



ReviewArticle

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## ***An Overview of Henoch Schonlein Purpura Diagnosis and Management Approach: Literature Review***

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### ABSTRACT

Henoch-Schönlein purpura (HSP) is a systemic autoimmune disease of childhood mainly, but can also be seen in adolescence and adults. It is considered the most common vasculitis in the pediatric group. Recently, it has been termed IgA vasculitis due to the presence of immunoglobulin A in these patients. Eventually, patients develop a generalized vasculopathy in multiple areas, including skin, and other systems such as the gastrointestinal, urinary, and pulmonary systems can be affected. Rarely, more severe involvement of the central nervous system and lungs can happen. We aimed to review the literature reviewing the pathophysiology of Henoch-Schönlein purpura, along with its clinical features, diagnosis, and management. PubMed database was used for article selection. Papers were selected, obtained, and reviewed based on our inclusion and exclusion criteria. Henoch Schonlein Purpura is the most common vasculitis in the pediatric population, which reflects the heavy burden it causes on the health care system and the patients themselves. As a result, pediatricians must have a complete understanding of this disease to approach this disease properly. Managing this disease, focuses on supportive care and subsiding the inflammatory status with corticosteroid in some cases. Early diagnosis and treatment are pivotal to avoid possible complications and the need for surgery in some severe presentations.

**Key words:** *Henoch schonlein purpura, Diagnosis, Clinical features, Management*

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### INTRODUCTION

Henoch-Schönlein purpura (HSP) is a systemic autoimmune disease of childhood mainly, but can also be seen in adolescence and adults [1-4]. Most cases present before the age of 10, but adults will likely present with a severe form of the disease and long-term complications (mainly renal). Nevertheless, recently, it has been termed as IgA vasculitis in some studies due to the presence of immunoglobulin A. This immunoglobulin is noted in the vessels

in these patients and is considered the main culprit of the disease [5]. Eventually, patients develop a generalized vasculopathy in multiple areas, including skin, and other systems such as the gastrointestinal, urinary, and pulmonary systems can be affected. Rarely, more severe involvement of the central nervous system and lungs can happen [6]. It is considered the most common vasculitis in a pediatric group with a prevalence of up to 20 cases per 100,000 yearly, while adults having a prevalence of approximately 14 cases per 100,000 per year. Recurrence of the disease is noted in many cases, with some papers reporting a rate of 2.7% to 30%, with varied average intervals between the first and second episodes [7-9]. In this paper, we will review this disease's pathophysiology, clinical features, diagnosis, and management.

## MATERIALS AND METHODS

PubMed database was used for articles selection, and the following keys used in the mesh ("Henoch Schonlein Purpura"[Mesh]) AND ("Diagnosis "[Mesh]) AND ("Clinical Features"[Mesh]) AND ("Management"[Mesh])). In regards to the inclusion criteria, the articles were selected based on the inclusion of one of the following topics; Henoch Schonlein Purpura and its clinical features, diagnosis, and management. Exclusion criteria were all other articles that did not have one of these topics as their primary endpoint.

### *Review*

This disease has been heavily studied due to its significant prevalence among all vasculopathy. As a result, many genetic, environmental, and antigenic factors have been associated with the etiology of Henoch-Schonlein purpura. One of the most reported associations is a preceding infection which is usually an upper respiratory tract infection. Other notable infections include pharyngeal and gastrointestinal tract infections. These findings drove many studies to culture for relevant bacteria in these patients. Group A Streptococcus was the most notable found bacteria with more than 30% of HSP nephritis. Many other pathogens precede HSP, such as hepatitis A, hepatitis B coxsackievirus, parvovirus B19, Mycoplasma, and adenoviruses among others [7].

### *Pathophysiology*

Henoch-Schonlein purpura is like many autoimmune diseases where its pathophysiology has not been fully figured. Nevertheless, IgA-antibody immune complexes have been proved to have a major role in the pathophysiology of the disease. These complexes usually form in response to an antigenic exposure from medication or infection. These will usually deposit in the vessels (mainly the small capillaries) of multiple organs, including kidneys, skin, gastrointestinal tract, joints, lung, and CNS. As a result, some inflammatory mediators (e.g. C3 receptor lymphocytes, and prostaglandins) can bind to these as well and deposit there too, adding to hyper-inflammatory response status. These deposits are believed to cause the clinical symptoms of HSP noted in the aforementioned organs [10, 11].

### *Clinical features*

In children, the course of HSP is rather benign, unlike adults. A typical triad of headache, fever, and aversion to food is seen in most cases of the disease. Other constitutional symptoms like lethargy and myalgia, along with fever have been associated with a higher risk of other symptoms (e.g. GI and renal). As the disease progresses insidiously, a characteristic rash develops on the lower limbs. This is often one of the most important initial signs of HSP. Upon examination, this rash is a red macular lesion, with often raised blanching papules. These eventually develop into palpable purpura, hence the term HSP. An interesting feature of these skin lesions is that they are gravity-dependent. They appear in the buttocks of newborns, on the face and trunk of crawling babies, and in the lower limbs of walking children. However, around 25% of patients do not present to the clinic with the rash, but rather with other symptoms, then develop the rash after a few days. Interstitial oedema may develop, encompassing the scrotum in boys. In young children, subcutaneous oedema may also appear on the face, and around the eyes, unlike in adults where it would affect the upper limb (specifically the hands). Children may also present with joint pain and swelling of the joints, especially of the lower limbs. These joints are tender on examination and appear visibly swollen. Commonly, gastrointestinal symptoms are present with nausea, vomiting, aversion to food, as well as a severe abdominal pain due to intestinal bleeding (which manifests as melena or hematemesis). The nearby renal system can be affected as well by the autoimmune disturbance, resulting in dark urine. Moreover, patients may develop nephrotic syndrome, hence, periorbital oedema finding upon examination [12, 13].

*Diagnosis*

Henoch-Schonlein purpura is commonly diagnosed based on clinical grounds. A pediatrician shall be able to identify the classical palpable purpura pattern on the lower limbs of an affected child. The other clinical manifestations of this autoimmune disease are required and should include at least one of the following; abdominal pain, joint arthralgia, nephrotic syndrome manifestations, and/or a renal biopsy supporting the diagnosis. Moreover, certain laboratory results will support the diagnosis of this disease, especially findings of reduced platelets and increased coagulation factors. However, unfortunately, there are no biomarkers diagnostically accurate for these conditions, and therefore, clinical examination combined with laboratory findings is currently the standard for diagnosis. **Table 1** shows the main diagnostic features according to European League Against Rheumatism (EULAR) and the Pediatric Rheumatology European Society (PReS) classification criterion. It is recommended that the pediatrician exclude any other potential causes of these manifestations [14]. Imaging techniques are mainly done to further confirm the disease and rule out another differential diagnosis. In boys with scrotal swelling, an ultrasound of the scrotum and abdomen should be taken to exclude herniation, testicular torsion, orchitis, and varicoceles. Varicoceles could be a result of a renal tumor, which will manifest with scrotal oedema and nephrotic disturbance. In children with abdominal pain or lower gastrointestinal bleeding, ultrasound and radiographic images should be taken, as intussusception and intestinal obstruction need exclusion. This is often followed by an endoscopy to look for the etiology, which are often purpuric lesions and ulcerations. In the case of HSP, these can be found anywhere along the gastrointestinal tract. Rarely, HSP patients may develop central nervous system symptoms and signs, and in these cases, magnetic resonance imaging is useful for the assessment of these abnormalities [15]. Moreover, a thorough work-up of renal functions is usually done in these patients, and a renal biopsy is indicated when an unremitting nephrotic syndrome is present. Patients with HSP usually have crescentic glomerulonephritis changes which signify a severe disease. However, in milder cases, the results would usually show a mesangial proliferation. If the latter includes pathognomonic diffuse immunoglobulin A deposits and C3 complement, then HSP pathology is confirmed. This is because HSP patients lack a classical complement pathway activation, a finding that is not characteristic of any other autoimmune inflammations of the glomerulus [11, 14, 15].

**Table 1.** Childhood IgA vasculitis classification criteria

Criterion	Description
Mandatory feature:	- Purpura or petechia (in a lower limb predominance fashion)
One of the following four features:	- Proteinuria or hematuria. - Confirmed histology showing HSP features (leukocytoclastic vasculitis or proliferative glomerulonephritis with predominant IgA deposition). - Acute onset diffuse abdominal colicky pain (and/or gastrointestinal bleeding and intussusception). - Acute onset arthralgia or arthritis.

*Management*

Before the diagnosis is confirmed, management shall be started with supportive therapy. This includes adequate fluid resuscitations, especially in children who lost fluids to vomiting or gastrointestinal bleeding. Patients with the latter or nephrotic syndrome should be monitored by laboratory tests of renal function. The child should be offered an oral diet whenever possible if the vomiting remits. Pain should be addressed and effort made to alleviate joint pain and scrotal or periorbital oedema. These can be managed using acetaminophen or NSAIDs such as ibuprofen or naproxen. Overall, this disease is usually self-limiting and the aforementioned therapy is sufficient in most cases. However, severe cases will need further treatment depending on the case. Patients with the persistent nephrotic syndrome usually will be managed with a dose of corticosteroids which is proved beneficial in many patients. This treatment option is also beneficial in patients with glomerulonephritis (confirmed with biopsy), active gastrointestinal melena, hematemesis, soft-tissue oedema, and/or scrotal swelling. Some studies reported evidence of a beneficial corticosteroid treatment even in cases with central nervous system involvement. In patients where the risk of nephrotic syndrome development is high, some prophylaxis to prevent its progression may be initiated. This can be offered with immunosuppressants (azathioprine if steroid-resistant has been suggested), angiotensin-converting enzyme inhibitors, and high-dose intravenous immunoglobulin G. There is some recent evidence in patients with progressive IgA-induced nephritis for tonsillectomy combined with steroidal pulse treatment. Moreover, plasmapheresis has been shown some benefits in some patients [10, 16-18].

Surgery is not uncommonly used in HSP patients but is limited to certain co-morbid conditions and presentations. Patients with intestinal obstruction and melena may have underlying bowel ischemia that requires surgical resection. If the above modalities fail to delay nephrotic disease progression, and the child develops chronic kidney injury, then the solution lies in renal organ transplantation [19, 20].

Even though most symptoms will mostly resolve within weeks, unfortunately, recurrence can happen. Some risk factors for recurrence in the pediatric group include underlying allergic rhinitis, steroid therapy for more than 10 days, and renal involvement. Thus, follow up is crucial in these patients where frequent urinalysis is done to screen for possible renal complications and any signs of recurrence. Usually, recurrence happens within 10 months of the first episode and after 6 months of the second episode. Any evidence of severe renal complications shall warrant an urgent referral to a nephrologist for aggressive therapy and possible kidney transplantation [9, 21, 22].

## CONCLUSION

Henoch Schonlein Purpura is the most common vasculitis in the pediatric population, which reflects the heavy burden it causes on the health care system and the patients themselves. This disease has certain classical presentations that shall raise clinical suspicion immediately in clinicians. As a result, pediatricians must have a complete understanding of this disease and the associated symptoms to approach this disease properly. Managing this disease, focuses on supportive care and subsiding the inflammatory status with corticosteroid in some cases. Early diagnosis and treatment are pivotal to avoid possible complications and the need for surgery in some severe presentations.

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