

HENOCH-SCHÖNLEIN PURPURA (IGA VASCULITIS)

WHAT IS HENOCH-SCHÖNLEIN PURPURA?

Henoch-Schönlein purpura (HSP) is a systemic vasculitis that causes the blood vessels in the skin to become inflamed, causing red spots. When the blood vessels in the skin get inflamed, they can bleed, causing a rash that is called purpura. This rash is typically seen on the lower legs or arms. The specific skin lesion is characterized by the tissue deposition of an immune system product, called IgA immunoglobulin, which is also found in kidneys of patients with a renal disease, called IgA nephropathy.

SYMPTOMS

The four main characteristics of Henoch-Schonlein purpura include:

1. **Rash (purpura).** Reddish-purple spots, which look like bruises, are the most distinctive and universal sign of Henoch-Schonlein purpura. The rash develops mainly on the buttocks, legs and feet, but it can also appear on the arms, face and trunk and may be worse in areas of pressure, such as the sock line and waistline.
2. **Swollen, sore joints (arthritis).** People with Henoch-Schonlein purpura often have pain and swelling around the joints — mainly in the knees and ankles. Joint pain sometimes precedes the classical rash by one or two weeks. These symptoms subside when the disease clears and leave no lasting damage.
3. **Gastrointestinal symptoms.** Many children with Henoch-Schonlein purpura develop gastrointestinal symptoms, such as abdominal pain, nausea, vomiting or bloody stools. These symptoms sometimes occur before the rash appears.
4. **Kidney involvement.** Henoch-Schonlein purpura can also affect the kidneys. In most cases, this shows up as protein or blood in the urine, which you may not even know is there unless you have a urine test done. Usually this goes away once the illness passes, but in a few cases, kidney disease may develop and even persist.

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CAUSE

In Henoch-Schonlein purpura, some of the body's small blood vessels become inflamed, which can cause damage in the skin, abdomen and kidneys. Why this initial inflammation develops isn't clear. It may be the result of the immune system responding inappropriately to certain triggers. Nearly half the people who have Henoch-Schonlein purpura developed the disease after an upper respiratory infection, such as a cold. Infectious triggers may include chickenpox, strep throat, measles and hepatitis. Other triggers may include certain medications, food, insect bites or exposure to cold weather

TREATMENT

Henoch-Schonlein purpura usually goes away on its own within a month with no lasting ill effects. Bed rest, plenty of fluids and over-the-counter pain relievers may help.

However, specific treatment is recommended in patients with marked proteinuria (protein in the urine) and/or impaired kidney function during the acute episode. A kidney biopsy can be performed to reveal the severity of the lesions which appears to be the best indicator of prognosis. Advanced disease, usually defined as crescentic nephritis, is treated with a regimen consisting of pulse intravenous corticosteroids, methylprednisolone followed by oral prednisone.

Other regimens that have been evaluated in children with kidney disease include corticosteroids and azathioprine and multidrug regimens such as cyclophosphamide. However, since spontaneous recovery is often observed in these patients, it remains unknown whether these regimens are superior to no or less aggressive therapy.