid or something that doesn't belong there, they are all very helpful and there for each other." *Maresa*

"All questions are taken seriously. No matter if it is little things about eating, about nutrition, about the condition, about any small side effects, about a cold. They're always very positive and want to help you." *Martina*

The future

"We both hope that both of us are doing well and that we can always be there for each other." *Maresa*

Patient stories—capturing the unique and individual patient experience across Europe

Patients from across the UK came together in Manchester to take part in Vifor Pharma's inaugural co-creation workshop. This presented a unique opportunity to gain an insight into the individuality of the AAV journey, with patients sharing their experiences around diagnosis, treatment and relapse. The collective outputs from this initial workshop went on to form the basis of the SEE ME. HEAR ME. campaign

Jeremy

Jeremy has microscopic polyangiistis (MPA). He enjoyed excellent health up until his late 50s, at which point he had heart trouble and required heart bypass surgery.

Primary disease presentation: Kidneys, skin.

First symptoms: A loss of kidney function was found when his doctor was checking for coronary heart disease. He was sent to a kidney specialist.

Delay to diagnosis: It took 10 months to be diagnosed with MPA due to a delay in seeing the appropriate specialists.

Initial treatment: Prescribed cyclophosphamide (an immunosuppressant) and enrolled in a clinical trial (unaware of the treatment received).

Maintenance therapy: Azathioprine (an immunosuppressant) started six weeks after initial treatment. Remained on azathioprine for four years. Experienced flares once or twice a year and treated with high-dose steroids (glucocorticoids) each time. Now on rituximab and no flare for over a year.

Patient support groups:

"There's a very excellent website called Vasculitis UK which is worth looking at; it gives you all sorts of reliable information about ANCA vasculitis."

Personal advice:

"Make sure you get the maximum possible information from the specialist... But don't be put off by what they say."

"Whatever the case, just keep positive."

Jeremy

Catherine

Catherine has eosinophilic granulomatosis with polyangiitis (EGPA). She lives in the UK and works part-time in communications for the government. Her cousin has recently been diagnosed with GPA.

Primary disease presentation: Lungs, nerves, heart, allergies.

First symptoms: Flu-like symptoms, headaches and tiredness. Was diagnosed as a viral infection despite three visits to the GP.

Delay to diagnosis: It took one year from first symptoms to ending up in A&E. Diagnosed with EGPA.

Treatment side effects: Struggled with steroids. Couldn't focus. Highs and lows of adrenaline all day and night. Suffered panic attacks after each cycle of cyclophosphamide and had to go to the emergency doctors.

Side effects of treatment:
"I found concentrating and focusing really difficult on high doses of steroids. I was pretty ill after each cycle of cyclophosphamide—a lot of it was panic because it set off all kinds of reactions in my body and it was like suffering from panic attacks."

Long-term impact of ANCA vasculitis:

"I have to pace myself and rest more often. I certainly adjust my social activities, if I am going out for the evening I often have to sleep for an hour during the day."

Catherine

ANCA-ASSOCIATED VASCULITIS

John

John has granulomatosis with polyangiitis (GPA). He is the head of the charity Vasculitis UK and a founding member of Vasculitis International. He lives with his wife and carer, Susan, in the UK.

Primary disease presentation: Kidneys, lungs, sinuses, joints.

First symptoms: In November 2000, John suffered from sinus problems and his ankles were aching.

Early progression: When skiing, he lost control of his legs and skis, continued with his life afterwards. A few days after doing repair work on his house John began coughing up blood. The doctor was unsure of the condition and John quickly became bedridden requiring oxygen.

Delay to diagnosis: Six months from first symptoms to ending up in hospital. Six weeks from coughing up blood. Had a lung biopsy and diagnosed with GPA in April 2001.

Struggles with fatigue: Used to go home and go to sleep directly after work. Felt that he had to sit down or he would fall down. All the strength felt drained from his muscles and his brain felt like it wasn't working very well.

Personal advice:

"Well I share my experiences, my knowledge with people on a daily basis and it's difficult: I can't say anything specific because each person is different."

John

Being a team:

"I think the main thing that got us through was—I know it's a bit soppy—love and a good sense of humour."

John & Susan

In partnership with Peter Verhoeven and Vasculitis Stichting, a total of 10 AAV patients and carers came together in Utrecht, Netherlands to share their experiences and build on the SEE ME. HEAR ME. campaign. While reinforcing the unpredictability of the patient journey, the workshop also outlined the significant challenges faced by carers on a daily basis

Wil

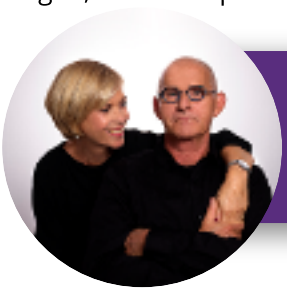
Wil has granulomatosis with polyangiitis (GPA). He lives together with his wife Marleen in the Netherlands.

Primary disease presentation: Ear, nose and throat.

First symptoms: Couldn't breathe through his nose, particularly at night. Had to breathe through his mouth and repeatedly woke up feeling thirsty, contributing to fatigue.

Delay to diagnosis: Went to the doctor but was not diagnosed with GPA at the time. Symptoms stayed the same until diagnosis was made by a specialist in 2001, approximately 18–24 months after he first visited the doctor.

Impact on life: The biggest problem has been the disease/treatment-induced fatigue, and its impact on his ability to work.



Disease awareness:

"I cannot talk about it with friends and family, because there has never been anybody in the family that has had the disease."

Marleen

Elisabeth

Elisabeth has granulomatosis with polyangiitis (GPA). She and Peter live in the Netherlands; they have been married for the past 53 years. Peter is chairman of Vasculitis Stichting and a founding member of Vasculitis International.

Primary disease presentation: Ear, nose and throat, lungs.

Problems before diagnosis: Had problems over a period of two years. Main issues were tiredness and a series of ear infections, eye infections and nose problems.

Diagnosis and treatment: Was diagnosed with GPA following referral to a specialist. Received steroids (glucocorticoids) and chemotherapy.

Steroid side effects: Her appetite increased substantially. Gained weight and found the changes in her appearance/weight challenging psychologically.

Treatment side-effects

"I don't think I was well educated by my doctor, because you gain weight with steroids. I was size 34 when I got GPA and I came back home a size 42."

Elisabeth

ANCA-ASSOCIATED VASCULITIS

Iva

Iva is 17 years old and has eosinophilic granulomatosis with polyangiitis (EGPA). She lives in the Netherlands with her parents.

Primary disease presentation: Face.

First symptoms: Had an unexplained fever, became pale in the face and had less energy than before. Her doctor carried out blood tests but was unable to find anything. Iva lost control of some facial features, and her mouth started to hang open. She was referred immediately to the regional hospital.

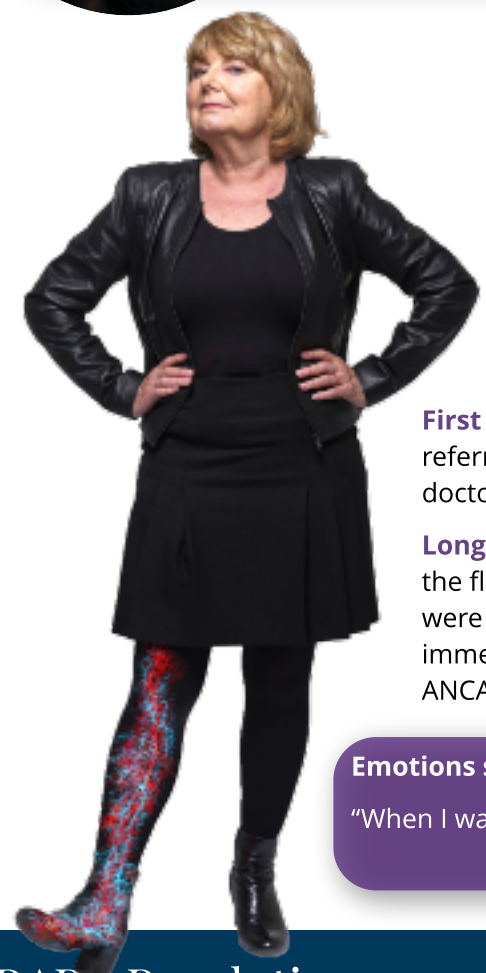
Diagnosis: After five months of uncertainty, developed sensitivity to light, sound and smell during her stay in regional hospital. She was diagnosed within five–six weeks.

Living with AAV: Fatigue and the risk of infection are key concerns. Always a sporty girl, Iva now only plays half matches of hockey and has moved to a lower level team as she could not keep up.

Parents' perspective:

"We're very worried about her future. We worry whether she will be able to get a full-time job, whether the disease will come back... We always have worries in the back of our minds."

Birgitta



Rina

Rina was diagnosed with microscopic polyangiistis (MPA) in 2004. She lives in the Netherlands with her husband and has two adult sons.

Primary disease presentation: Neurological failure symptoms, joint pain, spots on the lungs, bleeding under the nails.

First symptoms: From 1995 onwards had constant colds and joint pain. Was referred to a pulmonologist, a rheumatologist and an ear, nose and throat doctor, but her ANCA vasculitis was not recognised.

Long road to diagnosis: In 2004 she got up one morning and collapsed on the floor and was admitted to hospital. Her kidney function was poor, there were spots on her lungs and bleeding under her nails. The rheumatologist immediately thought of ANCA vasculitis. Diagnosis was finally confirmed after ANCA tests, nine years after her first visit to the doctor.

Emotions starting treatment:

"When I was hospitalised, I was very afraid I wouldn't make it."

Rina

ANCA-ASSOCIATED VASCULITIS

Anne

Anne has granulomatosis with polyangiitis (GPA). She lives in the Netherlands.

Primary disease presentation: Ear, nose and throat, lungs, joints.

First symptoms: In March/April 2015, flu-like symptoms, sinus inflammation, inner ear inflammation, crusts inside the nose. Doctor prescribed antibiotics, but they did not work. When she almost lost all hearing in one ear, she went to hospital. Sent home with painkillers and nose drops.

Diagnosis: When steroids (glucocorticoids) stopped, she started feeling worse again. Admitted to hospital. Doctor recognised the initial symptoms and suspected vasculitis. They did some tests including a biopsy from the nasal mucous membrane. Diagnosis of ANCA vasculitis received in June 2015.

Relapse: After a change of medicine three months into the treatment, a flare caused narrowing of the windpipe. Windpipe had to be stretched and rituximab was added to treatment.

Personal goal

"I really like to focus on the positive things and try not to let ANCA vasculitis hold me down."

Anne

Hellen

Hellen has granulomatosis with polyangiitis (GPA). She lives in the Netherlands with her husband Frank, and their three children.

Primary disease presentation: Lungs.

First symptoms: Woke up in the morning struggling to breathe, had difficulty walking up the stairs.

Journey to diagnosis: Diagnosis was finally confirmed following a biopsy, around five–six months after her first visit to the doctor. Experienced multiple headaches, joint pain and fatigue prior to diagnosis.

Specialist support: Has seen multiple specialists over the years including a lung specialist, an internist, an ear, nose and throat specialist, a rheumatologist, and an ophthalmologist.

Impact on daily life

"In the beginning it was difficult. Slowly I could do more and more things again, but still not enough to go to work or things like that. Then you adapt, so going out less often. Now the day is actually calm and I do a bit of volunteer work."

Hellen

Patients and carers came together in Stuttgart for a workshop and shared their compelling personal stories of being diagnosed and living with ANCA-associated vasculitis. Each patient had a very different tale to tell, emphasising the individual nature of the AAV patient experience

Hermann

Hermann was diagnosed with granulomatosis with polyangiitis (GPA) in 2005. He lives with his wife in Germany and runs the patient association group Vaskulitis Mainz.

Primary disease presentation: Ear, nose and throat (deafness in left ear), joint pain, fatigue.

First symptoms: What was initially thought to be middle ear inflammation quickly turned into almost complete deafness.

Diagnosis: An MRI scan was carried out and his eyes were examined; various blood tests were performed which showed increased ANCA values leading to the diagnosis of ANCA vasculitis.

Relapse: In autumn 2006, he was admitted to hospital and given high-dose steroids (glucocorticoids). Over the next six months he was given extremely high doses of steroids (glucocorticoids) which caused bloating and sleeplessness.

"In the beginning, the illness was associated with very great uncertainty as well as a lot of fear. I had no contact with other patients at all for many years so I was unable to talk about my experiences. And the information I found online wasn't really helpful."

"Together with the group I want to give people with ANCA vasculitis the opportunity to exchange experiences."

Hermann

Klaus

Klaus is 62 years old and was diagnosed with granulomatosis with polyangiitis (GPA) in 2008 after three years of worsening symptoms. He lives in Germany with his wife Heike and runs the patient association group Selbsthilfe Vaskulitis e.V.

Primary disease presentation: Kidneys, lungs, limbs.

First symptoms: Bloody cough, followed by pain in the limbs and swollen legs. Liver function was very poor. After three weeks symptoms subsided but soon returned with pain in the legs.

Diagnosis: Diagnosed with GPA in 2008 after three years of worsening symptoms. Klaus was 50. His wife Heike called an ambulance as his legs had become so swollen he couldn't walk. He was taken to hospital and when his kidneys started failing and he was transferred to a specialist hospital where GPA was diagnosed within an hour.

Effect of treatment: After 11 years of steroid (glucocorticoids) treatment, an MRI revealed an accumulation of fatty tissue behind his sternum. Steroid (glucocorticoid) treatment was discontinued.

Long-term effects of ANCA vasculitis:

"Klaus' quality of life is very, very limited. He used to walk or hike a lot. Unfortunately, he can't do that any more. Today he walks on crutches and it's slow progress if you want to go somewhere."

Heike

Personal advice:

"My advice for new patients, or anybody affected, is that they understand their disease. New patients practically stand in front of an invisible wall with so many question marks. They want these questions answered to understand what is going on in their body. I give them help and advise where they can receive further support."

Klaus

Maresa

Maresa was diagnosed with granulomatosis with polyangiitis (GPA) in 2019. She and her boyfriend and carer, Kai, are from Germany.

Primary disease presentation: Kidneys, lungs.

First symptoms: Maresa was constantly sick for two to three years with flu-like symptoms, tonsillitis, liver pain, fever and then eventually started coughing up blood.

Diagnosis: Five days after being hospitalised her lung collapsed; she was rushed to a specialist hospital to identify the cause. She was in an induced coma whilst she was examined by specialists from each department and then diagnosed with GPA.

Effect of treatment: Maresa underwent a significant change of appearance and suffered a heavy psychological and physical burden as well as fatigue.

Impact on her life: Passed police force entrance exam at age 19, but after being diagnosed with ANCA vasculitis just one month later, she had to give up her dream job. Is currently not working or studying due to symptoms such as fatigue.

Initial side effects of treatment:

"When I got home, we removed all the mirrors, because after the steroids I felt so disgusting that I couldn't look at myself any more."

Maresa

Personal advice:

"You have to set new goals. It is also quite normal to feel sad. But little by little, it will even out again. Then the sad moments become fewer and the beautiful moments more."

Kai

The typical ANCA-associated vasculitis patient journey

Emotional and psychological journey

DIAGNOSIS

1

There may be a long period of frustration and anxiety preceding diagnosis. Once diagnosis is confirmed, some patients may experience relief that the disease has been identified, while others will experience significant shock.

STARTING TREATMENT

2

This can be a period of some confusion, uncertainty, fear and fatigue. Patients can experience distress and isolation during the initial intensive treatment period. However there may also be relief that action is being taken.

GETTING TO REMISSION

3

Once undergoing treatment for remission, there may be issues to consider and discuss with one's physicians. At this stage it's also important to start thinking about a new life balance because changes may be needed. Personal relationships, work life and income, may all come under strain.

REMISSION AND RELAPSE

4

Remission can be achieved and maintained, but there may also be flares and relapses so it's important to look out for signs and be aware. Over time it may be unclear whether periods of feeling worse are down to the disease, the medications or simply getting older.

KEY CHALLENGES

5

The rare nature of the disease and the associated lack of awareness among the wider public is an additional problem. This can be addressed through more information and education; friends and family can also help increase understanding and awareness of the condition.

SEE ME
HEAR ME

#myANCAvasculitis

1. *De typische patiëntenreis bij ANCA-vasculitis*. Vasculitis Stichting N... 2. Rutherford P, et al. *Arthritis Rheumatol* 2016;Suppl 10[Abstract 2723].

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THANK YOU

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