

## IgA VASCULITIS (formerly Henoch-Schönlein Purpura)

### What is IgA vasculitis?

IgA vasculitis, formerly Henoch-Schönlein purpura, is a form of vasculitis—a family of rare disorders characterized by inflammation of the blood vessels, which can restrict blood flow and damage vital organs and tissues. IgA (immunoglobulin A) vasculitis causes inflammation and bleeding of the small blood vessels of the skin, joints, intestines and kidneys. Rarely, it can affect the lungs and central nervous system. It is the most common form of vasculitis in children.

IgA vasculitis is systemic, meaning it can affect all organ systems in the body. The most characteristic symptom is a raised, purplish skin rash that resembles bruises, mostly affecting the legs and buttocks. Other common symptoms or signs of organ involvement include abdominal pain, joint pain and swelling, and kidney inflammation.

When IgA vasculitis occurs in children younger than 16 years old, it is considered self-limiting because most patients recover on their own within one to two months, without treatment. Adults may have more severe and relapsing disease requiring immunosuppressive treatment. If the kidneys and intestines are affected, medical treatment is often needed, and ongoing medical follow-up is necessary.

### Causes

The exact cause of IgA vasculitis is not fully understood by researchers. Vasculitis is classified as an autoimmune disorder, a disease which occurs when the body's natural defense system mistakenly attacks healthy tissue. In IgA vasculitis, the immunoglobulin A antibody—a type of antibody that works to fight infections—is thought to play a role in blood vessel inflammation.

Studies show that 75 percent of people with IgA vasculitis develop the disease after an upper respiratory or gastrointestinal (GI) infection. Other possible triggers for IgA vasculitis include certain medicines, food reactions, insect bites, some vaccinations and, rarely, cancer. Although IgA vasculitis is seen throughout the year, there is an increase in cases during the fall and winter months. It is not considered contagious.

### Who gets IgA vasculitis?

IgA vasculitis can affect people of any age, but 90 percent of patients are children aged 3 to 10 years. It is more common in boys than in girls. Adults tend to have more severe disease than children, and therefore may be treated more aggressively.

The prevalence of IgA vasculitis in the United States is approximately 14 cases per 100,000 population. In the United Kingdom, the estimated annual incidence is 20.4 cases per 100,000 population.

### Symptoms

Patients with IgA vasculitis may experience several weeks of headache, fever and muscle aches before the primary symptoms set in.

- **Rash:** A raised, reddish-purple rash called purpura is the characteristic symptom that helps doctors diagnose the disease. Lesions develop primarily on the buttocks, legs and feet, but can also affect the elbows, arms, and trunk.

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- **Arthritis:** Painful swollen joints, especially affecting the knees and ankles. The arthritis generally goes away on its own without causing permanent joint damage.
- **GI problems:** Stomach pain, nausea, vomiting, or bloody stools, which can occur before the rash.
- **Kidney inflammation:** Blood or protein in the urine, detected in urinalysis test. Kidney inflammation usually dissipates as IgA vasculitis resolves, with most patients recovering completely. However, in some cases, IgA vasculitis can cause kidney damage/failure, and dialysis or a kidney transplant may be needed.

### Complications

Rarely, IgA vasculitis can cause bowel bleeding or bowel obstruction—a potentially severe complication caused by the bowel folding onto itself.

### Diagnosis

There is no single test for diagnosing IgA vasculitis. Your doctor will consider a number of factors, including a detailed medical history, physical examination, laboratory tests, and specialized imaging studies. A skin biopsy is frequently needed to help diagnose IgA vasculitis.

**Laboratory tests:** Blood tests and urinalyses can help rule out other diseases and detect signs of infection, anemia or kidney disease.

**Imaging studies:** These may include ultrasound (abdominal, scrotal/testicular); X-rays (chest, abdomen, small intestine); magnetic resonance imaging (MRI) to assess for neurological complications; and/or computed tomography (CT) scans of the head or abdomen.

**Biopsy:** This surgical procedure removes a small tissue sample from an affected organ, which is examined under a microscope for signs of inflammation or tissue damage. In IgA, a skin biopsy typically shows vasculitis with deposits of immunoglobulin A. A kidney biopsy is sometimes necessary.

### Treatment

Symptoms of IgA vasculitis typically last one to two months, and most patients recover on their own without treatment. Therefore, treatment is usually aimed at relieving minor symptoms until they dissipate. The following medications may be prescribed:

- Antibiotics (if an infection is suspected to be causing the IgA vasculitis)
- Over-the-counter painkillers, such as acetaminophen
- Anti-inflammatories such as ibuprofen, for joint pain and swelling
- Corticosteroids such as prednisone for severe stomach pain or kidney disease. (Steroids are prescribed cautiously due to potentially serious side effects, and should be avoided in children.)

In cases where the kidneys are affected or damaged, patients may require medications that suppress the immune system.

### Side effects

Some IgA patients may need to take corticosteroids or immunosuppressant drugs. These medications can have potentially serious side effects, such as lowering the body's ability to fight infection, and

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potential bone loss (osteoporosis), among others. Therefore, it's important to see your doctor for regular checkups. Medications may be prescribed to offset side effects. Infection prevention is also very important. Talk to your doctor about getting a flu shot, pneumonia vaccination, and/or shingles vaccination, which can reduce your risk of infection.

### **Relapse**

About one-third of individuals who have IgA vasculitis will experience a relapse, typically several months after the initial episode. Symptoms are usually less severe during a relapse. If your initial symptoms return or you develop new ones, report them to your doctor as soon as possible. Regular doctor visits and ongoing monitoring of lab and imaging tests are important in detecting relapses early.

### **Your medical team**

Effective treatment of IgA vasculitis may require the coordinated efforts and ongoing care of a team of medical providers and specialists. In addition to a primary care provider, IgA patients may need to see the following specialists: dermatologist (skin); gastroenterologist (digestive system); rheumatologist (joints, muscles, immune system); nephrologist (kidneys); or others as needed.

The best way to manage your disease is to actively partner with your health care providers. Get to know the members of your health care team. It may be helpful to keep a health care journal to track medications, symptoms, test results and notes from doctor appointments in one place. To get the most out of your doctor visits, make a list of questions beforehand and bring along a supportive friend or family member to provide a second set of ears and take notes.

Remember, it's up to you to be your own advocate. If you have concerns with your treatment plan, speak up. Your doctor may be able to adjust your dosage or offer different treatment options. It is always your right to seek a second opinion.

### **Living with IgA**

Although IgA vasculitis is not chronic for most patients, it can be, especially for adults. Living with a chronic condition can be overwhelming at times. Fatigue, pain, emotional stress, and medication side effects can take a toll on your sense of well-being, affecting relationships, work and other aspects of your daily life. Sharing your experience with family and friends, connecting with others through a support group, or talking with a mental health professional can help.

### **Outlook**

There is no cure for IgA vasculitis at this time. In general, children with IgA vasculitis have an excellent prognosis. In adults, the disorder can be chronic. Most patients with this form of vasculitis fully recover within eight weeks and few have ongoing symptoms. Because relapses can occur, however, follow-up medical care is essential. Patients whose kidneys are affected need treatment and ongoing monitoring.

Clinical studies are ongoing at multicenter research centers, including the Vasculitis Clinical Research Consortium (VCRC), to better understand the risk factors and causes of vasculitis, investigate more effective and safer treatments, and work toward a cure. The Vasculitis Foundation encourages patients

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to consider participating in clinical research studies to help further understanding of vasculitis. Patients are also encouraged to join the Vasculitis Patient Powered Research Network (VPPRN), where they can provide valuable disease insight and information. For more information on vasculitis research, visit: [www.vasculitisfoundation.org/research](http://www.vasculitisfoundation.org/research)

### **About Vasculitis**

Vasculitis is a family of nearly 20 rare diseases characterized by inflammation of the blood vessels, which can restrict blood flow and damage vital organs and tissues. Vasculitis is classified as an autoimmune disorder, which occurs when the body's natural defense system mistakenly attacks healthy tissues. Triggers may include infection, medication, genetic or environmental factors, allergic reactions, or another disease. However, the exact cause is often unknown.

### **A Family of Diseases**

- Anti-GBM (Goodpasture's) disease
- Aortitis
- Behcet's syndrome
- Central nervous system vasculitis
- Cogan's syndrome
- Cryoglobulinemia
- Cutaneous small-vessel vasculitis (formerly hypersensitivity/leukocytoclastic)
- Eosinophilic granulomatosis with polyangiitis (EGPA, formerly Churg-Strauss syndrome)
- Giant cell arteritis
- Granulomatosis with polyangiitis (GPA, formerly Wegener's)
- IgA vasculitis (Henoch-Schönlein Purpura)
- Kawasaki disease
- Microscopic polyangiitis
- Polyarteritis nodosa
- Polymyalgia rheumatica
- Rheumatoid vasculitis
- Takayasu's arteritis
- Urticarial vasculitis

### **About the Vasculitis Foundation**

The Vasculitis Foundation (VF) is the leading organization in the world dedicated to diagnosing, treating, and curing all forms of vasculitis. The VF provides a wide range of education, awareness and research programs and services. To learn more, and get the most updated disease and treatment information, visit [www.vasculitisfoundation.org](http://www.vasculitisfoundation.org)

### **Connect with the VF on Social Media**

- Instagram: [vasculitisfoundation](https://www.instagram.com/vasculitisfoundation)
- Twitter: [@VasculitisFound](https://twitter.com/VasculitisFound)
- VF Facebook Discussion Group: [www.facebook.com/groups/vasculitisfoundation](https://www.facebook.com/groups/vasculitisfoundation)

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### **Ways to Get Involved**

- Participate in research
- Join social media
- Host or participate in an event
- Attend a regional conference and/or symposium
- Give a donation toward long-term solutions
- Become an advocate for yourself, or for others
- Share your journey

### **Vasculitis Foundation Mission**

Building upon the collective strength of the vasculitis community, the Foundation supports, inspires and empowers individuals with vasculitis, and their families, through a wide range of education, research, clinical, and awareness initiatives.

### **Join the VPPRN!**

The Vasculitis Patient-Powered Research Network (VPPRN) seeks to improve the care and health of patients with vasculitis by exploring research questions that matter most to patients, and by advancing medical knowledge about vasculitis. For more information, visit: [www.VPPRN.org](http://www.VPPRN.org)

The Vasculitis Clinical Research Consortium (VCRC) is an integrated group of academic medical centers, patient support organizations, and clinical research resources dedicated to conducting clinical research in different forms of vasculitis. For more information, visit: [www.rarediseasesnetwork.org/cms/vcrc](http://www.rarediseasesnetwork.org/cms/vcrc)

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