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

Being an IgA mediated disorder, HSP, the most common systemic vasculitis in children, is also known as Immunoglobulin A vasculitis (IgAV).

Definition of HSP

Henoch-Schönlein Purpura as self-limited disease

Henoch-Schönlein Purpura (HSP) is an autoimmune disease characterized by IgA deposition in multiple tissues and small vessel walls of the skin, GIT, joints, kidneys and rarely the CNS and lungs. The acute and 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

Typically found in children of age between 3 and 10, HSP also affects adults, being slightly more common in females. The male-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 prevalence of HSP in the United States is 14-15 cases per 100,000 individuals, whereas in the UK, there’s an annual estimated incidence of 20.4 cases per 100,000 individuals.

HSP is more common among the whites compared to African Americans. The disease is preceded by an URTI or GIT infection for 1-3 weeks in half to two-thirds of the patients. Most patients 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 factors.
2. Environmental factors.
3. Autoimmune/antigenic factors. This theory is supported by the reports of URT, pharyngeal and GI infections in more than 75% of the 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 the immunoglobulin in the development of the disease. This is characterized by deposition 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**, which could be an infective organism or a drug. Group A of streptococcal organisms is the most common cause. Other possible triggers are listed above.

### Symptoms of HSP

#### Characteristic set of symptoms in HSP

IgA systemic deposition results in a characteristic set of symptoms. Not all symptoms may be experienced by the people suffering from HSP and are typically preceded by a prodrome of

- Headache
- Fever
- Anorexia

After the prodrome, the most common reported symptoms are:

![Image: “Henoch-Schönlein Purpura” by Madhero88/Thomas Habif. License: CC BY-SA 3.0](image)

**Palpable Purpura** – the hallmark of HSP (95-100 % the cases) characterized by reddish-purple spots mainly in the area of the buttocks, legs and feet and is worse on areas of pressure (waistline and sock line). In children, it often appears in crops and may not be the presenting feature.

**Arthralgia** – swollen, sore joints involving the ankle and **knee**, often preceding a rash by one or two days. It doesn’t cause any joint damage as it’s not true arthritis.

**Gastrointestinal symptoms** – Children often develop symptoms like nausea, vomiting, intussusception, bowel infarction and bloody stools. The triad of joint pain, abdominal symptoms and rash are often seen in children. Colicky abdominal pain is the most common symptom.

**Renal disease** – kidney involvement manifests as protein or blood in the urine 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 HSP diagnosis 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](image)

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’s pointless to perform in majority of the cases because the disease is benign and self-limited. Other lab investigations include:

- CBC - Leukocytosis, eosinophilia and thrombocytosis
- Platelet count - may be elevated
- ESR - mildly elevated (75 %)
- Factor XIII - reduced in 50 %
- Urinalysis - Hematuria or proteinuria may be present
- Renal profile - elevated BUN and Creatinine and deranged electrolytes
- Serum IgA - increased in 50 % of the cases
- Circulating immune complexes may be present
- C3 and C4 - occasionally reduced.

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

<table>
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<tr>
<th>Imaging Test</th>
<th>Significance</th>
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| Ultrasonography | Significant in:  
• Intussusception  
• Scrotal imaging |
| 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
- DIC
- ITP
- Ig A nephropathy
- IBD
- Meningitis

Therapy of HSP

Supportive treatment in Henoch-Schönlein Purpura

Management of HSP mostly involves supportive treatment as there has been no therapy effective enough to shorten its duration. Also, being 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. Usually resolution after 1-2 months, common to have recurrence up to 4 months later.

Management involves:

Supportive treatment, including:

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

Pharmacologic therapy

- Analgesics and NSAIDS for joint pain;
- Immunosuppressants like corticosteroids (in severe abdominal complaints, scrotal edema, CNS involvement, persistent nephritic syndrome);
- Anti hypertensive (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 involving urinalysis and blood pressure monitoring.

Progression and Prognosis of HSP

Being a benign disease, the prognosis is excellent with spontaneous resolution. Initial episodes of the condition may last several months and there may be relapses as well. Complete resolution chances are especially fair if:

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

Children younger than 3 years have a mild course as well. The long-term prognosis of HSP depends on the severity of renal and GI involvement, which are causes of considerable morbidity and mortality.

References


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