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Editorial

What We Know about Henoch-Schönlein Purpura in Children up to Date?

Hye Ran Yang

Division of Pediatric Gastroenterology and Hepatology, Department of Pediatrics, Seoul National University Bundang Hospital, Seoul National University College of Medicine, Seongnam, Korea

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Address for Correspondence:
Hye Ran Yang, MD, PhD
Division of Pediatric Gastroenterology and Hepatology, Department of Pediatrics, Seoul National University Bundang Hospital, Seoul National University College of Medicine, 82 Gumi-ro 173-beon-gil, Bundang-gu, Seongnam 13620, Republic of Korea.
E-mail: hryang@snubh.org

ORCID iDs
Hye Ran Yang
https://orcid.org/0000-0002-3423-6922

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Henoch-Schönlein Purpura (HSP) is the most common form of acute vasculitis affecting children, although it has also been reported in adults.[1] The etiology of developing HSP still remains unclear; various antigenic stimuli such as bacterial, viral, or parasitic infections, exposures to some drugs, and vaccinations may cause abnormal immune reactions characterized by the deposition of IgA immune complex in vessel walls and abnormal inflammatory process in genetically susceptible individuals.

As HSP is a systemic leukocytoclastic vasculitis involving small vessels of different organs such as the skin, joint, gastrointestinal (GI) tract, and kidney, clinical manifestations of HSP are diverse according to involved organs.[2] In practice, the diagnosis of HSP is based on clinical features of childhood HSP including non-thrombocytopenic palpable skin purpura, especially on the lower extremities, acute arthritis or arthralgia, diffuse colicky abdominal pain and/or GI bleeding, and renal symptoms of proteinuria or hematuria.

Laboratory tests are basically not so helpful in detecting HSP because even increased inflammatory markers such as leukocytes, platelets, and C-reactive protein may be nonspecific. Recently, some laboratory markers of activated coagulation such as D-dimer and fibrin degradation products were reported to be elevated during acute phase of the disease, reflecting the disease activity of HSP.[3]

HSP per se is a self-limiting disease. Although the prognosis of HSP usually shows a favorable clinical outcome, serious complications requiring hospitalization and surgical treatment such as massive GI bleeding, bowel ischemia and infarction, perforation, and intussusception can occur when the GI tract is involved.[4] Therefore, treatment with steroids has been indicated in HSP patients presenting with serious GI symptoms such as severe abdominal pain and hematochezia. If severe GI symptoms persist despite steroid therapy, additional treatment with intravenous immunoglobulins (IVIG), high dose methylprednisolone pulse therapy, immunosuppressants, or plasma exchange therapy are required consequently.[2] As previously reported, 2 of 10 children treated with only steroid for serious GI involvement underwent surgery for bowel perforation and intussusception, while the other 12 treated with additional IVIG did not.[4] Renal involvement of HSP with persistent

proteinuria and/or hematuria also requires therapeutic strategies, since long-term prognosis of HSP depends on the severity of renal involvement.[1]

Up to date, only limited information has been available regarding the epidemiological characteristics of childhood HSP; this may be one of the reasons for why there are no clinical guidelines or consensuses on the diagnosis and treatment of HSP yet, despite its clinical significance.

In the current issue of *Journal of Korean Medical Science*, Shim et al.[5] have reported the nationwide population-based epidemiological data on the clinical characteristics of HSP in Korean children for the first time. According to this study, the annual incidence of HSP was 55.9 per 100,000 children which was much higher in Korea than 6.1–30.0 per 100,000 children of those previously reported in other countries including Taiwan.[5] The peak age of HSP was 5 years, mainly involving young children, and HSP showed seasonal variation with the higher incidence in spring and winter, commonly accompanied by preceding upper respiratory infections, as reported previously.[1,5] Younger children showed more severe disease courses, and steroid therapy was more frequently used in complicated cases.[5] Because previous reports are mostly based on single center studies or multicenter surveys at most, this nationwide study by Shim et al.[5] provides valuable data on childhood HSP.

However, since childhood HSP differs from adult HSP both in clinical manifestations and outcomes, further nationwide surveys involving both children and adults are required in the future. Furthermore, development of practice guidelines for the diagnosis and treatment of childhood HSP based on nationwide data may be necessary for better practice because children with HSP should be approached differently from adult patients.

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