

UNDERSTAND AAV

Understand ANCA-associated vasculitis

Dialogue Tool

For Healthcare Professionals
& Patients

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What is ANCA-associated vasculitis (AAV)?

AAV is a group of rare autoimmune diseases^{1,2}

AAV is a disease of the small blood vessels^{1,2}

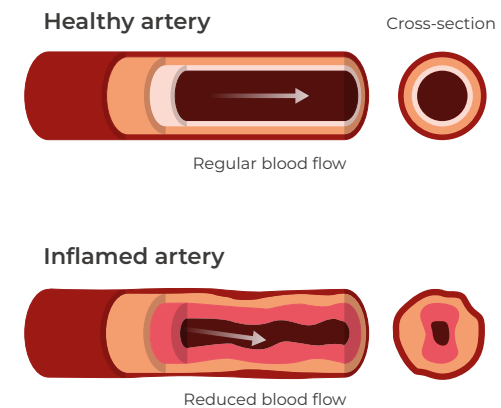
AAV is a long-term condition that may come and go, despite treatment^{3,4}



Number of affected patients^{5,6}

New onset patients / incidence in Europe:
13–20 cases / 1 million people / year

Affected individuals / prevalence worldwide:
46–184 cases / 1 million people



AAV affects the small blood vessels in the entire body and results in damage to blood vessels and organs^{1,7}



AAV is caused by the immune system inappropriately activating neutrophils,* causing inflammation and damage to blood vessels^{1,8}

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

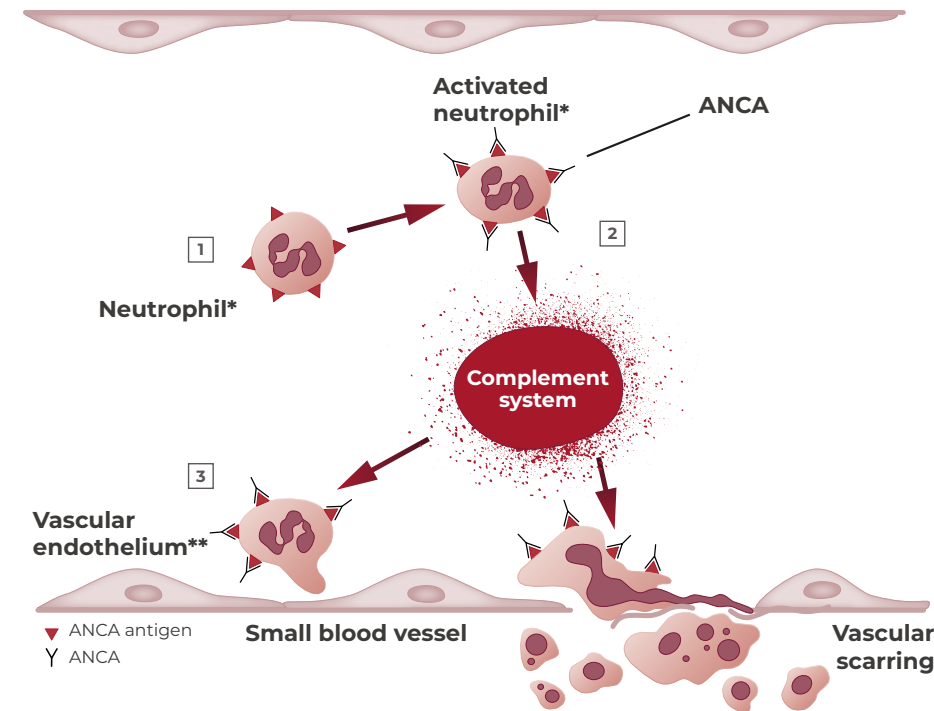
What is happening in my body with AAV?

What are autoimmune diseases?

Conditions that occur when the immune system goes into overdrive and builds antibodies that attack healthy cells.⁹

A microscopic view:

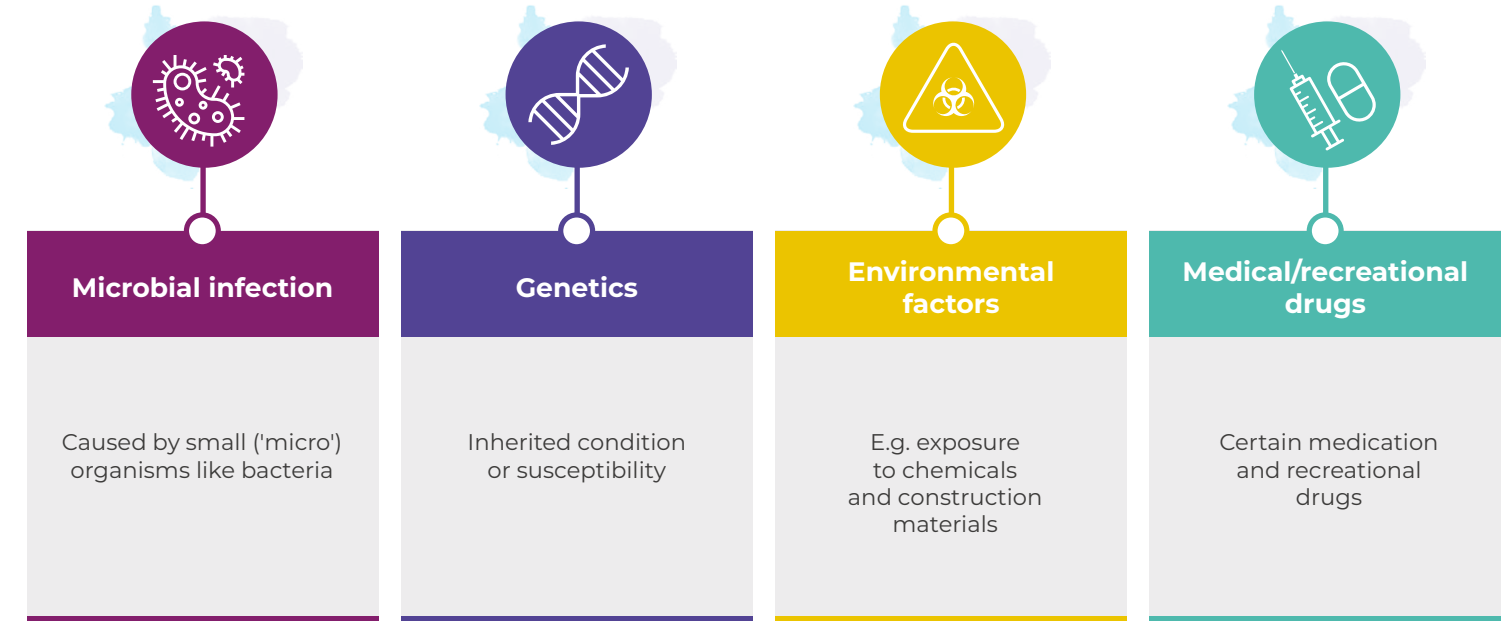
What does AAV look like inside a small blood vessel?



- 1 AAV onset**
In AAV, ANCA autoantibodies bind to proteins on neutrophils^{1,8}
- 2 Inflammation**
These 'activated' neutrophils cause inflammation and activate the complement system^{1,8}
- 3 Vessel damage**
Neutrophils cause damage, that over time can lead to scarring^{1,8}

What are the potential causes of AAV?

The causes of AAV have not been determined yet, but various factors may increase the likelihood of developing AAV^{1,2}



⁴ *A type of white blood cell that, among other functions, protects the body from infections
⁵ **The inner layer of cells lining blood vessels, playing an essential role in regulating vascular tone and cell activity

What are the different types of AAV?

There are three main types of AAV; sometimes you may experience characteristics of more than one type^{8,10,11}

Granulomatosis with Polyangiitis (GPA)

previously known as Wegener's granulomatosis



Common symptoms of GPA: chronically blocked nose, blood encrusted nostrils, red eyes, ear infections, renal, pulmonary and nerve problems^{7,10}

Microscopic Polyangiitis (MPA)



Common symptoms of MPA: bloody cough, purple rash, joint pain, renal and nerve problems⁷

Eosinophilic Granulomatosis with Polyangiitis (EGPA)

previously known as Churg-Strauss syndrome



Common symptoms of EGPA: pulmonary and sinus problems, also frequently associated with asthmatic complaints⁷

Signs and symptoms of AAV^{7*}

Kidneys

Blood in the urine



Lungs

Difficulty breathing, cough



Nose

Blood-encrusted nostrils



Eyes

Red, irritated eyes, blurred vision



Ears

Hearing loss, ear infections – symptoms: pain, headache, fluid excretion



Nerves

Neurological malfunction – symptoms: numbness, tingling sensation



Skin

Rashes, sore spots and blisters



Heart

High blood pressure[†]



Digestive system

Diarrhoea, abdominal pain



*You may experience some but not all of these symptoms

[†]Symptoms: severe headaches, tiredness or confusion, visual problems, chest pain, difficulty breathing, tachycardia

Diagnosing AAV

Diagnosing AAV involves a number of tests and your medical history is also considered⁷

Imaging techniques

Identification of areas affected by AAV such as the lungs, kidneys and other internal organs

Blood tests

Testing scope:

- ANCA autoantibodies*
- Inflammatory markers
- Kidney function

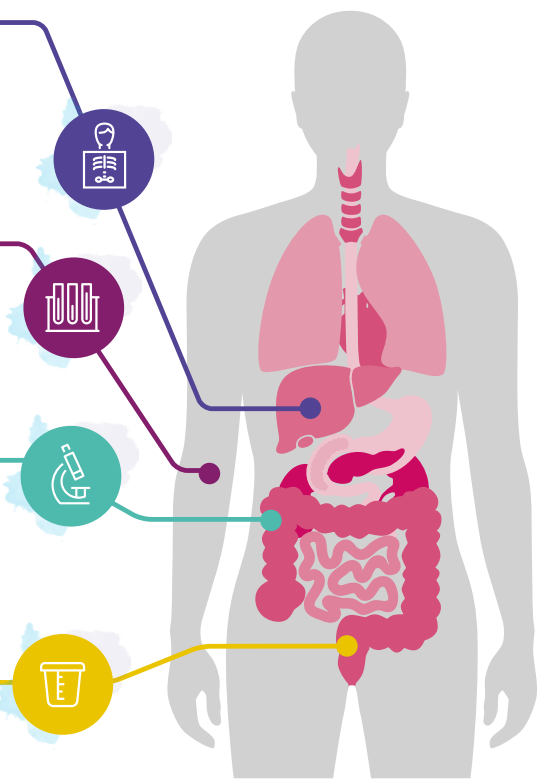
Biopsy

Examination of tissue samples affected by the condition

Urine tests

Testing scope:

- Kidney inflammation
- Kidney function



ANCA testing

There are two different ANCA autoantibodies that develop in people with AAV. These develop to two different proteins: proteinase 3 (PR3) or myeloperoxidase (MPO).⁸

PR3 and MPO are normally found inside neutrophils but if they appear outside, the ANCA can bind to them and activate the neutrophils, leading to damage.⁸ A blood test is used to see if one of these autoantibodies is present in your blood. This is the key test that your doctor will do to see if you have AAV.^{12,13}

Treating AAV (1/2)

Your individual AAV treatment depends on the type of AAV you are affected by and the severity of the inflammation. In all therapy options currently available, the body's own immune system activity is ramped down^{3,4}

Drug	Steroids*	Rituximab	Cyclophosphamide
What is it?	Glucocorticoids (a type of steroid), for the treatment of autoimmune diseases ¹⁴	For the treatment of autoimmune diseases ¹⁵	For the treatment of autoimmune diseases ¹⁶
When is it administered? ⁴	Induction therapy, remission and relapse [†]	Induction therapy, remission and relapse [†]	Induction therapy and relapse [†]
How it is administered? ¹⁷	High-dosage infusions or tablets (reducing the dosage over time)	Relapse: one infusion weekly for one month Remission: one infusion every 4 to 6 months	One infusion every two weeks initially and then at three-week intervals
Where do I receive my infusion?	Day clinic, infusion centre, or doctor's surgery (about 30 minutes for one infusion)	Day clinic, infusion centre, or doctor's surgery (about one half-day for one infusion)	Day clinic, or infusion centre (about one half-day for one infusion)

⁸ In some individuals with GPA, MPA or EGPA, it may not be possible to detect any ANCA autoantibodies

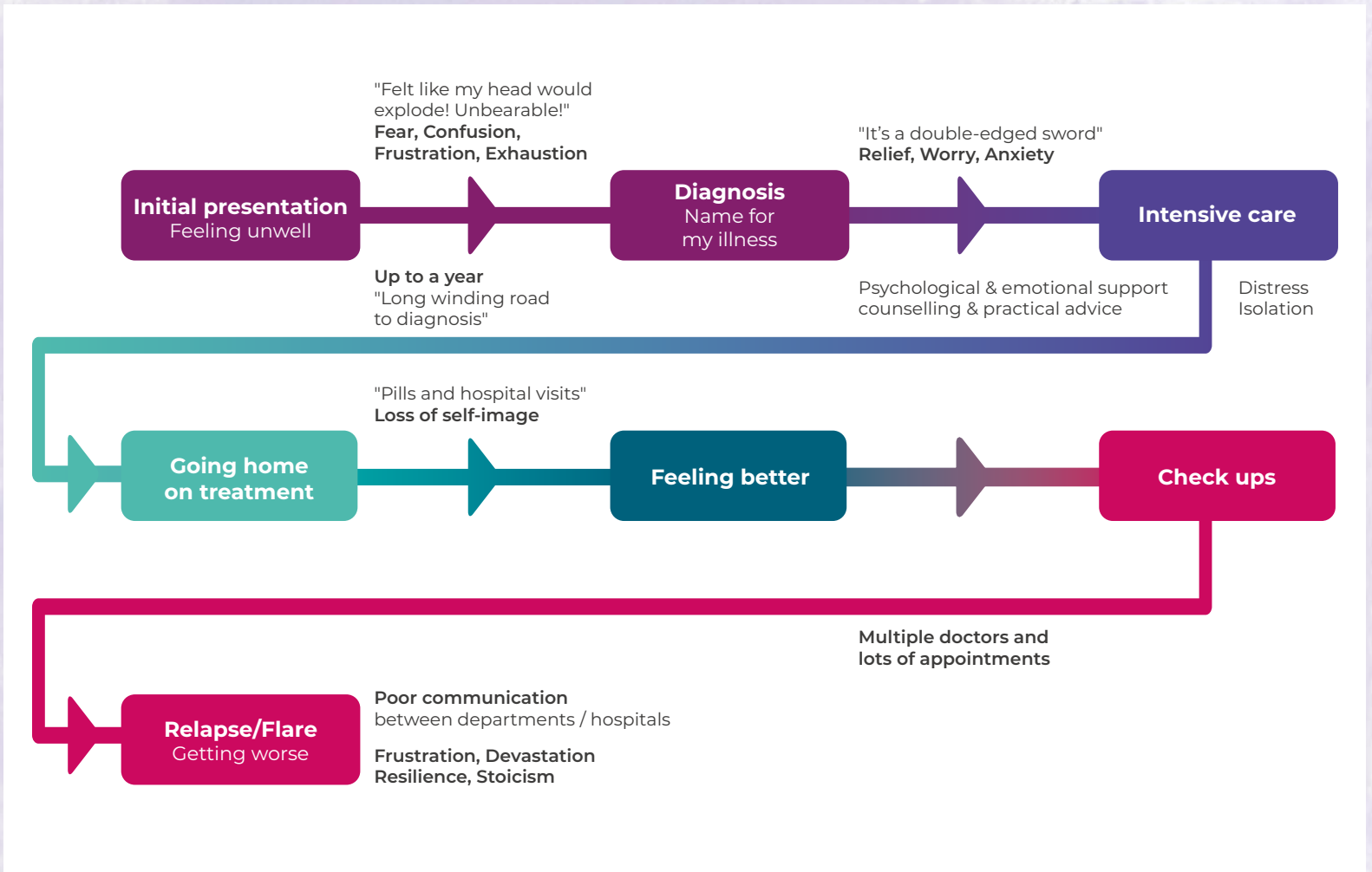
*Steroids can be taken in combination with rituximab or cyclophosphamide, but rituximab and cyclophosphamide are not taken together⁴
[†]Induction therapy: the first treatment AAV patients receive to obtain remission; Remission: a reduction in the severity of disease, remission can be full (i.e. there is no current disease activity) or partial (i.e. disease activity is contained); Relapse: refers to a disease that was previously under control with or without treatment, but has become active again⁴

Treating AAV (2/2)

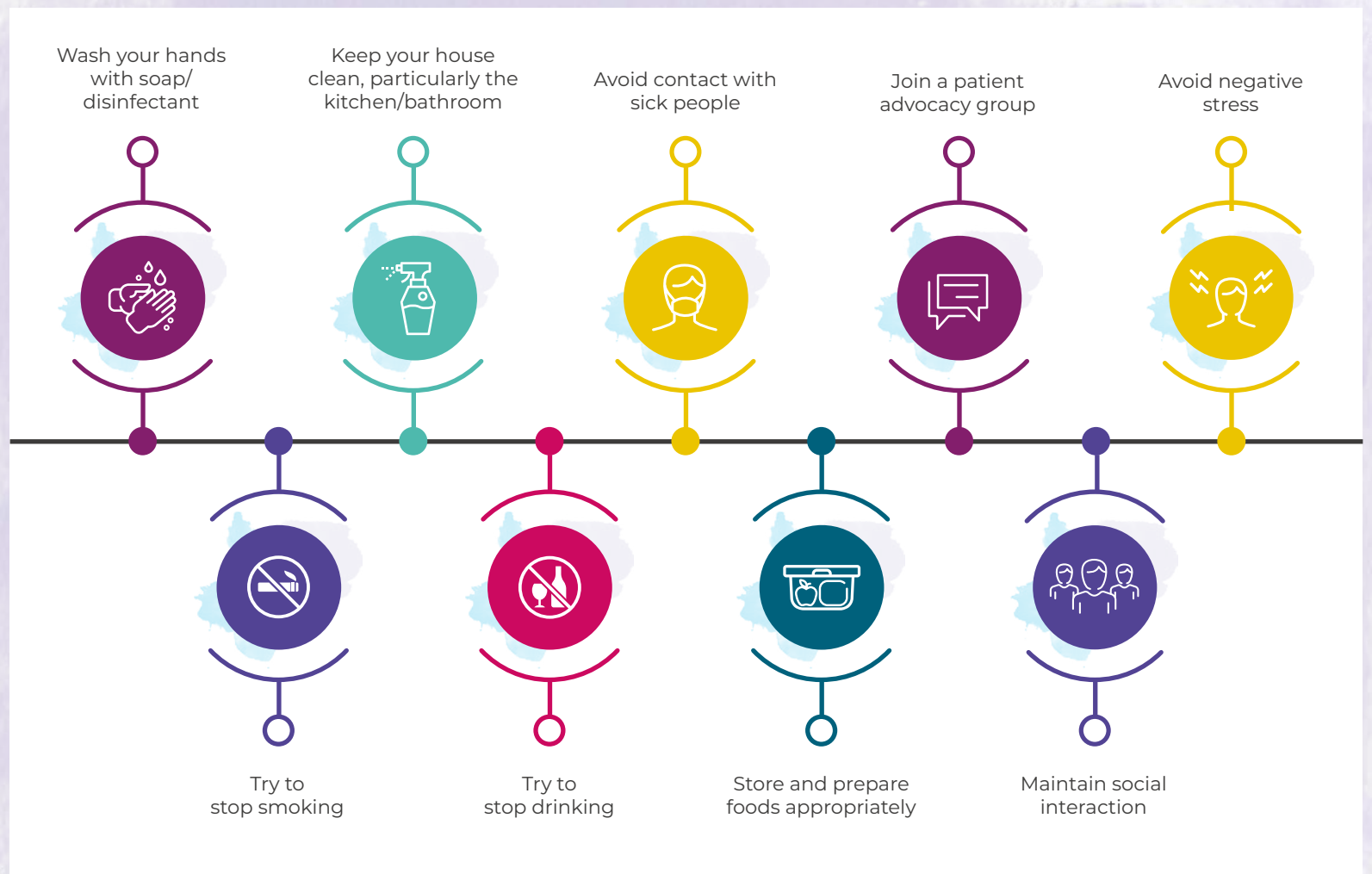
Your individual AAV treatment depends on the type of AAV you are affected by and the severity of the inflammation; in all treatment plans, the body's own immune system activity is ramped down^{3,4}

Drug	Azathioprine (off-label use)	Methotrexate (off-label use)	Mycophenolate mofetil (off-label use)
What is it?	For the treatment of rheumatoid arthritis and various autoimmune diseases ¹⁸	Anti-inflammatory agent for the treatment of bone, joint and autoimmune diseases ¹⁹	For the treatment of autoimmune and renal conditions and kidney transplants ²⁰
When it is administered? ⁴	Remission*	Remission*	Remission*
How it is administered?	Tablet, once daily ⁴	Tablet or infusion, once a week ¹⁷	Tablet, daily ^{17,20}
Where do I receive my infusion?	–	At home (infusion takes about one minute)	–

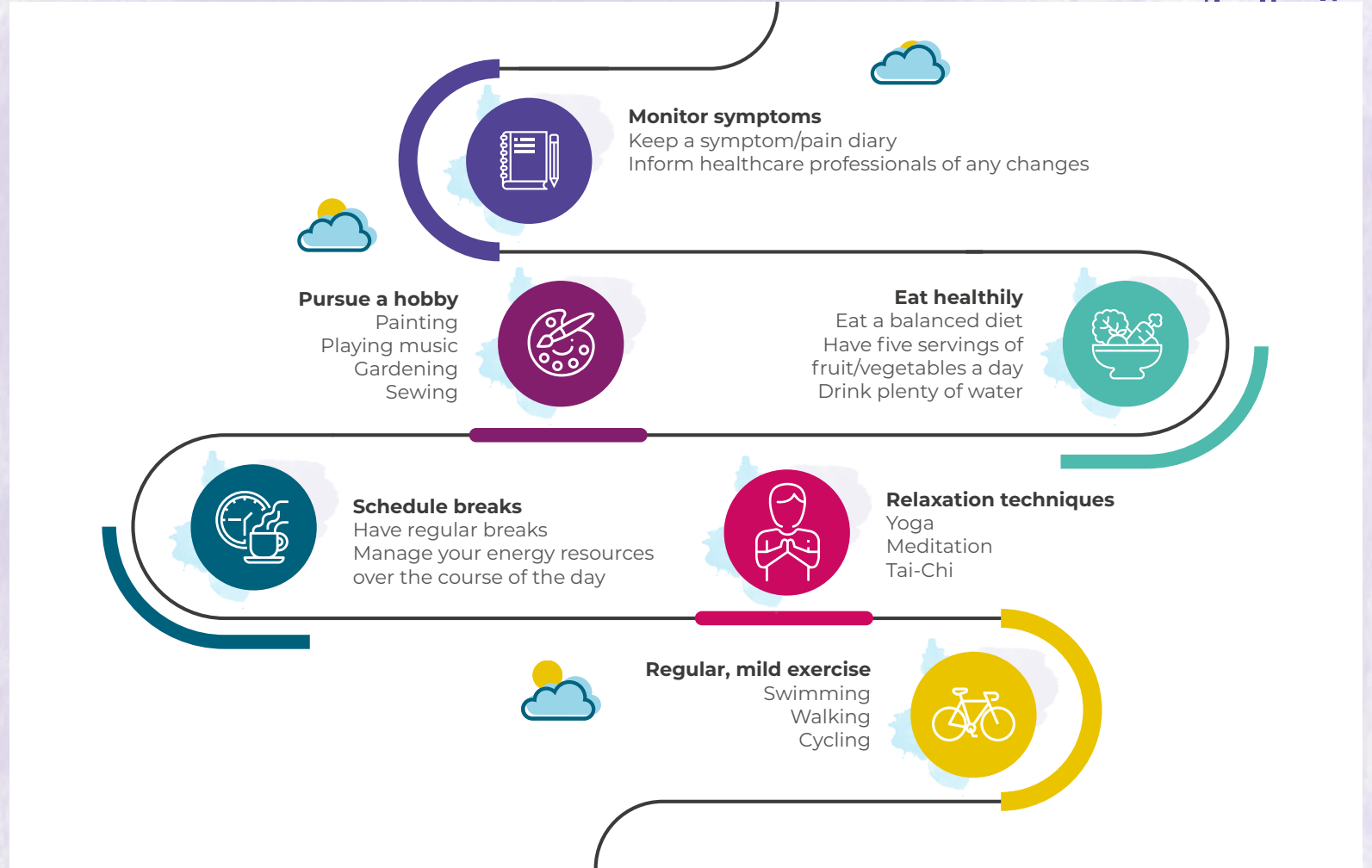
A typical patient journey²¹



Advice for when you are undergoing AAV treatment



Recommendations for living with AAV



Glossary (1/2)

A

ANCA

Abbreviation: anti-neutrophil cytoplasmic antibody.

ANCA-Test

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

Antibody

A protein produced by plasma cells that the immune system generates 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

A condition caused by the body's own immune system. This autoimmune reaction (or 'autoimmune attack') triggers the development of the disease.

Azathioprine

An immunosuppressant that is used to treat a number of inflammatory conditions, including AAV. Off-label use.

B

Biopsy

In a biopsy, a small tissue sample is taken to be examined under a microscope.

C

C5a

A protein involved in the complement system, which plays a key role in AAV associated inflammation.

Churg-Strauss syndrome

Previous name for eosinophilic granulomatosis with polyangiitis (EGPA). For a more detailed description, please see the entry under 'EGPA'.

Complement system

Proteins including C5a 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.

Cyclophosphamide

A medicine used to control autoimmune diseases like AAV by suppressing the immune system's response.

E

EGPA

Eosinophilic Granulomatosis with Polyangiitis, previously called Churg-Strauss syndrome, a type of AAV that often affects the lungs and/or sinuses.

G

GPA

Granulomatosis with Polyangiitis, previously called Wegener's granulomatosis, a type of AAV that usually begins in the nose, ears, eyes or mouth.

Genetics

The physical characteristics a person inherits from their parents.

Glucocorticoids

Steroid hormones that are used to treat diseases caused by an overactive immune system.

Granulomatosis

The small, nodule-like cell clusters that commonly form in GPA patients' sinuses and lungs.

I

Imaging techniques

In medicine, imaging techniques allow healthcare professionals to look into their patient's body, helping them to diagnose and treat diseases 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 infections and toxins.

Glossary (2/2)

Induction therapy

The first treatment in AAV in order to obtain remission.

M

MPA

Microscopic Polyangiitis, a type of AAV that typically affects the kidneys.

Methotrexate

An immunosuppressant that is used to treat AAV as well as a number of other conditions. Off-label use.

Microbial

Relating to small (micro) organisms, especially disease-causing bacteria.

Monoclonal antibody

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

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Mycophenolate mofetil

Another immunosuppressant used to treat AAV as well as various other conditions. Off-label use.

N

Neutrophil

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

P

Pulmonary

Relating to the lungs.

R

Rare disease

A disease affecting fewer than 1 in 2000 people.

Relapse

Refers to a condition that has flared up again after previously being under control with or without treatment.

Remission

A reduction in the severity of disease. Remission can be full (i.e. there is no current disease activity) or partial (i.e. disease activity is contained).

Rituximab

A certain type of monoclonal antibody used to treat AAV and other diseases.

V

Vascular system

The vascular system, also referred to as the circulatory system, is comprised of the vessels transporting blood and lymph (a fluid containing white blood cells).

W

Wegener's granulomatosis

Previous names of granulomatosis with polyangiitis (GPA). For a more detailed description, please refer to the entry under 'GPA'.

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Further information

The following links provide information and resources for people living with or affected by AAV:



**SEE ME
HEAR ME**
#myANCAvasculitis

www.myANCAvasculitis.com

This website is dedicated towards supporting people living with or affected by AAV.



UNDERSTAND AAV

www.understandAAV.com

More information for health care professionals about AAV.



www.vasculitis.eu

Their mission is to encourage and support Vasculitis Patient Advocacy Groups (VPAGs) to combine forces and to jointly develop relevant projects and activities.



www.viforpharma.com

