

## **What is Immunoglobulin A Vasculitis?**

Immunoglobulin A (IgA) vasculitis, formerly Henoch-Schönlein purpura, is a rare form of vasculitis, which is an inflammation of the blood vessels. Inflammation of blood vessels may result in narrowing of the vessel, which can block or slow down blood flow to vital organs and tissues.

IgA vasculitis is systemic, meaning it can affect many organ systems in the body. It commonly causes inflammation 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. It is rare in adults.

Vasculitis is classified as an autoimmune disorder, a disease that occurs when the body's natural defense system mistakenly attacks healthy tissue.

## **Causes**

The exact cause of IgA vasculitis is not fully understood. With 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.

Most people develop the disease after an upper respiratory or gastrointestinal (GI) infection.

Other possible causes for IgA vasculitis include:

- Certain medicines
- Food reactions
- Insect bites
- Some vaccinations
- Cancer (rarely)

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. Most children younger than 16 years old 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.

## Symptoms

Patients with IgA vasculitis may have headache, fever, and muscle aches for several weeks before the primary symptoms set in. These symptoms include:

- **Rash:** A raised, reddish-purple rash called purpura is the characteristic symptom. The rash develops primarily on the buttocks, legs, and feet, but can also affect the elbows, arms, and trunk.
- **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.
- **Kidney inflammation:** Blood or protein may be found in the urine, detected through a urinalysis test. Kidney inflammation usually goes away as IgA vasculitis gets better. Most patients recover completely; however, in some cases, IgA vasculitis can cause kidney damage/failure. 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 several factors, including a detailed medical history and physical examination, as well as:

- **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 to assess for neurological complications

- Computed tomography scans of the head or abdomen
- **Biopsy:** This surgical procedure removes a small tissue sample from an affected organ. It is examined under a microscope for signs of inflammation or tissue damage. With IgA, a skin biopsy typically shows vasculitis with deposits of immunoglobulin A. A kidney biopsy is sometimes necessary and it also typically shows deposits of immunoglobulin A.

## Treatment

Symptoms of IgA vasculitis usually last one to two months, and most patients recover on their own without treatment. Therefore, treatment is usually aimed at relieving 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 pain medications, 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 of Medications

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
- Potential bone loss (osteoporosis), among others

Therefore, it is important to see your doctor for regular checkups. Medications may be prescribed to relieve the side effects of the medications.

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:

- Rheumatologist (joints, muscles, immune system)
- Dermatologist (skin)
- Gastroenterologist (digestive system)
- Nephrologist (kidneys); or others as needed

The best way to manage your disease is to actively partner with your health care providers and 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 is 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**

Currently, there is no cure for IgA vasculitis. 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. However, because relapses can occur, follow-up medical care is essential. Patients whose kidneys are affected need treatment and ongoing monitoring.

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PO Box 28660, Kansas City, MO 64188 | 816.436.8211 | [vf@vasculitisfoundation.org](mailto:vf@vasculitisfoundation.org) | [www.VasculitisFoundation.org](http://www.VasculitisFoundation.org)

