Henoch-Schönlein Purpura

National Kidney and Urologic Diseases Information Clearinghouse

National Institute of Diabetes and Digestive and Kidney Diseases

NATIONAL INSTITUTES OF HEALTH

Henoch-Schönlein purpura (HSP) is a disease that causes small blood vessels in the skin to leak because of inflammation. The primary symptom is a rash that looks like many small raised bruises on the legs. The rash is most often on the legs and buttocks, but it can appear on other parts of the body. Some people with HSP also develop abdominal pain or joint pain (arthritis). The kidneys may be affected as well, causing blood or protein in the urine. HSP can occur any time in life, but it usually happens in children between the ages of 2 and 11.

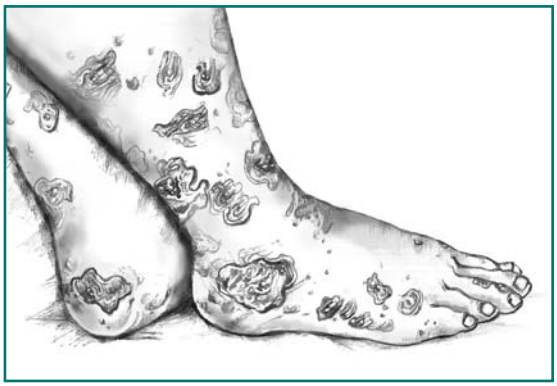
In most cases, HSP lasts 4 to 6 weeks, with no long-term consequences. Sometimes symptoms come and go during this time period. About one in three people have more than one episode (recurrence) of HSP. Recurrences usually occur within a few months and are usually less severe than the initial episode. Even when it lasts longer than a few months, HSP can still resolve completely. In a few cases, however, it can lead to kidney damage and permanent kidney failure. A person with severe kidney failure must receive a blood-cleansing treatment called dialysis or a kidney transplant if the damage is permanent.

Another rare complication of HSP is intussusception of the bowel, or intestine. With this condition, a section of the bowel folds into itself like a telescope. The bowel may become blocked as a result. Surgery may be needed to correct the problem.

What are the causes of HSP?

The causes of HSP are not fully understood. One theory is that it may develop as an immune response to an infection. In other words, the body's infection-fighting system, the immune system, continues to attack cells after the infecting organisms are gone. For example, HSP may develop after a cold. The cold germs cause your immune system to take action. Once the immune cells have rid the body of the germ cells, they normally rest. But with HSP, the immune cells continue to attack other cells in the body. This theory is also based on the fact that, in many cases, HSP symptoms recur or worsen during upper respiratory infections.

HSP has also been associated with insect bites and exposure to cold weather. Other cases have developed after a person received vaccination for typhoid, measles, cholera, hepatitis B, or yellow fever. Some foods, drugs, or other chemical toxins may trigger HSP as well. Often no cause can be found.



The primary symptom of Henoch-Schönlein purpura is a rash that looks like many small raised bruises on the legs.

U.S. Department of Health and Human Services

What are the symptoms of HSP?

HSP has four main symptoms:

- **Rashes and bruising.** Leaking blood vessels in the skin cause rashes that look like bruises or small red dots to develop on the legs, buttocks, and back of the arms. The rash may first look like hives, then change to look like bruises. Rarely, the rash may spread to the upper part of the body, but it is usually on the parts of the body that "hang down," like the legs, buttocks, elbows, and even earlobes. The rash does not disappear or turn pale when you press on it.
- **Abdominal pain.** About two-thirds of people with HSP experience pain in the stomach that may cause vomiting or blood in the stool. This pain and bleeding can vary from mild to severe.
- **Arthritis.** About 80 percent of people with HSP have pain and swelling in their joints, usually in the knees and ankles, less frequently in the elbows and wrists. These joint symptoms have no long-lasting effects, although they can be very uncomfortable while they're present.
- **Kidney involvement.** Blood in the urine (hematuria) occurs in about 40 percent of people with HSP. Often the blood cannot be seen by the naked eye, but it can be measured with a laboratory test called a urinalysis. In most people the hematuria goes away without permanent kidney damage. Protein in the urine or development of high blood pressure (hypertension) suggests more severe kidney problems.

How is HSP diagnosed?

When a typical rash, abdominal pain, and arthritis are present, the doctor can easily recognize HSP. But many people with HSP have only the rash, which can sometimes be seen as a symptom of other conditions and may delay the clinical diagnosis of HSP.

The doctor may need to perform a series of tests to confirm a diagnosis of HSP, as no single test for HSP exists.

- **Blood test.** Elevated levels of blood urea nitrogen and creatinine, which are waste products that are normally in the blood at low levels, indicate that the kidneys are affected. Healthy kidneys filter urea and creatinine from blood.
- **Urine sample.** A urine sample is needed to check for hematuria, which is blood in the urine, and to check for proteinuria, which is protein in the urine. Blood and high levels of protein in the urine indicate damage to the kidneys.
- **Skin biopsy.** If other testing is inconclusive, and a diagnosis is required, the doctor may take a small sample of your skin to examine with a microscope. The biopsy may reveal large numbers of white blood cells in the skin and deposits of IgA, one of the proteins normally made by the immune system to help fight off infections.
- **Kidney biopsy.** When the kidneys are affected by HSP, the nephrologist (kidney specialist) may take a small sample of kidney tissue to examine with a microscope. Examining the sample can help the doctor decide what specific medicines, if any, need to be given for the kidney disease. Very few patients with HSP need a kidney biopsy.

Several diseases share some of the symptoms of HSP. But consistent physical exam findings, along with blood, urine, and skin test results taken together, can help the doctor identify HSP.

How is HSP treated?

There is no specific treatment for HSP. The main goals of treatment are to relieve symptoms such as joint pain, abdominal pain, or swelling. In most cases, you can use over-the-counter medicines, such as acetaminophen (Tylenol), for the pain. In some patients with severe arthritis, the doctor may prescribe prednisone, a steroid medicine. As mentioned earlier, the rash and joint symptoms usually go away after 4 to 6 weeks without causing permanent damage.

Severe problems with the bowels are rare in HSP, especially in younger children. If you have severe pain or severe bleeding in the digestive tract your doctor may prescribe prednisone, or the problem may need to be corrected with surgery.

Your doctor will check your kidney function with blood and urine tests even after the main symptoms of HSP disappear. People who develop kidney disease usually show signs within 3 to 6 months after the initial rash appears. If signs of kidney disease appear, your doctor will refer you to a nephrologist, who may prescribe drugs to suppress the immune system. These immunosuppressive drugs may keep kidney disease from progressing to permanent kidney failure.

What is the long-term outlook?

Most cases of HSP resolve within 4 to 6 weeks without long-term problems. About one in three people has a recurrence of HSP. Recurrences usually occur within a few months and are usually less severe than the initial episode. When the symptoms recur or last longer than 6 weeks, they can be very frustrating and uncomfortable. The long-term outlook is still good, however, as long as your kidneys are healthy.

If you develop progressive kidney disease, you will need to have regular checkups to monitor your kidney function. In the early stages of kidney disease, you may not have any symptoms, but blood and urine tests may show that your kidney function is declining. If you continue to have blood and protein in your urine, you are at greater risk of developing chronic kidney disease.

Between 20 and 50 percent of children with HSP develop some kidney problems, but only 1 percent progress to total kidney failure. Progression to kidney failure may take as long as 10 years.

Hope Through Research

Through its Division of Kidney, Urologic, and Hematologic Diseases, the National Institute of Diabetes and Digestive and Kidney Diseases (NIDDK) supports several programs and studies devoted to improving treatment for patients with progressive kidney disease and kidney failure. The NIDDK maintains the Pediatric Nephrology Program, which supports research into the causes, treatment, and prevention of kidney diseases in children, including congenital malformations of the urinary tract, polycystic disease, primary glomerular disease, and postinfectious glomerulonephritis.

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The National Kidney and Urologic Diseases Information Clearinghouse (NKUDIC) is a service of the National Institute of Diabetes and Digestive and Kidney Diseases (NIDDK). The NIDDK is part of the National Institutes of Health under the U.S. Department of Health and Human Services. Established in 1987, the Clearinghouse provides information about diseases of the kidneys and urologic system to people with kidney and urologic disorders and to their families, health care professionals, and the public. The NKUDIC answers inquiries, develops and distributes publications, and works closely with professional and patient organizations and Government agencies to coordinate resources about kidney and urologic diseases.

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This fact sheet is also available at *www.kidney.niddk.nih.gov.*

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