

## Henoch-Schönlein Purpura (HSP, Spring Fever) — Symptoms and Diagnosis

[See online here](#)

**Henoch-Schönlein purpura (HSP), also called immunoglobulin A vasculitis (IgAV), is an IgA-mediated disorder and the most common systemic case of vasculitis in children.**



### Definition of HSP

#### Henoch-Schönlein Purpura as self-limited disease

HSP is an autoimmune disease characterized by the **depositing of IgA** in multiple [tissues](#) and small vessel walls of the [skin](#), GIT, joints, [kidneys](#), and, rarely, the central nervous system (CNS) and [lungs](#). This acute, self-limiting disease is a subset of **necrotizing vasculitis** and is characterized by **fibrinoid destruction of vessels and leukocytoclasia**.

# Epidemiology of HSP

## Mainly children affected by Henoch-Schönlein Purpura

Although HSP is typically found in **children between the ages of 3 and 10**, the disease also affects adults. It is slightly more common in males than females; the male-to-female ratio of the disease is 2:1. The disease is also seen in adults where the male: female ratio is estimated to be 1:1. The estimated annual prevalence of HSP in the United States is 14-15 cases per 100,000 individuals; in the United Kingdom, the incidence is 20.4 cases per 100,000 individuals.

HSP is more common among whites than African Americans in the United States. It is preceded by an URTI or GIT infection for 1-3 weeks in half to two-thirds of patients. Most patients also present with rashes, gastrointestinal symptoms, and joint pains.

## Etiology of HSP

The etiology of the disease is multifactorial with three main factors being incriminated:

1. Genetic
2. Environmental
3. Autoimmune/antigenic. (reportedly, upper respiratory tract, pharyngeal, and gastrointestinal infections occur in more than 75% of cases)

### Infections that may precede the development of HSP:

- Group A streptococcal infections (most common)
- Mononucleosis
- Hepatitis
- Mycoplasma infection
- *Campylobacter enteritis*
- Subacute bacterial endocarditis

### Vaccinations

- Typhoid and paratyphoid
- Measles
- Cholera
- Yellow fever

### Environmental factors

- Horse serum
- Insect bites
- Cold exposure
- Certain drugs (ampicillin, penicillin, quinidine, quinine, losartan)

It is also associated with IgA nephropathy.

## Pathology and Pathophysiology of HSP

## Deposition of IgA in Henoch-Schönlein Purpura

**Increased serum IgA concentrations** clearly indicate the critical role played by immunoglobulin in the development of HSP. This is characterized by depositing of IgA and complement C3 complexes in small vessels, resulting in purpura and petechiae. Findings depend on the area being supplied by the vessel.

Complex formation is triggered by **antigen exposure**, either an infective organism or a drug. Group A streptococcal organisms are the most common cause.

## Symptoms of HSP

### Characteristic set of symptoms in HSP

The systemic depositing of IgA results in a characteristic set of symptoms, not all of which may be experienced by patients and which are typically preceded by a prodrome of headache, fever, and/or anorexia.

**After the prodrome, the most common reported symptoms are:**



Image: "Henoch-Schönlein Purpura" by Madhero88/Thomas Habif. License: [CC BY-SA 3.0](https://creativecommons.org/licenses/by-sa/3.0/)

- **Palpable purpura:** This is the hallmark of HSP (occurs in 95%–100% of cases) and is characterized by reddish-purple spots mainly in the area of the buttocks, legs, and feet. The spots appear to be worse in areas of pressure (e.g., the waistline and where sock elastic typically sits on the calves). In children, it often appears in crops, although it may not be the presenting feature.
- **Arthralgia:** This produces swollen, sore joints of the ankle and knee, often preceding a rash by 1 or 2 days. It does not cause joint damage, however.
- **Gastrointestinal symptoms:** Colicky abdominal pain is the most common symptom. Children also often develop symptoms such as nausea, vomiting, intussusception, bowel infarction, and bloody stools. The triad of joint pain, abdominal symptoms, and rash is also often seen.

- **Renal disease:** Kidney involvement manifests as protein or blood in the urine, which can be detected in a urine test. Kidney disease may persist.

**Other symptoms are:**

- Subcutaneous edema
- Bloody stools
- Scrotal edema

## Diagnosis of HSP

### Henoch-Schönlein Purpura clinically diagnosed

The diagnosis of HSP is a clinically driven one, but lab investigations are required to rule out other possible causes of the condition. According to the American College of Rheumatology, the presence of 2 out of 4 of the features listed below has a diagnostic sensitivity of 87.1 % and specificity of 87.7%:

- Palpable purpura without [thrombocytopenia](#)
- Age of onset <20
- Bowel angina
- Biopsy showing granulocytes in the walls of small vessels.



*Image:* "Immunostaining for IgA in a patient with Henoch-Schönlein nephritis." by Lazarus Karamadoukis, Linmarie Ludeman and Anthony J Williams. License: [CC BY 2.0](#)

While the biopsy of tissue revealing deposition of immune complexes in vessel walls and mesangium (in case of the kidney) is the most accurate test, it is not necessary to undertake in the majority of the cases because the disease is benign and self-limited.

**Other lab investigations include:**

- Complete blood count for leukocytosis, eosinophilia, and thrombocytosis
- Platelet count; may be elevated
- ESR; may be mildly elevated (75%)
- Factor XIII; may be reduced by 50%
- Urinalysis; hematuria or proteinuria may be present
- Renal profile; elevated blood urea nitrogen, creatinine, and deranged electrolytes
- Serum IgA; increased in 50% of cases
- Circulating immune complexes
- C3 and C4; sometimes reduced

The following table shows the usefulness of imaging modalities in diagnosing HSP.

Imaging Test	Significance
Ultrasonography	Significant in: <ul style="list-style-type: none"> <li>• Intussusception</li> <li>• Scrotal imaging</li> </ul>

Radiography	Helpful in diagnosing intestinal obstruction and may be advised in case of hemoptysis
MRI and CT	In neurologic findings
Endoscopy	For evaluating abdominal symptoms

## Differential Diagnosis of HSP

### Similar diseases such as Henoch-Schönlein Purpura

Before diagnosing HSP, other conditions that should be taken into consideration and ruled out are:

- [Acute glomerulonephritis](#)
- [Acute renal failure](#)
- Bacterial endocarditis
- Disseminated intravascular coagulation
- Idiopathic thrombocytopenic purpura
- Ig A nephropathy
- [Inflammatory bowel disease](#)
- Meningitis

## Therapy of HSP

### Supportive treatment in Henoch-Schönlein Purpura

Management of HSP mostly involves **supportive treatment** as no therapy has been shown to be effective enough to shorten the duration of the disease. As well, as a self-limited disease, the condition resolves on its own over a period of several weeks.

Prednisone does not affect renal disease or long-term outcome. Symptoms usually resolve after 1 to 2 months, although it is common for patients to have a recurrence of the disease up to 4 months later.

Management involves:

#### **Supportive treatment includes:**

- Adequate hydration
- Observing for renal and abdominal complications
- Discontinuation of unnecessary drugs
- Symptomatic treatment of edema, malaise, and joint aches

#### **Pharmacologic therapy may include:**

- Analgesics and nonsteroidal anti-inflammatory drugs for joint pain
- Immunosuppressants such as corticosteroids (for severe abdominal complaints, scrotal edema, CNS involvement, and persistent nephritic syndrome)
- Antihypertensives (for renal involvement)
- Factor VIII concentrate (if steroids are contraindicated)
- Plasmapheresis to delay kidney disease progression
- Surgery for bowel ischemia or kidney transplantation
- Long-term monitoring with urinalysis and blood pressure monitoring

# Progression and Prognosis of HSP

Because HSP is a benign disease, the **prognosis is excellent**, with spontaneous resolution. Initial episodes of the condition may last several months and there may be relapses, however. Chances for complete resolution are positive if:

- There is mild kidney involvement.
- There are no neurologic complaints.
- The initial duration of the disease is <6 weeks.

Children younger than the age of 3 usually experience a mild course of the disease.

Long-term prognosis of HSP depends on the severity of renal and GI involvement, which can cause significant morbidity and mortality.

## References

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