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A retrospective study on Henoch-Schönlein purpura

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Abstract
Henoch-Schönlein Purpura (HSP) is a systemic vasculitis primarily affecting children, characterized by palpable purpura, arthritis, abdominal pain, and renal involvement. Henoch-Schönlein purpura is a prevalent form of vasculitis that mostly affects children, but may also occur in adults. The medical symptoms are believed to result from the deposition of IgA in the blood vessel walls of the afflicted organs, namely the skin, gastrointestinal system, joints, and kidneys. Corticosteroids may be useful in quickly resolving renal symptoms and alleviating joint and gastrointestinal pain. However, there is little evidence to support their effectiveness in treating organ manifestations and consequences, such as glomerulonephritis, bowel infarction, or intussusception. This study aims to provide a comprehensive overview of HSP, including its epidemiology, pathophysiology, clinical manifestations, diagnostic criteria, and management strategies. By synthesizing current research and clinical findings, this paper seeks to enhance understanding and improve patient outcomes.

Keywords: HSP, vasculitis, children, renal, corticosteroids

Introduction
Henoch-Schönlein Purpura, also known as IgA vasculitis, is the most common vasculitis in children, with a significant incidence in the pediatric population. Despite its self-limiting nature in many cases, HSP can lead to serious complications, particularly renal involvement, which necessitates prompt diagnosis and management. Henoch-Schönlein purpura (HSP) is a prevalent form of vasculitis in children, with an annual incidence rate of 10-20 cases per 100,000 children [1, 2]. The majority of patients, over 90%, are less than 10 years old, with an average age of 6 years. HSP is a kind of vasculitis that affects tiny blood vessels and involves the destruction of white blood cells [3, 4]. The clinical manifestation of this condition involves the presence of skin lesions that may be felt, joint discomfort, kidney involvement, severe stomach pain, and bleeding in the gastrointestinal tract. The majority of HSP cases occur during the fall and winter seasons. The suggested factors that might initiate a response include upper respiratory tract infections, medicines, vaccines, and malignancies [5]. The underlying mechanisms responsible for the development of HSP are not yet fully understood. HSP, also known as Henoch-Schönlein purpura, is often a benign and self-limiting condition [8]. However, if nephritis occurs concurrently, it may lead to serious consequences [6, 7]. The diagnosis of HSP is based on certain criteria. In 2010, the European League Against Rheumatism (EULAR), the Paediatric Rheumatology International Trials Organization (PRINTO), and the Paediatric Rheumatology European Society (PRES) released an updated set of criteria that demonstrated both high sensitivity and specificity. The therapy is determined by the degree of the illness and the organs that are affected [9]. Typically, the therapy for HSP without renal involvement is focused on managing symptoms. HSPN is often managed with corticosteroids or other immunosuppressive and immunomodulatory medications. Current research has not reached a definitive conclusion about the preferred drug.

Epidemiology
HSP predominantly affects children between the ages of 3 and 15, with a peak incidence at around 5 years old. The annual incidence rate ranges from 10 to 20 cases per 100,000 children. There is a slight male predominance, with a male-to-female ratio of approximately 1.5:1. Seasonal variations suggest a higher incidence in autumn and winter. Henoch-Schönlein purpura has less severe symptoms in newborns and toddlers who are under the age of two [10]. HSP is prevalent form of vasculitis amongst children and have slight male predilection too [11].

Criterion	Description
Mandatory criterion	Purpura or petechiae with lower limb predominance
Minimum 1 out of 4 criteria	1. Diffuse abdominal pain with acute onset 2. Histopathology showing leukocytoclastic vasculitis or proliferative glomerulonephritis, with predominant immunoglobulin A (IgA) deposits 3. Arthritis or arthralgia of acute onset 4. Renal involvement in the form of proteinuria or haematuria

Fig 1: Diagnostic criteria: HSP

Pathophysiology: The exact etiology of HSP remains unclear, though it is widely believed to be an immune-mediated disorder. The deposition of IgA-containing immune complexes in small vessel walls is a hallmark of the disease. Triggers such as infections, medications, and vaccinations have been implicated in precipitating the immune response. Henoch-Schönlein purpura is characterized by the deposition of immunoglobulin A (IgA) immune complexes in tiny blood vessels, leading to the development of petechiae and palpable purpura. Gastrointestinal bleeding may arise when immune complexes form in the tiny capillaries of the gut wall [12]. Immune complexes impacting the renal mesangium may result in a range of glomerulonephritis, from moderate proliferative to severe crescentic. The presence of an antigen resulting from an illness, treatment, or other environmental element may initiate the development of antibodies and immunological complexes [13]. Group A streptococcus has been detected in cultures from over 30 percent of children diagnosed with Henoch-Schönlein nephritis. Additionally, individuals with Henoch-Schönlein nephritis are more likely to have positive serum antistreptolysin-O titers. Considered viral and bacterial causes of HSP include Coxsackie virus, adenovirus, hepatitis A and B viruses, mycoplasma, and methicillin-resistant Staphylococcus aureus [14].

Clinical Manifestations: The typical tetrad of HSP comprises visible purpura, joint pain, gastrointestinal symptoms, and renal involvement (Fig. 2). These clinical manifestations may emerge gradually over a period of days to weeks [15]. The sequence of delivery may differ. The typical initial manifestation includes purpura and arthralgia. In 2016, a research examined the clinical signs of 260 individuals with HSP [16]. All patients displayed purpura at diagnosis. Among the patients, 62% had joint involvement, 70% had glomerulonephritis, and 53% had gastrointestinal involvement [17]. A study compared indicators in 75 adults and 208 children diagnosed with HSP [18]. Joint involvement and stomach discomfort were more prevalent in children compared to adults. There was a higher prevalence of adult people with lower extremities edema and hypertension. Uncommon clinical signs of HSP include inflammation of the blood vessels in the brain, bleeding in the testicles, and bleeding in the lungs [19].

HSP is characterized by a tetrad of symptoms

1. **Palpable Purpura:** Non-thrombocytopenic purpura predominantly on the lower extremities and buttocks.
2. **Arthritis/Arthralgia:** Transient, migratory joint pain, often affecting the knees and ankles.
3. **Abdominal Pain:** Colicky pain, which may be associated with gastrointestinal bleeding.
4. **Renal Involvement:** Hematuria, proteinuria, and in severe cases, nephrotic syndrome or acute renal failure.

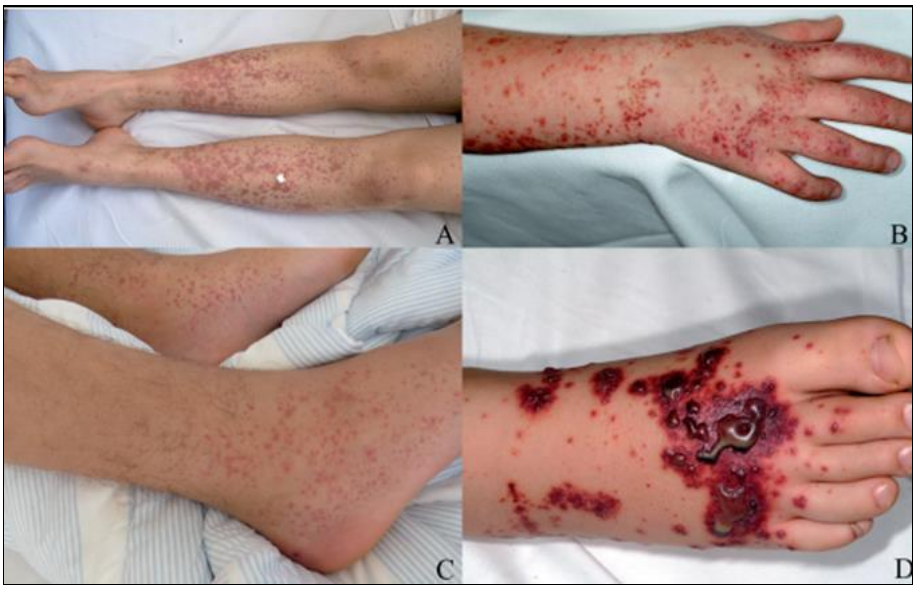

Fig 2: (A and B) Skin lesions (C) Arthritis and purpura (D) Necrotic lesions

Diagnostic Criteria

A conclusive diagnostic test for HSP does not exist. The presence of purpura, stomach discomfort, and arthritis together should be a cause for worry in a clinical setting.

Palpable purpura, which is evident without thrombocytopenia, is very indicative and may be seen in all individuals. The punch biopsy of the skin is a valuable diagnostic tool for demonstrating the distinctive leukocytoclastic vasculitis. The renal biopsy will reveal a membranoproliferative glomerulonephritis that is comparable to IgA nephropathy [21, 22]. The inclusion criteria for this study were as follows: patients who were 20 years old or younger at the time of commencement, presence of palpable purpura (without thrombocytopenia), experiencing bowel angina (characterized by widespread abdominal pain or diagnosis of bowel ischemia), and histologic evidence of granulocytes in the tiny walls of arterioles and venules (leukocytoclastic vasculitis). In 2006, the criteria underwent revisions that included making palpable purpura a required characteristic, eliminating the age requirement, introducing arthritis as a criterion, and substituting granulocytes in biopsy specimens with IgA deposition [23].
Diagnosis is primarily clinical, supported by laboratory and histopathological findings.

Management
HSP is generally self-limiting, with a good prognosis in most cases. Management focuses on symptomatic relief and monitoring for complications. However, NSAIDs may worsen gastrointestinal symptoms and should not be taken in individuals with established kidney problems. During the active period of the sickness, it is advisable to keep the afflicted extremities elevated and at rest in order to avoid the occurrence of purpura. Patients should be informed that they may encounter repeated episodes of purpura as they increase their degree of physical activity [24].
Inpatient care may be necessary in cases when there is a lack of sufficient outpatient monitoring or when hospitalization is needed for managing conditions such as dehydration, bleeding, or pain control. A referral to a nephrologist is advised in cases when there is substantial renal involvement. Renal biopsy is necessary in individuals with severe renal illness to establish a conclusive diagnosis and direct the appropriate treatment [25]. Early use of steroids is more suitable for children who have kidney involvement or severe symptoms outside of the kidneys. Additionally, it may assist in alleviating edema in the scrotum. Administering oral prednisone at a dosage of 1 to 2 mg per kg per day for a duration of two weeks has been used as a treatment for children with mild to severe stomach and joint symptoms, as well as to expedite the clearance of Henoch-Schönlein purpura (25). An experiment conducted using a double-blind randomized trial revealed that administering prednisone at an early stage resulted in a reduction in the degree of stomach and joint pain in children. While prednisone did not have a preventive effect on renal illness, it proved to be beneficial in the treatment of renal disease after it had already developed. A meta-analysis revealed that the use of corticosteroids in children diagnosed with Henoch-Schönlein purpura resulted in a decrease in the average duration of stomach discomfort and a reduction in the likelihood of developing permanent renal impairment [26]. The use of steroids did not have any impact on the clearance of the purpura, and no adverse effects were seen. Children and adults with significant renal damage are advised to undergo early aggressive treatment [27]. Possible treatment options include the administration of large doses of steroids with immunosuppressants, high doses of intravenous immunoglobulin, plasmapheresis, and kidney transplant.

Key aspects include
- **Pain Management:** NSAIDs for arthritis and abdominal pain.
- **Corticosteroids:** Indicated for severe abdominal pain, gastrointestinal bleeding, or renal involvement.
- **Monitoring:** Regular follow-up for renal function, particularly in patients with initial renal involvement.

Complications
Renal involvement is the most significant complication, with potential progression to chronic kidney disease. Gastrointestinal complications, such as intussusception, can also occur. Long-term follow-up is essential to monitor for these complications.

Conclusion
Henoch-Schönlein Purpura, while often self-limiting, can lead to serious complications, particularly renal. Early recognition and appropriate management are crucial to improving outcomes. Recent years have seen significant advancements in research, resulting in an improved, but incomplete, comprehension of the pathophysiology. Treatment involves the use of drugs that alleviate symptoms and ultimately reduce or modify the immune response. We suggest limiting the use of immunosuppressants and immunomodulators to instances that are chronic, persistent, recurring, or complex. Subsequent multicentre investigations including both children and adults should ascertain the appropriateness of corticosteroids and evaluate other medications that may reduce the need for steroids, such as rituximab or dapsone, for specific organ-related symptoms. A clinically-oriented therapy protocol for HSP, depending on the symptoms of the illness, is required. Additional study is required to clarify the fundamental processes and enhance therapeutic options.

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