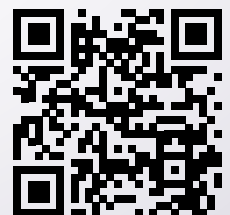


# SEE ME HEAR ME

## What is AAV?

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



[myANCAvasculitis.com/uk](http://myANCAvasculitis.com/uk)

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

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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.

# AAV: AN AUTOIMMUNE DISEASE

Hellen,  
GPA patient,  
Netherlands



## What are autoimmune diseases and how does AAV develop?

Normally, your immune system will make antibodies to fight foreign bodies like bacteria or germs. However, in people with autoimmune diseases, antibodies develop to a specific part of their own cells (autoantibodies), leading to their immune system attacking their own cells.<sup>1</sup>

In AAV, the autoantibody that develops is called ANCA (which stands for anti-neutrophil cytoplasmic antibody). ANCAs bind to a type of white blood cell called neutrophils, activating them.<sup>2,3</sup>

Activated neutrophils contribute to inflammation, which can cause damage to the walls of the blood vessels, as seen in AAV.<sup>2,3</sup>

### Understanding the terminology...

- **ANCA:** stands for anti-neutrophil cytoplasmic antibody, a specific autoantibody that develops in AAV
- **Antibody:** a protein produced by the immune system to target and attack foreign cells like bacteria
- **Autoantibody:** a type of antibody that develops against one of the body's own cells, instead of a foreign cell
- **Complement system:** proteins that form part of the immune system and increase the body's ability to fight infection, but can lead to unwanted inflammation in people with AAV
- **Neutrophil:** a type of white blood cell that forms part of the immune system, and is involved in AAV development

More terms can be found on page 12 within the glossary

## What types of AAV are there?

There are three types of AAV; granulomatosis with polyangiitis (GPA), microscopic polyangiitis (MPA) and eosinophilic granulomatosis with polyangiitis (EGPA)<sup>4</sup>

What do the types of AAV look like?	
<b>GPA</b> Granulomatosis with polyangiitis (Wegener's Granulomatosis)	<ul style="list-style-type: none"> <li>• General symptoms include fever, weight loss and fatigue</li> <li>• Areas of the body that can be impacted include; lungs, ears, nose and throat, the eyes, the kidneys, the peripheral nervous system and the skin</li> <li>• There is granulomatous inflammation (clumps of immune cells)</li> </ul>
<b>MPA</b> Microscopic polyangiitis	<ul style="list-style-type: none"> <li>• General symptoms include fever, weight loss and fatigue</li> <li>• Areas of the body that can be impacted are the same as in GPA, but the ears, nose and throat are impacted less</li> <li>• There is no granulomatous inflammation</li> <li>• The kidneys and skin are commonly impacted</li> </ul>
<b>EGPA</b> Eosinophilic granulomatosis with polyangiitis (Churg-Strauss Syndrome)	<ul style="list-style-type: none"> <li>• General symptoms include fever, weight loss, and fatigue</li> <li>• Areas of the body that can be impacted include; ears, nose and throat, lungs, skin, peripheral nervous system, kidneys, heart and the gastrointestinal tract</li> <li>• Asthma and eosinophilia are present in nearly all EGPA cases and usually come before vasculitis</li> </ul>

For a more detailed explanation of this process, visit [myANCAvasculitis.com/uk/what-is-aav](http://myANCAvasculitis.com/uk/what-is-aav)

For more information on the different types of AAV, visit [myANCAvasculitis.com/uk/what-is-aav](http://myANCAvasculitis.com/uk/what-is-aav)

# SIGNS AND SYMPTOMS



Hermann,  
GPA patient, Germany

## What are the signs and symptoms of AAV?

### The patient perspective

#### Martina's story

"I was suffering from flu-like symptoms, headaches and I was always tired. I went to see my physician and was told I probably had a viral infection. But I couldn't get over it."

"The symptoms would return, sometimes less severe, sometimes worse. For years, the physician told me the same thing. One day, it was so bad I went straight to the Accident & Emergency room. A rheumatologist in the hospital diagnosed MPA really quickly, but tests showed my kidney function had already deteriorated. **The nephrologist who intervened took good care of me and prevented me ending up on dialysis**".

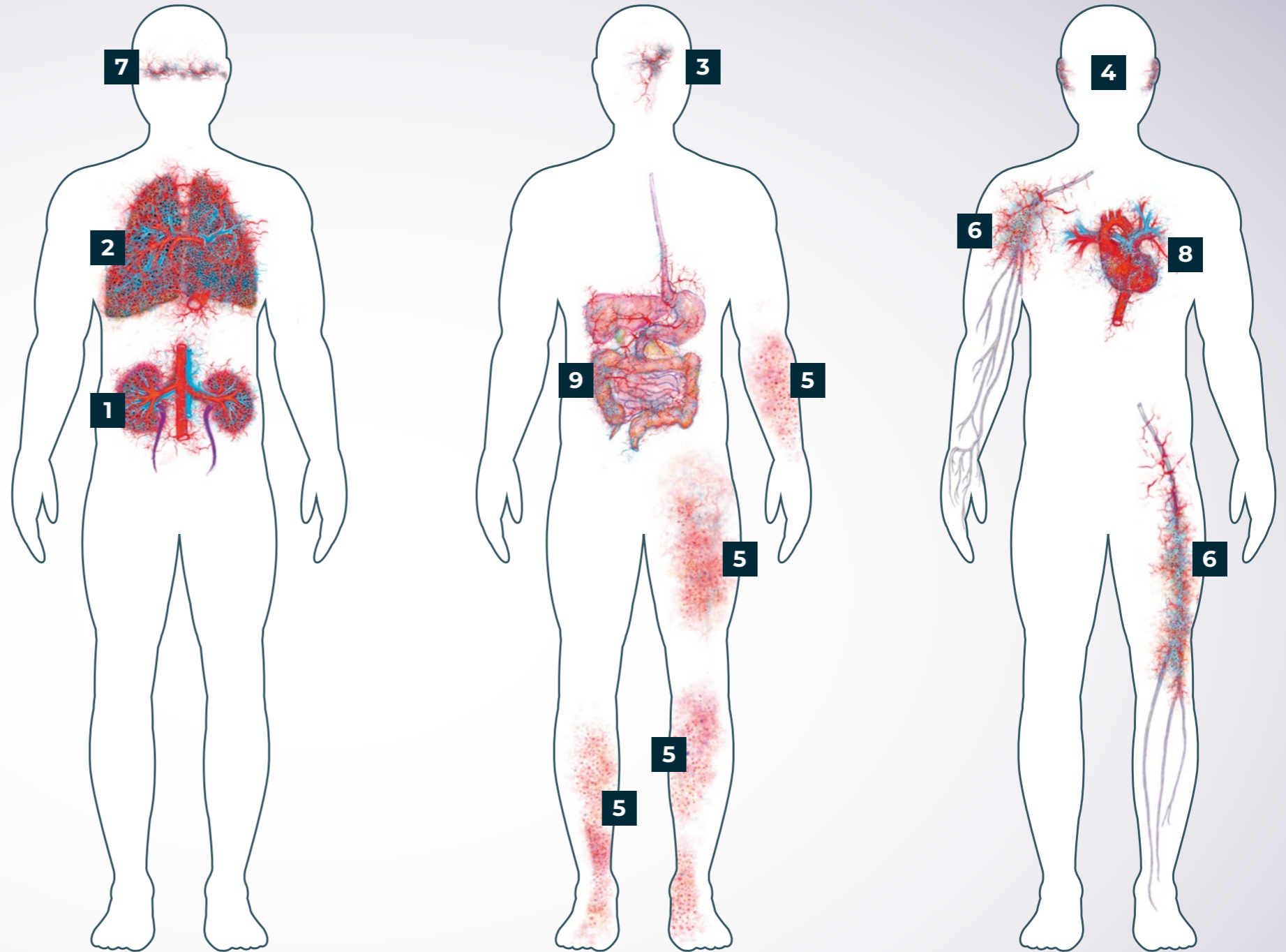


Martina, single mother with two young children, Germany

# What are the signs and symptoms of AAV?

## Discuss your symptoms with your doctor

Whatever you're going through, there is no need to feel embarrassed by talking about it with your doctors. They will be used to discussing a wide range of health issues and telling them about everything you are experiencing, even the smallest of symptoms, can be very useful in helping them to understand your disease.



- |  |   |  |  |   |  |   |   |  |
|--|---|--|--|---|--|---|---|--|
| <p><b>1</b> Kidneys</p> <ul style="list-style-type: none"> <li>• Blood in urine</li> </ul> | <p><b>2</b> Lungs</p> <ul style="list-style-type: none"> <li>• Difficulty breathing/shortness of breath</li> <li>• Cough</li> </ul> | <p><b>3</b> Nose</p> <ul style="list-style-type: none"> <li>• Sinus pain/pressure/congestion/infections</li> <li>• Nosebleeds (nose crusting)</li> </ul> | <p><b>4</b> Ears</p> <ul style="list-style-type: none"> <li>• Hearing loss</li> <li>• Earache</li> </ul> | <p><b>5</b> Skin</p> <ul style="list-style-type: none"> <li>• Rash and sores</li> </ul> | <p><b>6</b> Nerves</p> <ul style="list-style-type: none"> <li>• Numbness, tingling, or weakness (peripheral neuropathy)</li> </ul> | <p><b>7</b> Eyes</p> <ul style="list-style-type: none"> <li>• Red, painful eyes</li> <li>• Blurred vision</li> <li>• Watery eyes</li> </ul> | <p><b>8</b> Heart</p> <ul style="list-style-type: none"> <li>• Heart failure</li> </ul> | <p><b>9</b> Digestive system</p> <ul style="list-style-type: none"> <li>• Diarrhoea</li> <li>• Abdominal pain</li> </ul> |
|--|---|--|--|---|--|---|---|--|

## Glossary

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**AAV** ANCA-associated vasculitis

**ANCA** Anti-neutrophil cytoplasmic antibody

**Antibody** A protein produced by plasma cells that the immune system generate to respond to a specific antigen

**Antigen** A substance that the immune system perceives as 'foreign' or dangerous. The body responds to antigens by producing antibodies

**Autoantibody** A type of antibody produced by the immune system that binds to one of the body's own proteins instead of a foreign object, leading to autoimmune diseases

**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

**Churg-Strauss syndrome** The original name for eosinophilic granulomatosis with polyangiitis, please see 'EGPA' for the full description

**Complement system** Proteins that form part of the immune system, increasing the body's ability to fight infection, but can lead to unwanted inflammation in people with AAV

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

**Eosinophils** A type of white blood cell that help the body's allergic responses

**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

**Granulomatosis** The small, nodule-like groups of cells which commonly form in GPA patients' respiratory tracts

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

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

**Neutrophil** A type of white blood cell that protects the body from infections, among other functions

**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)

**Wegener's Granulomatosis** The original name for granulomatosis with polyangiitis, please see 'GPA' for full description

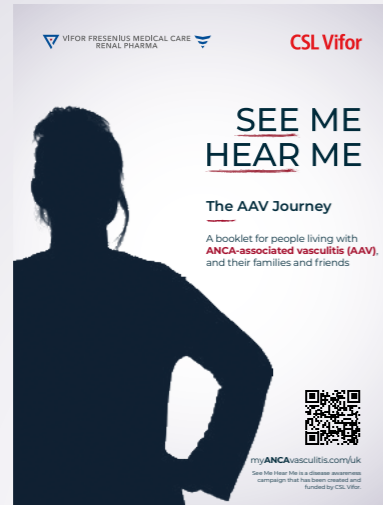
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## Other brochures in this series

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To learn about the AAV journey, from diagnosis and clinical management to treatment and self-management options, read our 'The AAV Journey' 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

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