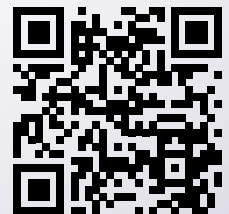


SEE ME HEAR ME

The AAV Journey

A booklet for people living with **ANCA-associated vasculitis (AAV)**, and their families and friends



myANCAvasculitis.com/uk

See Me Hear Me is a disease awareness campaign that has been created and funded by CSL Vifor.

Contents

The patient journey	3–5
<ul style="list-style-type: none">• Emotional and psychological patient journey	
<hr/>	
Diagnosis and clinical management	6–12
<ul style="list-style-type: none">• Testing in AAV• What kinds of doctors could be involved in managing your AAV?• Tips when receiving treatment• What does remission mean in AAV?	
<hr/>	
Self-management	13–16
<ul style="list-style-type: none">• Questions to ask at your medical appointment• How can I cope with AAV?	
<hr/>	
Glossary	17
<hr/>	
References	18

Disclaimer: Although every effort has been taken to ensure the accuracy of information in this booklet, it is not intended to be a substitute for professional medical advice, diagnosis, or treatment. If you have any unanswered questions about AAV or your treatment, please speak to your specialist medical team.

THE PATIENT JOURNEY



Shanali,
Vasculitis patient,
rheumatologist,
UK

Emotional and psychological patient journey

Each patient's journey with AAV is different. However, there are some key milestones commonly experienced from diagnosis to ongoing treatment.

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.¹

2

Starting treatment

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.^{1,2}

Getting to remission

3

Once undergoing treatment for remission, there may be issues to consider and discuss with your 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.²

4

Remission and relapse

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.^{3,4}

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.

DIAGNOSIS AND CLINICAL MANAGEMENT



Jeremy,
MPA patient, UK

Testing in AAV

Your doctors will have to perform a combination of tests to support your AAV diagnosis, but some of these tests may also be repeated throughout your journey, to monitor your disease activity.^{3,5}

Blood tests

ANCA tests

The presence of ANCAs (auto-antibodies that attack the body's own cells) in your blood may help your physician diagnose one of the three types of AAV (GPA, MPA and EGPA), and determine a prognosis.^{6,7} However, they are not perfect – a negative ANCA result doesn't rule out an AAV diagnosis, and a positive ANCA result doesn't mean a patient has AAV.^{6,7}

Kidney function

If your doctors suspect you may have kidney disease, they may investigate how well your kidneys are functioning with a blood test.⁵ They may look for levels of urea (the breakdown product from protein in the urine) and creatine (the protein from muscle breakdown) in your blood.⁵

Inflammatory markers

Signs of inflammation (called inflammatory markers) can be detected in the blood. These may be tested during diagnosis and at follow-up clinic visits to monitor disease activity.^{3,5} Your doctors may look at how fast the particles in your blood move in a test tube (blood cell sedimentation rate) or the levels of a specific type of protein produced by the liver, which are higher when inflammation occurs (C-reactive protein).^{5,8}

Full blood count

A full blood count may be completed to confirm diagnosis or at follow-up appointments.^{3,5} They measure the amounts of different cells in your blood, which may indicate to specific signs of AAV, such as anaemia due to inflammation.⁵

Biopsy

A biopsy is a procedure where a small piece of tissue is taken from an organ to examine under a microscope for signs of AAV. Kidney or lung biopsies may be taken by speciality services to confirm a diagnosis of AAV or exclude other diagnoses such as cancer.⁵

Urine tests

Urine tests are a quick and easy way to see whether inflammation from vasculitis is present in the kidneys for diagnosis. The tests usually look for blood and protein in the urine. During follow-ups, patients should have further urine tests to look for infection, signs of relapse and how well you are responding to treatment.^{3,5}

Imaging procedures

Imaging allows healthcare professionals to look inside different areas of the body, to determine whether they've been impacted by AAV. Examples include X-rays and CT scans on the chest to search for damage.⁸

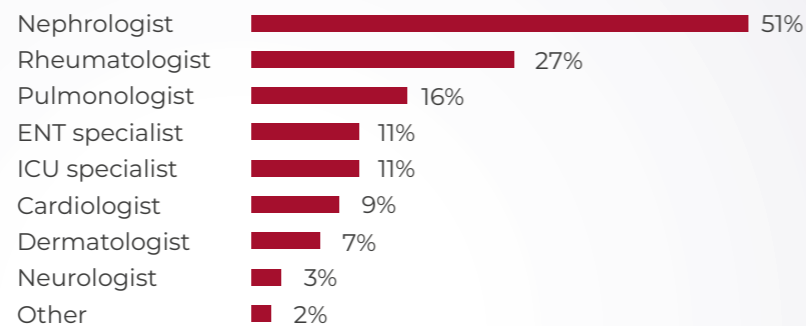
What kind of doctors could be involved in managing your AAV?

As AAV is a disease that affects multiple systems of the body, patients are often treated by a multidisciplinary team of doctors and nurses.^{3,5,9} A variety of different specialists may be involved in your treatment at different stages of your AAV journey.



17% of patients are managed by only one physician for AAV.

Other physicians involved:
(other than main managing physician)



Adapted from: Rutherford P, et al.¹

Some of the specialists that may be involved in your treatment include:¹⁰

Rheumatologist: A doctor who specialises in the diagnosis and management of diseases affecting the **muscles, bones, joints, ligaments, and tendons**

Nephrologist: A doctor who specialises in the diagnosis and management of diseases affecting the **kidneys**

Pulmonologist: A doctor who specialises in the diagnosis and management of diseases affecting the **respiratory system (nose, mouth, lungs)**

Otolaryngologist: Also known as **ENT doctors**, they specialise in the diagnosis and treatment of diseases affecting the **ears, nose and throat**

Cardiologist: A doctor who specialises in treating diseases of the **cardiovascular system (heart and blood vessels)**

Dermatologist: A doctor who specialises in disorders of the **skin, hair and nails**

Neurologist: A doctor who specialises in treating diseases of the **nervous system (nerves, spinal cord, brain)**

As well as doctors, nurses play an important role in managing your inflammation and pain, and also assisting doctors throughout diagnosis and treatment.¹¹ Nurses are also there to help with patient education, and providing emotional support for you and your family.¹¹

Why would I need to see a specialist?

The symptoms and signs of AAV can affect many different areas of the body, from the kidneys and lungs to your ears or nervous system.^{3,5} Although your primary doctor will oversee most of your treatment and disease management, sometimes it may be necessary to bring in a specialist for symptoms specific to an organ or organ system.



Catherine,
EGPA patient, UK

Ongoing clinic visits

Ongoing clinic visits allow your doctors to check that your disease is under control and monitor you for any signs of **relapse** or new organ involvement.³



Urine testing is typically performed at each visit to screen for infection, kidney problems or bladder complications. Every 1-3 months your inflammatory markers and renal (kidney) function will be measured to monitor your disease.³



You will also have blood tests to monitor blood count and liver function so your doctors can see how your medication is affecting your body. Patients on glucocorticoid therapy should also have their blood glucose (how much sugar you have in your blood) measured periodically.³



If you have a **relapse**, you will start **therapy** to induce remission again.¹²

Actively monitoring your AAV between visits can help you better understand your disease and spot the signs of any possible **relapses** as quickly as possible. Some people do this by keeping a diary to track their symptoms. If you decide to keep a diary, you can use it to discuss your AAV with your doctors.

You can write in your diary each day or just use it when your circumstances change. Useful topics you can include are your symptoms, treatment, side effects, and AAV's impact on your everyday life, as well as any other health issues that seem unrelated to the disease.

You can also write out questions for your doctor along with points for discussion at your next appointment.

If your doctor recommends it, you may also wish to actively monitor your AAV by checking your blood pressure regularly (such as once a week) using a blood pressure monitor. Get in touch with your doctors if you notice ongoing changes.

Tips when receiving treatment

AAV treatments can have many side effects, so here are some tips designed to help your body be in the best condition possible when receiving treatment.¹³⁻¹⁵



Minimise situations where infections may be more common. When attending appointments in clinics or hospitals ensure you protect yourself with a face mask and disinfectant



Try to decrease stress through activities such as meditation or yoga



If you smoke, try to stop (your doctor will be able to tell you if any services to help you stop smoking are available in your area)



Make sure to keep within sensible alcohol limits. This would be no more than 3-4 units of alcohol per day for men, and no more than 2-3 for women. One or two days a week should be alcohol-free



Stay up-to-date on your vaccinations, and discuss with your doctor what vaccines are suitable for you

What does remission mean in AAV?

Remission in AAV means that there is no detectable inflammatory disease activity. The term 'remission' implies that the disease is not cured, and you could experience relapses in the future.⁴

- **Remission** is defined as the absence of typical signs, symptoms or other features of active AAV with or without immunosuppressive therapy¹²
- **Sustained remission** is defined as the absence of typical signs, symptoms or other features of active AAV over a defined time period with or without immunosuppressive therapy¹²

It is important to remember that despite being in remission, it doesn't mean you will necessarily feel perfectly well. You may still require treatment and experience ongoing symptoms, particularly pain and fatigue.⁴ This is because symptoms can be caused by active disease, organ damage due to the disease or side effects from treatment. For example, even if you are in complete remission, you may still experience symptoms due to accrued organ damage or medication side effects.³

If you are experiencing any symptoms, it is important to contact your doctor immediately so they can examine you and give you the most appropriate treatment.

For more information on remission in AAV, visit myANCAvasculitis.com/uk/what-is-aav

SELF MANAGEMENT



Iva, EGPA patient and Birgitta, mother/carer, Netherlands

Questions to ask at your medical appointment

Many people find that asking questions helps them to better understand their disease and feel more confident about managing their AAV. You may find it useful to prepare for medical appointments by making a list of questions you want to ask and bringing a pen and paper so you can note down the answers.

What type of AAV do I have?

When and how will you be checking my progress?

Is my condition currently organ- or life-threatening?

Who should I contact if I feel unwell?

What signs or symptoms should I be aware of to spot a potential relapse?

Are any clinical trials available for me to join and, if so, what are the benefits and risks?

Are there any patient support groups I can join to help me manage the impact of my diagnosis?

Are counsellors or other professionals available to discuss the emotional impact of my AAV?

What do people in my situation usually do about work?

What short-term goals may be realistic for me to work towards?

What long-term goals may be realistic for me to work towards?

How can I cope with AAV?

Everyone reacts differently to being diagnosed with AAV and there is no right or wrong way to feel. It may affect you physically and emotionally, and impact your personal and professional relationships in a variety of ways.

AAV can affect people's everyday lives in many ways. Whatever you are feeling, you don't have to cope alone. Your doctors will be able to advise you on how to cope with the impact of AAV.

Find a detailed list of tips at myANCAvasculitis.com/uk/living-with-aav

What goals do you want to aim for?

Use this space to note down any goals you would like to discuss with your doctor.

Where do you see yourself in...

3 Months? _____

6 Months? _____

1 Year? _____

You may also wish to seek support by contacting a local patient support service or talking to your doctor about what help is available to you.

How can goal setting help?

Many people with AAV find that setting goals helps them become a more active participant in their disease management. Realistic short- or long-term goal setting is important to avoid disappointment or frustration.

Your doctors will be able to discuss which goals might be suitable for you, based on factors such as your disease, treatment, and personal, social or professional ambitions.

Glossary

AAV ANCA-associated vasculitis

ANCA Stands for anti-neutrophil cytoplasmic antibody

ANCA test A type of blood test used to detect the presence and level of ANCA within your blood

Autoimmune disease Occurs when the disease is caused by the body's own immune system. This autoimmune reaction (or 'attack') triggers the development of the disease

Biopsy A medical procedure where a small sample of tissue is removed from the body so it can be examined under a microscope

Cardiologists Doctors specialising in the cardiovascular system

Dermatologists Doctors specialising in the skin, hair and nails

EGPA Eosinophilic granulomatosis with polyangiitis, previously called Churg-Strauss syndrome, is a type of AAV that often affects the lungs and/or sinuses

Fatigue Extreme tiredness and exhaustion that doesn't go away with rest or sleep and may affect people emotionally and physically

GPA Granulomatosis with polyangiitis, previously called Wegener's, is a type of AAV that usually begins in the nose, ears, eyes or mouth

Imaging In medicine, imaging is a technique which allows healthcare professionals to look inside the patient's body, helping them diagnose and treat disease by revealing internal structures that would otherwise be hidden by skin and bones

Immune system The organs and processes within the body that enable it to respond to and fight infection and toxins

Induction therapy The first treatment in AAV used to get remission

Monoclonal antibody Monoclonal means all of the same type, so each monoclonal antibody (for MAB) reacts to a single antigen. MABs are made in laboratory

MPA Microscopic polyangiitis, a type of AAV that typically affects the kidneys

Nephrologists Doctors specialising in the kidneys

Neurologists Doctors specialising in the nervous system, including the nerves, spinal cord and brain

Otolaryngologists Doctors specialising in the ears, nose and throat, also known as ENT doctors

Progressive A progressive disease is one that becomes more severe over a period of time

Pulmonologists Doctors specialising in the respiratory system

Relapse Refers to disease that was previously well-controlled with or without treatment, but has become active again

Remission A reduction in the severity of disease. Remission can be full (meaning that no activity is currently occurring as a result of active disease) or partial (meaning that disease activity has been reduced)

Rheumatologists Doctors specialising in the muscles, bones, joints, ligaments and tendons

X-ray A type of imaging procedure where high-energy rays are used to create a picture of an area inside the body

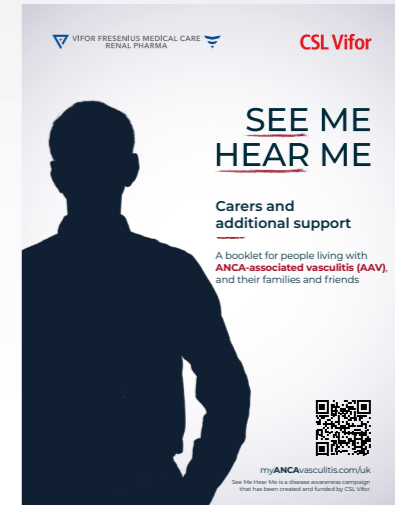
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Other brochures in this series



To find out more about AAV and its signs and symptoms, read our 'What is AAV?' brochure



To find out more about the experience of being a carer for AAV and the additional support available for carers, read our 'Carers and additional support' brochure

For more information on AAV, visit myANCAvasculitis.com/uk



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