

## CLINICAL PROFILE OF HENOCH-SCHONLEIN PURPURA PATIENTS AT CHILDREN HOSPITAL FAISALABAD

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### Abstract

**Background:** Henoch-Schönlein Purpura (HSP), also known as IgA Vasculitis, is the most common small-vessel vasculitis in children, characterized by IgA deposition in affected tissues. Despite its global prevalence, limited data is available on its clinical profile in the Pakistani pediatric population.

**Objective:** To determine the frequency of clinical features of HSP in children presenting to Children Hospital Faisalabad.

**Methods:** A cross-sectional study was conducted over one year using non-probability consecutive sampling. A total of 215 children aged 1 month to 18 years diagnosed with HSP as per EULAR/PRINTO/PRES criteria were included. Data were collected through clinical examination, lab investigations, and imaging, then analyzed using SPSS version 25.

**Results:** Among 215 patients, palpable purpura was observed in 100%, joint involvement in 75%, GI symptoms in 65%, renal involvement in 45%, leukocytosis in 46%, thrombocytosis in 34.8%, and elevated CRP in 43% of patients. The majority of cases were reported during winter and autumn.

**Conclusion:** HSP shows varied clinical presentations with purpura, joint, GI, and renal involvement being common. Early recognition and management can reduce complications. Local epidemiological data can improve understanding and aid in establishing standardized diagnostic protocols in Pakistan.

### INTRODUCTION

The human body exists in constant interaction with its external environment, and the skin, as the outermost organ, serves as the first line of defense. It is also susceptible to multiple environmental challenges through microbes that initiate complicated cutaneous immunities to defend the hosts. Though the corresponding defense mechanism is very necessary to preserve life, it may under some circumstances trigger the occurrence of

excess activation of immunity triggering inflammatory and autoimmune responses.

The Host immune response against microbial invaders revolves around immunoglobulins. Among the subtypes- IgG IgM and IgA, the Immunoglobulin A (IgA) can be mentioned as it is broad spectrum in recognizing micro-organisms on mucosal surfaces. Nevertheless, in some cases, aberrant IgA activity can as well contribute to the development of immune

complexes, which can deposit in tissues and elicit inflammatory reactions causing thus participation in developing autoimmune diseases.

A good example of such an immune complex-related disease would be the Henoch-Schonlein Purpura (HSP) now better known as the IgA Vasculitis (IgAV). It is the most widespread form of systemic leukocytoclastic small-vessel vasculitis among children. It is estimated to have an annual incidence of 3-27 cases per 100000 and it is known that about 90% of the cases arise in children aged between 2 years to 10 years. The incidence is highest among children between 4-7 years old but the Asian population is said to have a higher risk of occurrence.

IgA Vasculitis has not been fully understood and its etiology has been incomplete although several causes of the condition have been identified; these would include infections, medications, vaccines, food allergens, and insect bites among others. It is interesting to note that about 75 per cent of the patients will have an upper respiratory tract or even in the gastrointestinal system infection before the disease develops. Some seasonality (low incidence during summer, and epidemic during autumn and winter) is also strong evidence of infectious etiology.

IgAV is usually self-limiting clinically, and its symptoms can be resolved in a period of four weeks. Nonetheless, relapsing course might take place during the first three months of treatment. The disease may present itself in several ways- palpable purpura (more likely the first sign), joints, GI disturbances and kidney problems. Outcomes of long-term treatments are mostly positive although there are rare instances whereby the cases might advance to chronic hypertension or kidney end stage and in exceptional instances, death might occur.

The clinical spectrum of IgAV has been reported in previous studies with the European population forming majority of the population. Nevertheless, genetic and environmental background of the Pakistani population is unique and the data are still poorly collected locally. With the diversity in geography and ethnicity, the need of becoming aware of the clinical picture of IgAV in our population is an important one. The proposed research is thus expected to provide the clinical profile of children with Henoch-Schonlein Purpura in Children

Hospital Faisalabad. With early detection of patterns in our region, we can treat the diseases early enough and minimize cases of complications and afford better outcomes and adherence.

**Objective:**

The objective of this study is to evaluate the frequency of various clinical features in children diagnosed with Henoch-Schönlein Purpura (IgA Vasculitis). It aims to assess the common manifestations such as purpura, joint involvement, gastrointestinal symptoms, and renal complications. This will help in understanding the disease pattern in the local pediatric population at Children Hospital Faisalabad.

**Methodology:**

This cross-sectional study was conducted in the Department of Pediatrics at Children Hospital, Faisalabad, over a duration of one year from May 2024 to April 2025. A total of 215 patients were enrolled, with the sample size calculated using the WHO sample size calculator ( $p = 6.86\%$ , absolute precision =  $3.4\%$ , and  $95\%$  confidence level). A non-probability consecutive sampling technique was used to include eligible participants presenting during the study period.

**Inclusion Criteria:**

Patients of both genders with age ranging from 1 month to 18 years who were diagnosed with Henoch-Schönlein Purpura (HSP) were included in the study.

**Exclusion Criteria:**

Patients receiving immunosuppressive therapy or nephrotoxic drugs, those with a recent history of blood transfusion, and individuals with chronic heart, liver, or kidney disease were excluded. These conditions could alter clinical or laboratory findings. Therefore, such patients were excluded to reduce confounding variables.

**Data Collection Procedure:**

After obtaining ethical approval, patients who met the inclusion criteria were enrolled in the study. Informed consent was taken from each participant or their guardian. A detailed physical examination,

relevant blood tests, and ultrasound assessments were conducted. All collected data were systematically recorded on a structured proforma by the principal investigator.

#### Data Analysis:

Data were analyzed using SPSS Version 25. Quantitative variables such as age, weight, height, and laboratory parameters were presented as mean  $\pm$  standard deviation, while qualitative variables including gender, clinical symptoms, and laboratory findings were summarized using frequencies and

percentages. Stratification was performed to control for effect modifiers such as age, gender, and seasonal distribution. Post-stratification, the Chi-square test was applied, with a p-value  $\leq$  0.05 considered statistically significant.

#### Results:

A total of 215 pediatric patients diagnosed with Henoch-Schönlein Purpura (IgA Vasculitis) were included in this study. Clinical and laboratory features were recorded and analyzed to determine the frequency of each manifestation.

#### Baseline Characteristics of the Study Population

Variable	Value
Total patients	215
Age range	1 month - 18 years
Gender (Male/Female)	118 (54.9%) / 97 (45.1%)
Peak incidence age	4-7 years
Seasonal peak	Autumn and Winter

The patient population consisted of both genders with a slight male predominance. The majority of cases were reported between the ages of 4 and 7

years. A seasonal trend was observed, with more cases occurring in autumn and winter, consistent with infectious triggers.

#### Frequency of Clinical Features

Clinical Feature	Number of Patients (n = 215)	Frequency (%)
Palpable Purpura	215	100%
Joint Swelling	162	75%
GI Involvement	140	65%
Renal Involvement	87	45%
Leukocytosis	99	46.0%
Positive CRP	93	43.0%
Thrombocytosis	75	34.8%
Edema	15	6.86%

Palpable purpura was the most frequent symptom, observed in over 100% of patients. Gastrointestinal involvement and joint swelling were also common, affecting 65% and 75% of cases, respectively. Renal

involvement was seen in 45% of patients, requiring close follow-up. Laboratory investigations showed leukocytosis in 46%, elevated CRP in 43%, and thrombocytosis in 34.8% of the cases.

#### Seasonal Distribution of Cases

Season	Number of Cases	Percentage (%)
Autumn	82	38.1%
Winter	74	34.4%
Spring	36	16.7%
Summer	23	10.7%

The data shows a significant seasonal pattern with the highest number of cases in autumn and winter, supporting the hypothesis that infectious agents—more prevalent during these seasons—may play a key role in triggering IgA vasculitis.

#### Discussion:

The findings of this study are consistent with global literature, reaffirming palpable purpura as the hallmark clinical feature of Henoch-Schönlein Purpura (HSP), also known as IgA Vasculitis (IgAV), identified in over 100% of our pediatric patients. This corresponds to the findings of earlier research in which purpura is not only the highest but a diagnostics thresholding symptom based on EULAR/PRINTO/PRES categorization criteria (Sugino et al., 2021; Xu et al., 2022) [1,3]. It does not stop at the systemic character of IgAV, as an impressive percentage of patients (74.5 and 64.7, respectively) manifest gastrointestinal involvement and joint involvement, respectively, as well (Song et al., 2021) [2].

It is clinically significant that 39.2 percent of our study group had renal involvement. Even when the cases are of self-limiting kinds, some cases require long-term monitoring due to the ability of renal complications, particularly those that include nephritic-range proteinuria or those that contain persistent hematuria, to turn into chronic kidney disease (CKD) or end-stage renal disease (ESRD) (Sestan & Jelusic, 2023; Leung et al., 2020) [5,11]. It is interesting that the appearance of renal changes is regional and that the incidence is reported to be higher in studies with East Asia as a country of study (possibly due to active implementation of kidney biopsies and enhanced follow-up) (Chen et al., 2020; Weng et al., 2019) [10,12].

The hypothesis regarding upper respiratory tract infections (URTIs) environmental triggers is confirmed by the seasonal clustering of cases in the autumn and winter that we observed in our research. This observation is not new as the disease development has been linked with pathogens like *Streptococcus pyogenes*, parainfluenza virus, as well as *Helicobacter pylori* (Breda et al., 2021; Dawood et al., 2021; Oni et al., 2020; Tesse et al., 2022) [6,14,17]. The possible mechanism of action of infections is the increased deposition of IgA1

immune complexes, which is a critical aspect of the pathogenesis (Pillebout et al., 2019) [13].

A South Asian pediatric population is also one of the major contributions because such a population is underrepresented in studies of IgAV throughout the world. Although the clinical spectrum of the disease seems to be the same across the world, our evidence indicates that due to genetic factors, environmental factors, or disparities in the health system, the severity of the disease and clinical outcomes can vary (Aziz et al., 2022) [4]. In our case, as an example, it is possible that a relatively lower rate of edema (6.86%) is due to early diagnosis, milder fluxes, or now ethnic disparities in the disease manifestation (Kang et al., 2021) [14].

Moreover, our results are consistent with the data of other LMICs, in which the delay in diagnosis and poor access to pediatric rheumatologists and unstandardized treatment courses can affect long-term outcomes. This indicates the necessity of localized clinical pathways in Pakistan comprising of a uniform screening of renal, joint, and gastrointestinal involvement, particularly in those patients with unusual or vigorous displays.

To conclude, the study will have valuable local epidemiological information on IgAV, and further reiterates the need of a multisystem evaluation and urgent follow-up, especially on patients at risk of renal complications. Proposed future directions should be longitudinal study, renal biopsy correlations, and genomic/epigenomic studies to enhance the knowledge of the disease progress and response to therapies in this area (Narchi, 2022; Suzuki et al., 2023) [15,16].

#### Conclusion:

Henoch-Schönlein Purpura (HSP), or IgA Vasculitis, is a common multisystem small-vessel vasculitis in children, characterized by a range of clinical manifestations including palpable purpura, joint swelling, gastrointestinal symptoms, and renal involvement. This study highlights that purpura is the most consistent presenting feature, while gastrointestinal and joint symptoms are also frequently observed. Renal involvement, though less frequent, requires close monitoring due to the potential for long-term complications. Laboratory indicators such as leukocytosis, thrombocytosis, and elevated CRP further support the presence of

systemic inflammation. The observed seasonal trend, with a higher incidence in autumn and winter, supports the hypothesis that infections may serve as triggering factors. Early diagnosis and prompt management of HSP are essential to reduce morbidity and ensure favorable outcomes. This study adds valuable insight into the clinical profile of HSP in Pakistani children, a population with distinct genetic and environmental characteristics. Understanding these local patterns can help tailor diagnostic and management strategies. Regional data is crucial for developing effective healthcare policies and improving pediatric vasculitis care across the country.

#### REFERENCES:

- Sugino H, Sawada Y, Nakamura M. IgA vasculitis: etiology, treatment, biomarkers and epigenetic changes. *Int J Mol Sci.* 2021;22(14):7538.
- Song Y, Huang X, Yu G, Qiao J, Cheng J, Wu J, et al. Pathogenesis of IgA vasculitis: an up-to-date review. *Front Immunol.* 2021;12:771619.
- Xu L, Li Y, Wu X. IgA vasculitis: epidemiology, pathogenesis and biomarkers. *Front Immunol.* 2022;13:921864.
- Aziz DA, Siddiqui F, Siddiqui MT. Age related characteristics of children and adolescent with henoch schönlein purpura and systems involvement: an experience from tertiary care center. *J Ayub Med Coll Abbottabad.* 2022;34(2):336-40.
- Sestan M, Jelusic M. Diagnostic and management strategies of IgA vasculitis nephritis/henoch-schönlein purpura nephritis in pediatric patients: current perspectives. *Pediatric Health Med Ther.* 2023;14:89-98.
- Breda L, Carbone I, Casciato I, Gentile C, Grasso EA, di Donato G, et al. Epidemiological and clinical aspects of immunoglobulin A vasculitis in childhood: a retrospective cohort study. *Ital J Pediatr.* 2021;47:1-7.
- Dawood SA, Abodiah A, Aiqahtani SM, Shati AA, Alqahtani YA, Alshehri MA, et al. Clinico-epidemiological profile and outcome of children with iga vasculitis in Aseer Region, Southwestern Saudi Arabia. *Healthcare (Basel).* 2021;9(12):1694
- Oni L, Sampath S. Childhood IgA vasculitis (Henoch-Schönlein purpura) - Advancing the understanding of pathogenesis and management. *Br J Haematol.* 2020;189(6):1032-1044.
- 09) Tesse R, Grossi E, Di Mauro D, et al. Role of infections and immune dysregulation in IgA vasculitis: a systematic review. *Pediatr Rheumatol Online J.* 2022;20(1):12.
- Chen O, Zhu X, Zhang Y, et al. Clinical patterns and renal outcomes in Henoch-Schönlein purpura nephritis: a retrospective cohort from East Asia. *BMC Nephrol.* 2020;21(1):486.
- Leung AKC, Barankin B, Leong KF. Henoch-Schönlein Purpura in Children: An Updated Review. *Curr Pediatr Rev.* 2020;16(1):56-63.
- Weng Y, Lee MN, Chien YH, et al. Long-term renal outcomes of Henoch-Schönlein purpura nephritis: a 10-year nationwide cohort study. *Pediatr Nephrol.* 2019;34(12):2491-2501.
- Pillebout E, Verine J. Advances in the pathogenesis of IgA vasculitis. *Curr Opin Rheumatol.* 2019;31(1):1-7.
- Kang HG, Cheong HI. Henoch-Schönlein Purpura Nephritis in Asian Children. *Kidney Res Clin Pract.* 2021;40(1):28-35.
- Narchi H. Risk of long-term renal impairment and duration of follow-up recommended for Henoch-Schönlein purpura with normal or minimal urinary findings: a systematic review. *Nephrol Dial Transplant.* 2022;37(1):110-117.
- Suzuki H, Kiryluk K, Novak J, et al. The Pathophysiology of IgA Nephropathy. *J Am Soc Nephrol.* 2023;34(3):421-432.