Henoch-Schönlein Purpura in Children: a 17-year Experience

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ABSTRACT : A descriptive study of one hundred and five Henoch-Schönlein purpura (HSP) patients (57 males, 48 females) treated during 1987-2003 in Department of Pediatrics, Faculty of Medicine Siriraj Hospital, was conducted. The male to female ratio was 1.2: 1. The mean age of the patients was 7.1 years (range 2-15). Most patients lived in Bangkok and the central region of Thailand. HSP most commonly occurs in the rainy season. Clinical manifestations were rash (100%), arthralgia (61.9%), arthritis (25.7%), abdominal pain (66.7%), gastrointestinal bleeding (14.3%: stool occult blood 11.4% and hematemesis or melena 2.9%), nephritis (37.1%: microscopic hematuria 29.5%, gross hematuria 7.6%), proteinuria (23.8%), nephrotic syndrome (1%), and hypertension (1%). The most characteristic rash was purpura, mainly on the lower extremities. Arthritis and/or arthralgia commonly affected feet and ankles. The abdominal pain was commonly localized at epigastrium and umbilical area. Gastrointestinal complications included upper gastrointestinal bleeding, appendicitis, duodenal ulcer, gangrenous intussusception, and gastritis. The most common presenting signs and symptoms were rash. Of all patients, 34.3% developed recurrent symptoms including abdominal pain, nephritis, and rash mostly occurring within the first 3 months after the initial resolution (range 2 days-9.6 years).

The mainstay of management was supportive care. The patients with severe abdominal pain received prednisolone. Prednisolone and cyclophosphamide were only given to severe nephritis patient with a good outcome.

Key words : Henoch-Schönlein purpura, children, clinical manifestation

Henoch-Schönlein Purpura (HSP) is primarily a children disease and the most common acute vasculitis affecting children. The dominant clinical manifestations are purpura, abdominal pain, nephritis, arthritis, and gastrointestinal bleeding[1]. Renal involvement is the most severe complication of this disease[2]. Although the exact cause of the vasculitis is unknown, the clinical features of HSP are a consequence of IgA deposition in vessel walls causing leukocytoclastic vasculitis[3]. Epidemiological studies showed annual incidence of HSP in about 13.5-18/100,000 children[4]. Although this disease can occur in children as young as 6 months of age as well as adults, 75% of cases occur in children under 10 years of age and more commonly in boys[5].

In this report, we present epidemiologic data, the clinical features, the rate of recurrence, the timing between recurrence and initial resolution, management, and outcome of 105 Thai children with HSP.

MATERIALS AND METHODS

A retrospective study of 105 patients diagnosed with HSP in the Department of Pediatrics, Faculty of Medicine Siriraj Hospital, from January 1987 through December 2003 was undertaken.

Inclusion criteria

1. HSP is defined as non-thrombocytopenic purpura plus one or more of the following; joint pain and swelling, renal involvement, abdominal pain, and gastrointestinal bleeding.
2. Age at presentation was less than 15 years.

Nephritis was defined as the presence of gross hematuria or microscopic hematuria and having the characteristic of dysmorphic red blood cells, whereas severe nephritis was defined as the presence of acute nephritic syndrome (i.e., hematuria with the following: hypertension, raised plasma urea and creatinine or oliguria) with or without nephrotic syndrome. Joint manifestations were described as well-defined arthralgia or arthritis. Gastrointestinal manifestations were described as the following: bowel angina presented as diffuse abdominal pain, gastrointestinal bleeding presented as melena, hematochezia, or a positive test for occult blood in the stool. A recurrence or relapse was defined as reappearance of the rash or other symptoms following resolution of disease.

Data on the following items were analyzed: age, sex, season of diagnosis, presenting symptoms, clinical manifestations (including location of skin and joint lesions, gastrointestinal and renal manifestations), interval from the onset of symptoms to the development of recurrent symptoms, clinical manifestation of recurrent symptoms, and corticosteroids requirement.

Statistical analysis: Data present in frequency, percent and mean.

RESULTS

Epidemiology

The patient population consisted of 57 boys (54.3%) and 48 girls (45.7%) ranging from 2 to 15 years of age (Figures 1). The mean age was 7.1 years. The male to female ratio was 1.2 to 1. Eighty-three percent of patients were less than 10 years of age. Patients were presented every month of the year. 59, 25, and 21 patients were presented in the rainy, cool and hot seasons, respectively. Most patients lived in Bangkok and the central provinces of Thailand. Eight patients came from north and 2 each came from the south, northeast, and east of Thailand.

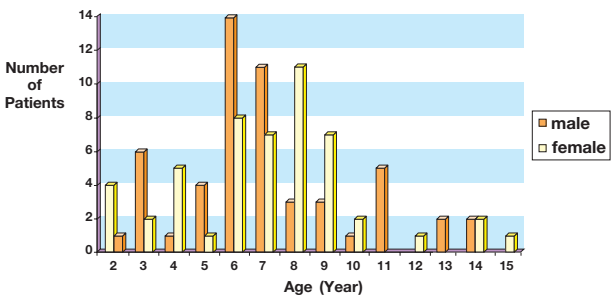


Fig 1. Age and gender distribution of 105 children with Henoch-Schönlein purpura.

Clinical Manifestations

The major clinical manifestations of the 105 patients are shown in Table 1. All patients had skin manifestations mostly described as non-thrombocytopenic palpable purpura except for one patient who had petichiae. Ninety-five (90.5%) patients had skin lesions concentrated on lower extremities only or lower extremities and buttocks. Six (5.7%) patients had lesions on bodies and buttocks in addition to legs and arms. Three (2.9%) patients had skin involvement limited to legs and arms and one patient had only truncal lesion. The distribution of skin manifestation is shown in Table 2.

TABLE 1. Clinical manifestations in 105 children.

Clinical features, signs and symptoms	Number of patients	(%)
Skin lesions	105	(100)
Arthralgia (no arthritis)	65	(61.9)
Arthritis	27	(25.7)
Abdominal pain	70	(66.7)
Gastrointestinal bleeding		
Stool occult bleeding	12	(11.4)
Gross bleeding (hematemesis, melena)	3	(2.9)
Nephritis		
Microscopic hematuria	31	(29.5)
Gross hematuria	8	(7.6)
Significant proteinuria	25	(23.8)
Nephrotic syndrome	1	(0.9)
Orchitis	7	(6.7)
Hypertension	1	(0.9)
Appendicitis	1	(0.9)
Duodenal ulcer	1	(0.9)
Gangrenous intussusception	1	(0.9)
Peritonitis	1	(0.9)

TABLE 2. Distribution of skin involvement in 105 patients.

Location of skin lesions	Number of patients	(%)
Legs	77	(73.3%)
Legs and buttocks	18	(17.1%)
Legs, arms, and trunk	3	(2.9%)
Legs, arms, and buttocks	3	(2.9%)
Legs and arms	3	(2.9%)
Trunk	1	(0.9)

Arthralgia occurred in 65 (61.9%) patients. In 54 of the 65 (83.1%) patients, arthralgia was limited to the lower extremities. Six (9.2%) patients had involvement limited to hands and wrists and five (7.7%) to both wrists and ankles. The distribution of arthralgia is shown in Table 3.

TABLE 3. Distribution of joint involvement in 65 patients with arthralgia

Location of joints	Number of patients	(%)
Feet and ankles	35	(53.8%)
Knees	12	(18.5%)
Knees and ankles	7	(10.8%)
Wrists and hands	6	(9.2%)
Wrists and ankles	5	(7.7%)

Arthritis occurred in 27 (25.7%) patients. In 22 of the 27 (81.5%) patients, arthritis was limited to the lower extremities. Four (14.8%) patients had hands and ankles involvement in addition to wrists and only one (3.7%) patient had elbow involvement. The distribution of arthritis is shown in Table 4.

TABLE 4. Distribution of joint involvement in 27 patients with arthritis

Location of joints	Number of patients	(%)
Feet and ankles	14	(51.9%)
Knees	5	(18.5%)
Knees and ankles	3	(11.1%)
Wrists and hands	2	(7.4%)
Wrists and ankles	2	(7.4%)
Elbow	1	(3.7%)

Gastrointestinal involvement presented as abdominal pain occurred in 70 (66.7%) patients. Fifteen of the 70 (21.4%) had gastrointestinal bleeding. The occult gastrointestinal bleeding was found in 12 (80%) patients and grossly bloody or melanotic stools occurred in 3 (20%) patients. One patient each had confirmed appendicitis, duodenal ulcer, peritonitis, and gangrenous intussusception. The locations of abdominal pain are shown in Table 5.

TABLE 5. Distribution of locations in 70 patients with abdominal pain.

Areas of abdominal pain	Number of patients	(%)
Epigastria	42	(60%)
Umbilicus and peri-umbilicus	13	(18.6%)
Generalized	8	(11.4%)
Right lower quadrant	3	(4.3%)
Left upper quadrant	2	(2.9%)
Left lower quadrant	1	(1.4%)
Mid lower quadrant	1	(1.4%)

Nephritis with dysmorphic red blood cells in the urine occurred in 39 (37.1%) patients. Nephritis was presented as microscopic hematuria in 31 (79.5%) patients and as gross hematuria in the remaining 8 (20.5%) patients. Twenty-six of the 39 (66.7%) patients with nephritis also had significant proteinuria. All patients with proteinuria had concomitant hematuria. One patient had nephrotic syndrome defined as urinary protein excretion greater than 50 mg/kg per 24 hours plus edema, hypercholesterolemia, and hypoalbuminemia. One patient had hypertension together with microscopic hematuria.

Presenting signs and symptoms

Purpura was the chief complaint that brought 73 (69.5%) patients to see physicians. Abdominal pain, arthritis, and arthralgia were the initial manifestations in 18 (17.1%), 9 (8.6%), 5 (4.8%) patients, respectively. These are shown in Figure 2.

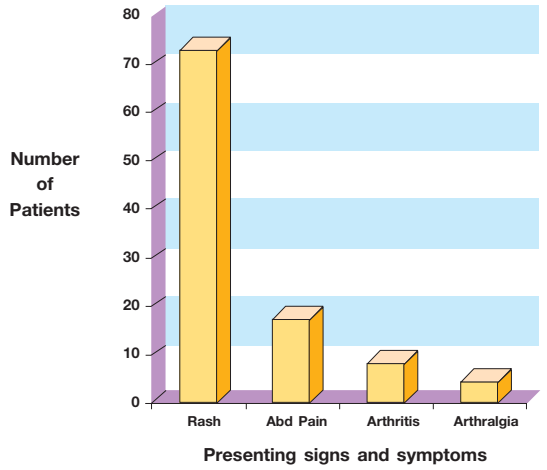


Fig 2. Illustration of chief complaint in 105 children with Henoch-Schönlein purpura

Recurrences

A recurrence was defined as a reappearance of the rash or other symptoms following resolution of the disease. Thirty-six (34.3%) patients had recurrences. The symptoms of recurrences were abdominal pain in 16 (44.4%) patients, nephritis in 12 (33.3%) patients, rash in 3 (8.3%) patients, abdominal pain/nephritis in 3 (8.3%) patients, and abdominal pain/rash in 2 (5.6%) patients. The time between initial resolution and recurrence ranged from 2 to 3,500 days, with a mean of 141 days. However, 33 of 36 (91.7%) patients had recurrences within 13

weeks (3 months) after the initial resolution. These are illustrated in Figure 3.

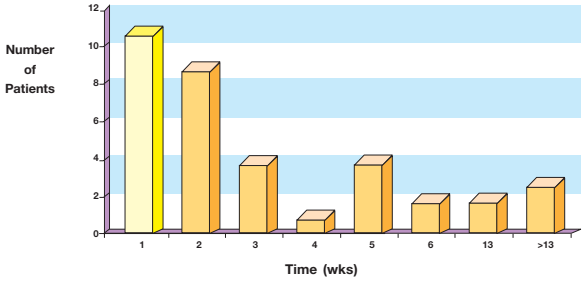


Fig 3. Illustrate duration between initial resolution and recurrence in 36 patients.

Treatment

Sixty-four patients (61%) received prednisolone with a dosage ranging from 1-2 mg/kg/day for 3-7 days for abdominal pain and 7-30 days in patients with nephritis. The average duration of prednisolone treatment was 9.7±2.9 days. There were three severe nephritis patients who received oral cyclophosphamide. None of the patients in this study had renal failure.

DISCUSSION

HSP is the most common vasculitic disease in children. It is a small-sized blood vessel vasculitis characterized by involvement of the skin, joint, gastrointestinal tract, and kidneys. Histopathologic features are infiltration of blood vessels with polymorphonuclear leukocytes called leukocytoclastic angiitis resulting from immunoglobulin A (Ig A)-mediated inflammation.

Although this condition can occur from age 6 months to adulthood, it is predominantly in children. However, it rarely affects children younger than 2 years of age. The mean age at onset is 4 to 5 years[6]. In most reports, HSP is more common in boys with male to female ratio, 1.4-1.7:1[7]. In this study, most patients were in the range from 4 to 10 years of age and the mean age was 7.1 years. The male to female ratio was 1.2 to 1. In Thailand, at King Chulalongkorn Memorial Hospital, most HSP children were in the range 6-9 years of age with an average age of 8.5 years[8]. The male to female ratio was 1 to 1.2. A report on 79 HSP children from our hospital showed an average age of 7.76 years[9]. The male to female ratio was 1 to 1.07. Although the average age in our study did not differ from the previous 2 studies in our country, there were differences in the male to female ratio. Our study found more boys than girls, which followed the pattern in western countries. This could reflect the higher number of patients in our study than previous studies. Our patients mainly developed this disease in the rainy season, which also had been reported at King Chulalongkorn Memorial Hospital[8]. However, HSP patients mainly were reported in the fall and winter in western countries[10].

Skin lesions were present in all identified patients. The lesions usually appeared on the extensor surface of the lower extremities and buttocks but may have involved the upper extremities, trunk, and face. Skin manifestations were extremely variable. The classic lesion begins as a small wheal or erythematous maculopapule. Lesions initially blanch on pressure but later lose this feature and generally become petechial or purpuric. The purpura is often palpable. Purpuric areas evolve in the usual manner of ecchymoses, changing from red to purple, becoming rusty, and eventually fading. Angioedema involving the scalp, eyelids, lips, ears, dorsa of the hands and feet, back, scrotum, and perineum is common and may be striking in young children[11].

Our study found palpable purpura in 104 children (99%) and petechiae in 1 child (1%). Skin lesions were concentrated on the upper or lower extremities and buttocks in over ninety-five patients (90.5%). Skin lesions were also found on the bodies and arms but not on the face. Most patients develop skin lesions prior to other symptoms. Purpura preceded other signs and symptoms in 73 (69.5%) patients which was comparable to the 69.2% reported by Calvino et al[12]. Abdominal pain and arthritis were the initial manifestation in 27 (25.5%) patients, whereas as high as 43% of patients having abdominal pain and arthritis preceded the onset of the purpura by 1-14 days[13]. We also found orchitis in 7 (12.3% of boys) patients.

Joint symptoms were the second most significant clinical manifestation. Ninety-two (87.6%) patients had joint involvement. Most patients had joint symptoms involving the knees, ankles, or feet. Sixty-five (61.9%) and twenty-seven (25.7%) patients had arthralgia and arthritis, respectively. None received joint aspiration. Few patients had synovial fluid analyses in the previous reports and it was unclear whether the joint involvement was true synovitis. Our report found arthritis in 8.6% of patients as a presenting symptom compared to 24%-25% in previous reports[5,13,14]. Although patients with arthritis may have severe pain on ambulation, it is self-limited and non-deforming. But it can recur. Like the purpura, arthralgia and arthritis tend to subside with bed rest and exacerbate with ambulation.

Gastrointestinal involvement occurs in 50%-75% of patients. Abdominal pain is the most common; however, symptoms vary from nausea, vomiting to gastrointestinal bleeding, intussusception, pancreatitis, and hydrops of the gall bladder. Mesenteric adenitis can lead to intussusception in rare patients. In the present report, most patients had gastrointestinal symptoms followed by skin lesions and joint involvement. Eighty-five (81%) patients had gastrointestinal involvement. Seventy (66.7%) patients had abdominal pain, and fifteen (14.3%) had gastrointestinal bleeding. Abdominal pain was presented as the chief complaint in 18 (17.1%) patients. Most abdominal pain was presented as colicky pain. The gastrointestinal bleeding was occult in 12 and gross bleeding as hematemesis or melanotic stools in 3 patients. Gastrointestinal bleeding occurred in the presence of abdominal pain. Three severe abdominal pain patients received surgery, later diagnosed as acute appendicitis, gangrenous intussusception, and peritonitis. Gastrointestinal symptoms result from bowel wall edema and hemorrhaging owing to the vasculitis. Both edema and hemorrhage can happen together. If gastrointestinal symptoms are present prior to skin lesions, patients may be misdiagnosed as surgical cases. It would be difficult to differentiate abdominal pain in HSP from surgical cases. Abdominal pain in HSP may also need surgery as in appendicitis, gastric hemorrhage or intussusception. One patient had a duodenal ulcer diagnosed from a gastroduodenoscope. Abdominal pain was the most common recurrent symptoms found in 21 of 36 patients who had a recurrence of HSP. A study from Rosenblum[15] reported 75% of the abdominal pain HSP patients presented within eight days of the onset of the rash. It occurred within 30 days in 42 of 43 patients, except one patient who developed pain 150 days following the onset of rash. In their report stool occult bleeding, hematemesis and melenotic stools were found to be much more com-

mon than in our report.

The incidence of nephritis varies widely from 30%-90% of patients with HSP[2]. Nephritis is usually mild and not progressive. Nephritis was found more commonly in older than younger children. The true incidence of nephritis may be slightly higher since there was a report of severe hypertension without urinary abnormalities in a patient with HSP[16]. Unlike arthritis or abdominal pain, it is uncommon for nephritis to precede the appearance of the rash. Approximately 80% and 95% of patients manifest nephritis within 4 and 13 weeks, respectively, after the onset of other symptoms. It is uncommon to have nephritis after 3 months even though it has been reported in a few patients[13]. In this report, nephritis was present in 39 (37%) patients. The severity of nephritis was not related to the severity of other symptoms. Fifteen (40.5%) of 37 recurrent HSP patients had nephritis, which occurred within 20 weeks. However, most nephritic patients were asymptomatic except for abnormal urine findings, which were represented as microscopic hematuria. Data on the recurrence of nephritis might be lower than actual data since some patients had a short period of follow-up and microscopic hematuria might not bring patients to our clinic. In the present study, none of the nephritic patients had renal impairment. HSP is generally benign and self-limited in children and more severe in adults. The frequency of severe renal manifestations or renal insufficiency during the course of the disease in children was significantly reduced compared to that of adults[17,18]. In an unselected patient population, renal insufficiency was found in only 1.5% and progression to end-stage renal disease may take several years after onset[19]. Goldstein et al[20] reported a long-term outcome of patients who had had Henoch-Schönlein nephritis during childhood until pregnancy and found that 16 of the 44 (36%) successful pregnancies were complicated by hypertension, persistent proteinuria, or both. However, the majority of women had Henoch-Schönlein nephritis with complete clinical recovery. HSP nephritis also tends to recur in living-related renal grafts in children whose original disease was HSP nephritis. Prognosis of the graft was poorer in patients who had a pathological recurrent graft, hematuria, and proteinuria[21].

We found recurrences of symptoms in 34.3% of patients which was close to the 33% reported by Saulsbury[13] but there were some differences in the definition of recurrences. His study defined recurrences as a reappearance of the rash or other symptoms following the resolution of the disease for at least 2 weeks. Recurrences of symptoms were as low as 14.5% when Calvino et al[12] defined recurrences as a new flare of skin lesions or other systemic complications after diagnosis with HSP and asymptomatic for at least 1 month. In the present report, recurrences of symptoms in HSP were presented as nephritis in 15 patients with Ig A deposition by immunofluorescent. There were 21 recurrences of HSP patients with gastrointestinal symptoms who had abdominal pain alone or together with other symptoms. In this group, 2 patients had gastritis. There were 5 patients with complications. One each had upper gastrointestinal bleeding, duodenal ulcer, gangrenous intussusception, peritonitis, and appendicitis. However, 91.7% of recurrences happened within 3 months after resolution, whereas Saulsbury[13] reported 94% of recurrences within 4 months.

There is no specific treatment for HSP. Symptomatic treatment for joint pain, fever, and malaise with acetaminophen or NSAIDs is generally recommended. We used prednisolone for severe abdominal pain. Abdominal pain patients received prednisolone for 3 to 7 days. Glasier et al[22] reported that abdominal pain was resolved within 3 to 7 days in most patients treated with or without corticosteroid. However, a retrospective review from Rosenblum[15] found that corticosteroid hastens the resolution of abdominal pain. 44% and 65% of corticosteroid-treated patients had a resolution of abdominal pain within 24 and 48 hours, respectively, whereas only 14% and 45% of untreated patients had a resolution at the same period. Data suggest no effect of corticosteroids on the purpura, the duration of illness, recurrences rate, or the prevention of recurrences. The value of corticosteroids in preventing intussusception in HSP remains difficult to assess due to the small number of patients. Mollica et al reported a prospective, randomized, controlled study of corticosteroids in the prevention of HSP nephritis[23]. In contrast, a retrospective study by Saulsbury[24] reported a similar incidence in developing nephritis in HSP patients treated or not treated with corticosteroids. Niaudet's prospective study showed an improvement in outcome in severe nephritis patients treated with intravenous pulsed methylprednisolone followed by oral prednisolone[25]. Foster et al demonstrated that early treatment with prednisolone and azathioprine could prevent the progression of chronic changes and improve outcome[26]. Oner et al[27] also described a good response in rapidly progressive glomerulonephritis HSP patients treated with methylprednisolone, oral cyclophosphamide, dipyridamole, and prednisolone. Tarshish et al[28] reported no differences in outcome in severe HSP nephritis treated with oral cyclophosphamide, 90 mg/m^2/day for 42 days. In the present study, nephritic patients received prednisolone for 7-30 days and 3 patients with severe nephritis received prednisolone and cyclophosphamide with good results.

Due to the nature of this retrospective study the data for presenting signs and symptoms and recurrence might not represent the true data because we relied on recall data from patients and parents, and some patients received treatment from other hospitals prior to being referred to us. (Some information also might be lack due to patients fail to recognize their symptoms).

CONCLUSION

HSP is the most common type of vasculitis in children. The etiology of HSP remains inconclusive. It is clear that IgA plays an important role in the immunopathogenesis as evidenced by IgA deposition in the kidney. In the current report, we present the clinical features of 105 Thai children with HSP. The dominant clinical features are purpura (99%), arthralgia (61.9%), arthritis (25.7%), abdominal pain (66.7%), gastrointestinal bleeding (14.3%), and nephritis (37.1%). About one-third of the patients will have recurrences of symptoms. Abdominal pain was the most common recurrent symptoms. Even though retrospective studies of HSP in children suggests that abdominal pain in this disorder is largely self-limited, treatment with corticosteroids may have a role in accelerating the resolution of abdominal pain. Corticosteroids together with cyclophosphamide may benefit patients with severe nephritis.

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บทคัดย่อ

ประสบการณ์ 17 ปี ในผู้ป่วยเด็ก Henoch-Schönlein Purpura

สุโรจน์ ศุภเวคิน พ.บ.*, เพ็ญจันทร์ ทองพิว พ.บ.*, อนิรุธ ภัทรากาญจน์ พ.บ.*, วิบูล สุนทรพจน์ พ.บ.*, อรุณ วงษ์จิราษฎร์ พ.บ.*, อัจฉรา สัมบุณณานนท์ พ.บ.*

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ศึกษาย้อนหลังในผู้ป่วยเด็กโรค Henoch-Schönlein purpura จำนวน 105 คนที่ได้รับการวินิจฉัย และรักษาที่ภาควิชากุมารเวชศาสตร์ คณะแพทยศาสตร์ศิริราชพยาบาล ในระหว่างปี พ.ศ.2530-2546 เป็นชาย 57 คน หญิง 48 คน อัตราส่วนชายต่อหญิง 1.2 :1 อายุเฉลี่ย 7.1 ปี (พิสัย 2-15 ปี) ผู้ป่วยส่วนใหญ่มาจากกรุงเทพมหานครและจังหวัดในภาคกลางของประเทศ พบผู้ป่วยในช่วงฤดูฝน ลักษณะทางคลินิกพบว่ามีผื่น ร้อยละ 100 ปวดข้อ ร้อยละ 61.9 ข้ออักเสบ ร้อยละ 25.7 ปวดท้อง ร้อยละ 66.7 เลือดออกในทางเดินกระเพาะอาหารและลำไส้ ร้อยละ 14.3 (ตรวจพบเลือดในอุจจาระ ร้อยละ 11.4 และถ่ายเป็นเลือด ร้อยละ 2.9) ไตอักเสบ ร้อยละ 37.1 (ตรวจพบมีเม็ดเลือดแดงในปัสสาวะ ร้อยละ 29.5 และปัสสาวะเป็นเลือดสด ร้อยละ 7.6) มีโปรตีนในปัสสาวะ ร้อยละ 23.8 เป็น nephrotic syndrome ร้อยละ 1 และมีภาวะความดันโลหิตสูง ร้อยละ 1 ลักษณะผื่น ส่วนใหญ่เป็น purpura ที่ขา อาการปวดข้อและข้ออักเสบพบมากที่สุดที่เท้าและข้อเท้า อาการปวดท้องพบบ่อยที่สุดคือบริเวณลิ้นปี่และบริเวณสะดือ ส่วนอาการแทรกซ้อนของทางเดินกระเพาะอาหารและลำไส้ คือ มีเลือดออกในกระเพาะอาหารและลำไส้ส่วนต้น ไส้ติ่งอักเสบ แผลที่ลำไส้เล็กส่วนดูโอดีนัม ลำไส้เน่าจากลำไส้กลืนกันและเยื่อบุกระเพาะอาหารอักเสบ อาการสำคัญที่มาพบแพทย์คือ ผื่นผิวหนัง ผู้ป่วยร้อยละ 34.3 เกิดอาการกลับเป็นซ้ำ ได้แก่ ปวดท้อง ไตอักเสบและผื่น ส่วนใหญ่อาการกลับป็นซ้ำพบในช่วง 3 เดือนแรก การรักษาในผู้ป่วยส่วนใหญ่เป็นการรักษาประคับประคองและมีผู้ป่วยจำนวนหนึ่ง ที่มีอาการปวดท้องอย่างรุนแรงได้รับ prednisolone และผู้ป่วยที่มีอาการไตอักเสบ อย่างรุนแรง ที่ต้องได้รับ ยา prednisolone และ cyclophosphamide ผลการรักษาอยู่ในระดับที่ดี