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# Epidemiological Traits and Seasonal Variation of Henoch-Schonlein Purpura (HSP) Iin Children

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## Abstract

***Background:*** *HSP is typically a disease of children between the ages of 3 and 10 years. Although adult cases have been described, 50% of all cases occur at or before the age of 5 years. Males are affected twice as often as females. In North America, Caucasians have the highest prevalence, and African Americans have the lowest prevalence. Although the cause of HSP is unknown, it commonly follows an upper respiratory tract infection. As a result, the disease is more common in winters.*

***Objectives:***

1. *To determine the prevalence of Henoch-Schonlein purpura (HSP) in children.*
2. *To determine the seasonal variation of Henoch-Schonlein purpura (HSP) in children.*

***Materials and Methods:*** *This was a cross sectional study conducted at Jinnah Postgraduate Medical Center & Karachi Medical & Dental College. Children (<17 yr of age) diagnosed with HSP were included on the basis of non-probability convenient sampling. The diagnosis of HSP was based on the standard defined by the American College of Rheumatology. The sample size calculation was done using the World Health Organization, Geneva software where α=5%, 1-Beta=80, Po=0.15, Pa=0.20, sample size=342. Continuous variables like age and number of patients admitted at various time of year were presented as mean ± standard deviation. Categorical variables, gender and age (<14, >14 years) were presented as proportions.*

***Results:*** *Mean age of children with HSP was 11 ± 1.5 years. Majority of the children 43.6% were <14 years of age in 2017 and 51.2% in 2018. The annual prevalence of HSP from 2017 till 2018 increased to 9.2%. The frequency of hospitalization also increased, 2017: 40%; 2018: 59%. Males were 71% and females were 28%. Male to female ratio was 2.0. Males >10 years were 29% and females >10 years were 70% (p<0.05) Table1. Number of cases increased from 8-12 years of age and were less frequent in <8 years and >14 years of age. Majority, 20% of cases were of 10 years age. HSP occur all year round with a high prevalence in the winter season. Number of patients increased from 2% in June and July to 58% from November till February.*

***Conclusion:*** *The overall prevalence of HSP was 19.5 per 1000 affecting children of 9 to 11 years in winters.*

***Keywords:*** ***Henoch-Schonlein purpura, children, seasonal variation.***

## Background

Henoch-Schönlein purpura (HSP) is a kind of systemic small vessel vasculitis characterized by non-thrombocytopenic palpable purpura, arthritis, bowel angina and haematuria/proteinuria. The real pathogenesis is still unknown, it is well recognized as a specific clinicopathological entity by the vascular deposition of immunoglobulin A (Ig A)-containing immune complexes and elevated serum Ig A levels. This disease is usually self-limiting. Involvement of internal organs such as kidney, intestine and central nervous system is a major complication.[1-3] Although HSP is not uncommon in children, there are few large-scale epidemiological studies of childhood HSP, especially nationwide surveys.[4-6] Like most previous studies that have been limited to hospital-based or regional-based sampling data, the latest survey conducted in the West Midlands, UK has shown the annual frequency of HSP to be around 20.4 per 100 000 children aged <17 yr.[7] In France, the annual occurrence of HSP among children before 15 years of age was reported to be 21.75/100 000 in a multicenter survey between 1992 and 1995.[8] Features of HSP in Thailand were reviewed, and the authors found that of 47 children with HSP, renal involvement was detected 6 months after diagnosis in six of 22 cases of children who ultimately had renal disease.[9] HSP occurs throughout the year, but a number of studies have noted seasonal skewing, with most patients presenting from fall through spring, and a paucity of cases during the summer months.[10-12] Clusters or epidemics of HSP are rare. Farley et al reported a cluster of 16 cases of HSP, including 2 pairs of siblings during a 7-month period in Connecticut.[13] Seasonal distribution variation of HSP in England and Scotland showed that the occurrence of the disease is lowest in the June-August period.[14]

***Rationale:*** The prevalence of HSP is not known in Pakistan. We are also not aware of the seasonal variation of HSP in our country. We plan to determine the annual prevalence and seasonal discrepancy of HSP in Pakistan. Using the data from government hospital the aim of this study is to determine the annual prevalence of Henoch-Schonlein purpura (HSP) in children and to determine

the seasonal variation of Henoch-Schonlein purpura (HSP) in children.

## Materials and Methods

**Study design and study cases**

This was a cross sectional study conducted at Jinnah Postgraduate Medical Center and Karachi Medical & Dental College. Children (<17 yr of age) diagnosed with HSP throughout the study period were included on the basis of non-probability convenient sampling. The diagnosis of HSP confirmed by medical doctors in Karachi, including pediatricians and dermatologists, was based on the standard defined by the American College of Rheumatology (ACR),[15] which require individuals to have at least two of four criteria: age less than or equal to 20 yr at disease onset, palpable purpura without thrombocytopenia, acute abdominal pain or gastrointestinal bleeding and biopsy showing granulocytes in the walls of small arterioles or venules. Physicians were able to distinguish HSP from other forms of vasculitis according to these criteria with a sensitivity of 87.1% and a specificity of 87.7%.[15] The sample size calculation was done using the W.H.O. software for "Sample Size Calculation" edited by L. Lemeshow and S. K. Lwanga, where α=5%, 1-Beta=80, Po=0.15, Pa=0.20, sample size=342. Continuous variables like age and number of patients admitted at various time of year were presented as mean ± standard deviation. Categorical variables, gender and age (<14, >14 years) were presented as proportions.

## Results

Mean age of children with HSP was 11 ± 1.5 years. Majority of the children, 43.6% and 51.2% were <14 years of age respectively. The average annual prevalence of HSP increased to 19.5%. The frequency of hospitalization also increased from 40% to 59%. Males were 71% and females were 28%. Male to female ratio was 2.0. Males >10 years were 29% and females >10 years were 70% (p<0.05) Table1. Number of cases increased from 8-12 years of age and were less frequent in <7 years and >14 years of age. Majority, 20% of cases were of 10 years age, Table 2 and Figure 1. The annual prevalence ranged from 14.9 per 1000 to 24.1 per 1000 in children among <14 years with an average overall prevalence of 19.5 per 1000, Figure 3.

**Table 1: Demographic profile of children with Henoch-Schonlein purpura**

| Variables | Percent |
|---|---|
| **Mean Age** | 11 ± 1.5 years |
| **<14 years 2010** | 43.6% |
| **<14 years 2011** | 51.2% |
| **Annual incidence 2010/1000** | 14.9% |
| **Annual incidence 2011/1000** | 24.1% |
| **Frequency of hospitalization 2010** | 40% |
| **Frequency of hospitalization 2011** | 59% |
| **Males** | 71% |
| **Females** | 28% |
| **Male to female ratio** | 2 |
| **Male >10 years** | 29% |
| **Females >10 years** | 70% |

**Table 2: Age distribution of Henoch-Schonlein purpura**

| Age in years | Percent |
|---|---|
| **7** | 8% |
| **8** | 13% |
| **9** | 17% |
| **10** | 20% |
| **11** | 17% |
| **12** | 13% |
| **13** | 7% |
| **14** | 2% |

**Table 3: Seasonal variation of Henoch-Schonlein purpura**

| Seasons | Percent |
|---|---|
| **January** | 18% |
| **February** | 11% |
| **March** | 8% |
| **April** | 5% |
| **May** | 4% |
| **June** | 2% |
| **July** | 2% |
| **August** | 4% |
| **September** | 5% |
| **October** | 7% |
| **November** | 11% |
| **December** | 18% |

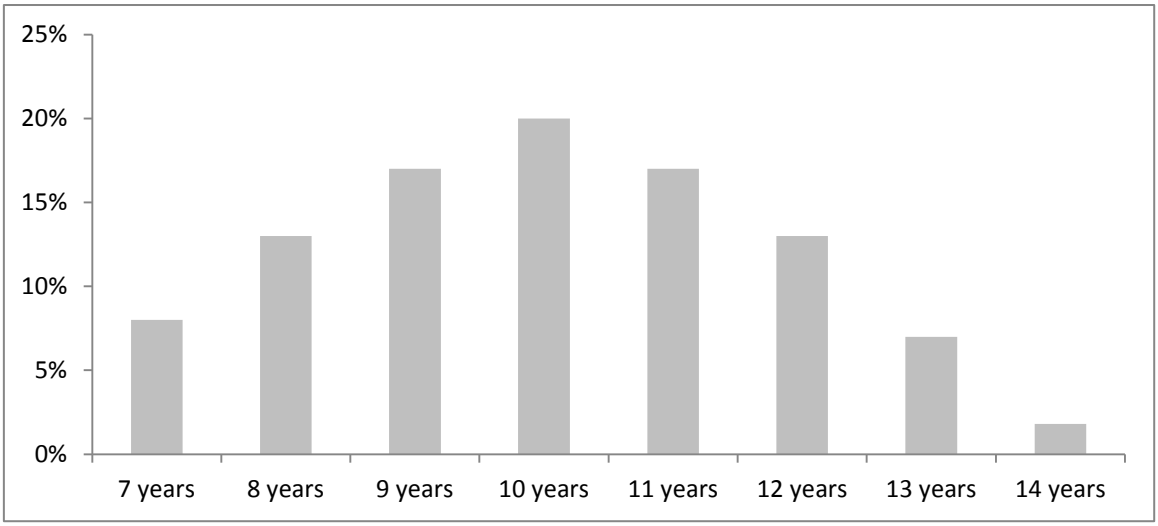


**Figure 1: Age distribution of Henoch-Schonlein purpura**

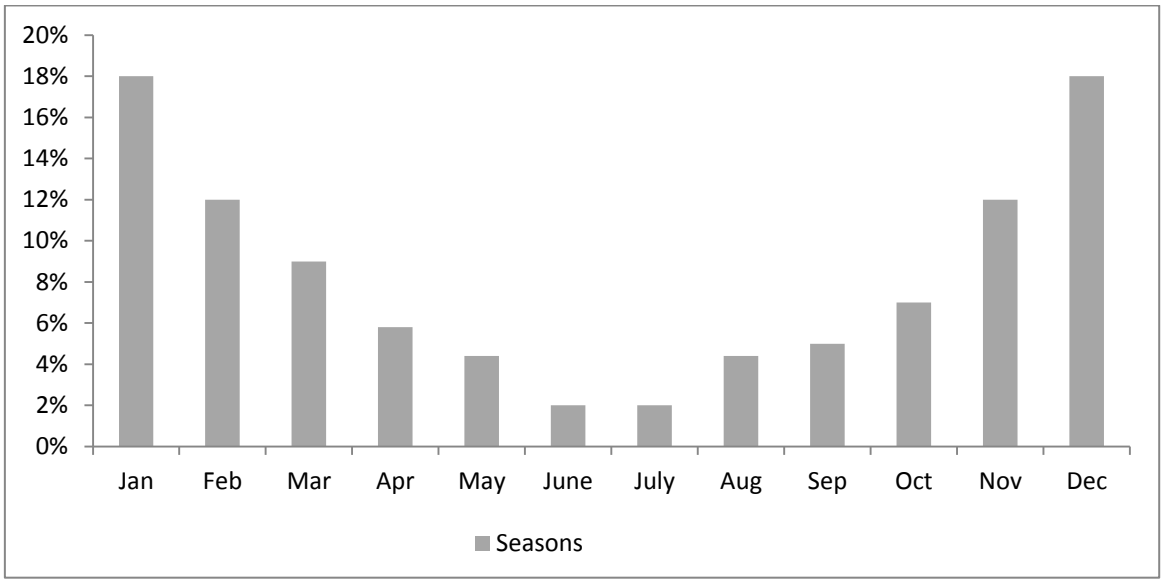


**Figure 2: Seasonal variation of Henoch-Schonlein purpura**

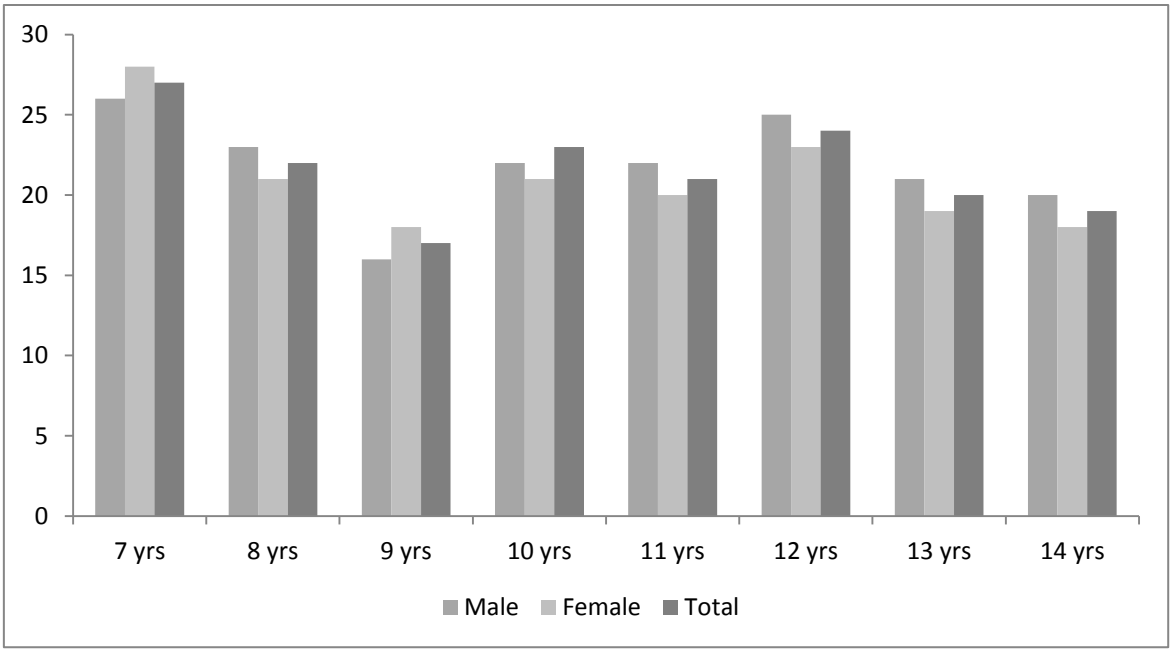


**Figure 3: Annual prevalence (per 1000 children) of Henoch–Schonlein purpura in male, female and total number of children by age**

***seasonal variation:***
HSP occur all year round with a high prevalence in the winter season. Number of patients increased from 2% in June and July to 58% from November till January, Table 3 and Figure 2.

## Discussion

The data was used from hospital records and HSP clinic services, this study provided local population survey on childhood HSP. These results show that overall 49.5% of patients were hospitalized in HSP wards. The annual prevalence of HSP was recently studied by Gardner-Medwin et al.[7] Using questionnaires in one region of the United Kingdom, they found the estimated annual prevalence from 1996 to 1998 to be 20.4 per 100 000 children <17 yr of age. Asian children accounting for 10% of the study population had a higher prevalence (24.0 per 100 000) than White and Black. Among them, most were Indian (53%) followed by Pakistani (33%) and Bangladeshi (7%).[7] In Czech Republic, there is a lower prevalence of 10 per 100,000 in children <17 years of age with a peak prevalence at five to seven years of age.[16,17] The rate of HSP is significantly higher (approximately 5 percent) in patients with familial Mediterranean fever.[18,19] Our findings suggested annual prevalence of HSP in children younger than 14 yr of age (14.9 per 1000 in 2010 and 24.1% per 1000 in 2011.)

The modification of imbursement scheme and easily accessible medical services may influence physicians' performance and might encompass rising hospitalization rate throughout the 2 year study period. An additional prospect is that the number of cases increased progressively. In Karachi, it is recommended that HSP children with vital organ involvement and those who suffer from a more intractable disease course are hospitalized for additional management. Further studies are required to assess the complications of these patients. The male to female ratios of childhood HSP varied among different studies. Most series have described a higher prevalence in males (the ratio ranged from 1.2 to 1.6),[20,21] while some studies have shown a female

predominance.[9,22] These variations may be attributable to the small sample sizes in the majority of previous studies and different time frames, races and geographical areas from which the data were recorded and analysed. In this study, the overall male to female ratio was 2. Our study also revealed an increased frequency of hospitalization in 2017; nevertheless, the duration of hospitalization in males and females was not different. Infection shortly before the onset of vasculitis, which is a predisposing factor for the episode of HSP is, by contrast a protective factor for relapsing disease.[23] Therefore, further study is needed to ascertain whether the disease course in male patients is more complicated and severe than in female patients. Close contact with schoolmates in younger children is suggested to be a significant risk factor for many infectious diseases, especially respiratory infections.[24,25] The age-related epidemiological variations in HSP, together with the disease clustering in autumn and winter and the histories of preceding upper respiratory infections recorded in many HSP patients provide clues to the possibility that HSP is infection-related.[24] There are several limitations in this study. Comprehensive medical reports for every patient are not accessible from the hospital research center. As a result, the medical presentations such as cases with kidney involvement, complications, restorative plans and prognosis in these patients are unknown. Patients might be hospitalized in the similar period, exclusive of medical records; we in addition could not conclude the recurrence and the time between two events of childhood HSP.

## Conclusion

Henoch-Schonlein purpura affects children with a peak prevalence in 9 to 11 years in winters.

**Key messages:**

1. The annual prevalence of childhood HSP in Karachi is 19.5 per 1000 children.
2. The occurrence of HSP had a peak at the age of 9 to 11 years.
3. Disease onset was more common in winters.
4. Evidence-based medicine in the pathogenesis and treatment of HSP remains limited and most treatment remains conservative and experience-based.

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The authors have declared no conflicts of interest.

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