Henoch-Schönlein Purpura

National Kidney and Urologic Diseases Information Clearinghouse

U.S. Department of Health and Human Services

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What is Henoch-Schönlein purpura (HSP)?

Henoch-Schönlein purpura is a disease that causes small blood vessels in the body to become inflamed and leak. The primary symptom is a rash that looks like many small raised bruises. HSP can also affect the kidneys, digestive tract, and joints. HSP can occur any time in life, but it is most common in children between 2 and 6 years of age.[1] Most people recover from HSP completely, though kidney damage is the most likely long-term complication. In adults, HSP can lead to chronic kidney disease (CKD) and kidney failure, described as end-stage renal disease when treated with blood-filtering treatments called dialysis or a kidney transplant.

What are the causes of HSP?

Henoch-Schönlein purpura is caused by an abnormal immune system response in which the body's immune system attacks the body's own cells and organs. Usually, the immune system makes antibodies, or proteins, to protect the body from foreign substances such as bacteria or viruses. In HSP, these antibodies attack the blood vessels. The factors that cause this immune system response are not known. However, in 30 to 50 percent of cases, people have an upper respiratory tract infection, such as a cold, before getting HSP.[2] HSP has also been associated with

- infectious agents such as chickenpox, measles, hepatitis, and HIV viruses
- medications
- foods
- insect bites
- exposure to cold weather
- trauma

Genetics may increase the risk of HSP, as it has occurred in different members of the same family, including in twins.

What are the symptoms of HSP?

The symptoms of HSP include the following:

- **Rash.** Leaking blood vessels in the skin cause a rash that looks like bruises or small red dots on the legs, arms, and buttocks. The rash may first look like hives and then change to look like bruises, and it may spread to the chest, back, and face. The rash does not disappear or turn pale when pressed.

[1]McCarthy JH, Tizard EJ. Clinical practice: diagnosis and management of Henoch-Schönlein purpura. *European Journal of Pediatrics.* 2010;169:643–650.

[2]Appel GB, Radhakrishnan J, D'Agati VD. Secondary glomerular disease. In: Brenner BM, ed. *Brenner & Rector's The Kidney.* Vol. 1. 9th ed. Philadelphia: Saunders Elsevier; 2012: 1192–1277.

- **Digestive tract problems.** HSP can cause vomiting and abdominal pain, which can range from mild to severe. Blood may also appear in the stool, though severe bleeding is rare.
- **Arthritis.** Pain and swelling can occur in the joints, usually in the knees and ankles and less frequently in the elbows and wrists.
- **Kidney involvement.** Hematuria—blood in the urine—is a common sign that HSP has affected the kidneys. Proteinuria—large amounts of protein in the urine—or development of high blood pressure suggests more severe kidney problems.
- **Other symptoms.** In some cases, boys with HSP develop swelling of the testicles. Symptoms affecting the central nervous system, such as seizures, and lungs, such as pneumonia, have been seen in rare cases.

Though the rash affects all people with HSP, pain in the joints or abdomen precedes the rash in about one-third of cases by as many as 14 days.[1]

What are the complications of HSP?

In children, the risk of kidney damage leading to long-term problems may be as high as 15 percent, but kidney failure affects only about 1 percent of children with HSP.[1] Up to 40 percent of adults with HSP will have CKD or kidney failure within 15 years after diagnosis.[3]

A rare complication of HSP is intussusception of the bowel, which includes the small and large intestines. With this condition, a section of the bowel folds into itself like a telescope, causing the bowel to become blocked.

Women with a history of HSP who become pregnant are at higher risk for high blood pressure and proteinuria during pregnancy.

How is HSP diagnosed?

A diagnosis of HSP is suspected when a person has the characteristic rash and one of the following:

- abdominal pain
- joint pain
- antibody deposits on the skin
- hematuria or proteinuria

[3]Feehally J, Floege J. IgA nephropathy and Henoch-Schönlein nephritis. In: Floege, J, Johnson RJ, Feehally J, ed. *Comprehensive Clinical Nephrology.* 4th ed. St. Louis, MO: Elsevier Saunders; 2010: 270–281.

Antibody deposits on the skin can confirm the diagnosis of HSP. These deposits can be detected using a skin biopsy, a procedure that involves taking a piece of skin tissue for examination with a microscope. A skin biopsy is performed by a health care provider in a hospital with little or no sedation and local anesthetic. The skin tissue is examined in a lab by a pathologist—a doctor who specializes in diagnosing diseases.

A kidney biopsy may also be needed. A kidney biopsy is performed by a health care provider in a hospital with light sedation and local anesthetic. The health care provider uses imaging techniques such as ultrasound or a computerized tomography scan to guide the biopsy needle into the organ. The kidney tissue is examined in a lab by a pathologist. The test can confirm diagnosis and be used to determine the extent of kidney involvement, which will help guide treatment decisions.

Hematuria and proteinuria are detected using urinalysis, which is testing of a urine sample. The urine sample is collected in a special container in a health care provider's office or commercial facility and can be tested in the same location or sent to a lab for analysis. For the test, a nurse or technician places a strip of chemically treated paper, called a dipstick, into the urine sample. Patches on the dipstick change color when blood or protein are present in urine.

How is HSP treated?

No specific treatment for HSP exists. The main goal of treatment is to relieve symptoms such as joint pain, abdominal pain, and swelling. People with kidney involvement may receive treatment aimed at preventing long-term kidney disease.

Treatment is rarely required for the rash. Joint pain is often treated with nonsteroidal anti-inflammatory medications, such as aspirin or ibuprofen. Recent research has shown corticosteroids—medications that decrease swelling and reduce the activity of the immune system—to be even more effective in treating joint pain. Corticosteroids are also used to treat abdominal pain.

Though rare, surgery may be needed to treat intussusception or to determine the cause of swollen testicles.

HSP that affects the kidneys may be treated with corticosteroid and immunosuppressive medications. Immunosuppressive medications prevent the body from making antibodies. Adults with severe, acute kidney failure are treated with high-dose corticosteroids and the immunosuppressive cyclophosphamide (Cytoxan).

People with HSP that is causing high blood pressure may need to take medications that—when taken as prescribed by their health care provider—lower their blood pressure and can also significantly slow the progression of kidney disease. Two types of blood pressure lowering medications, angiotensin-converting enzyme (ACE) inhibitors and angiotensin receptor blockers (ARBs), have proven effective in slowing the progression of kidney disease. Many people require two or more medications to control their blood pressure. In addition to an ACE inhibitor or an ARB, a diuretic—a medication that helps the kidneys remove fluid from the blood—may be prescribed. Beta blockers, calcium channel blockers, and other blood pressure medications may also be needed.

Blood and urine tests are used to check the kidney function of people with HSP for at least 6 months after the main symptoms disappear.

Eating, Diet, and Nutrition

Eating, diet, and nutrition have not been shown to play a role in causing or preventing HSP.

Points to Remember

- Henoch-Schönlein purpura (HSP) is a disease that causes small blood vessels in the body to become inflamed and leak.
- HSP is caused by an abnormal immune system response in which the body's immune system attacks the body's own cells and organs. The factors that cause this immune system response are not known.
- The symptoms of HSP include the following:
  - rash
  - digestive tract problems
  - arthritis
  - kidney involvement
- In children, the risk of kidney damage leading to long-term problems may be as high as 15 percent, but kidney failure affects only about 1 percent of children with HSP. Up to 40 percent of adults with HSP will have CKD or kidney failure within 15 years after diagnosis.
- A diagnosis of HSP is suspected when a person has the characteristic rash and one of the following:
  - abdominal pain
  - joint pain
  - antibody deposits on the skin
  - hematuria or proteinuria
- Antibody deposits on the skin can confirm the diagnosis of HSP.
- No specific treatment for HSP exists. The main goal of treatment is to relieve symptoms such as joint pain, abdominal pain, and swelling. People with kidney involvement may receive treatment aimed at preventing long-term kidney disease.

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.

Participants in clinical trials can play a more active role in their own health care, gain access to new research treatments before they are widely available, and help others by contributing to medical research. For information about current studies, visit *www.ClinicalTrials.gov*.

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