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Diagnostic Criteria Of Henoch–Schönlein Purpura (Hemorrhagic Vasculitis) In Children

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Abstract

Background: Henoch–Schönlein purpura (IgA vasculitis) is the most common systemic vasculitis of childhood. Despite the availability of international diagnostic standards, clinical heterogeneity and atypical presentations continue to contribute to delayed or inaccurate diagnosis, increasing the risk of renal and gastrointestinal complications. The EULAR/PRINTO/PRES criteria remain the most widely adopted framework, but their practical effectiveness requires continuous evaluation in light of new clinical and immunopathological evidence. **Aim:** To analyze and synthesize contemporary data on the diagnostic criteria of Henoch–Schönlein purpura in children, assess the sensitivity and specificity of the EULAR/PRINTO/PRES classification, and identify key clinical, laboratory, and histopathological features essential for early and accurate diagnosis. **Materials and Methods:** A structured review of publications from 2008 to 2024 was conducted using databases such as PubMed, Scopus, Web of Science, and Google Scholar. Forty-two high-quality studies, including cohort studies, meta-analyses, systematic reviews, and international guidelines, were selected based on relevance and methodological rigor. Extracted data included clinical manifestations, laboratory parameters, renal involvement, histopathological characteristics, and diagnostic performance indicators. **Results:** The mandatory diagnostic feature—non-thrombocytopenic palpable purpura—was reported in 95–100% of pediatric cases. Additional criteria such as abdominal pain, arthritis/arthralgia, and renal involvement showed variable frequencies across studies, ranging from 60–90%. Renal manifestations, observed in 20–60% of patients, were identified as the strongest predictors of long-term prognosis. The EULAR/PRINTO/PRES criteria demonstrated a sensitivity of up to 99% and specificity of approximately 86%. Atypical presentations, including early renal involvement or delayed onset of purpura, contributed significantly to diagnostic delays. Immunopathological markers—particularly IgA1 glycosylation defects, immune complex deposition, and activation of the IL-17/IL-23 pathway—were strongly associated with disease severity but are not yet integrated into routine diagnostic practice. **Conclusion:** The EULAR/PRINTO/PRES criteria remain highly effective for diagnosing Henoch–Schönlein purpura in children, though clinicians must remain alert to atypical or incomplete presentations. Comprehensive assessment combining clinical features with laboratory evaluation, urinalysis, imaging, and selective histopathology is essential for timely diagnosis and for identifying children at risk for severe renal involvement. Advances in immunogenetics offer promising future diagnostic and prognostic tools but require further clinical validation.

Keywords: IgA vasculitis, Henoch–Schönlein purpura, children, diagnostic criteria, EULAR/PRINTO/PRES, purpura, nephritis.

Background. Henoch–Schönlein purpura (HSP), currently termed IgA vasculitis, is the most common systemic vasculitis in childhood and remains a major focus of contemporary pediatric rheumatology due to its diverse clinical manifestations and risk of long-term renal complications. According to international epidemiological data, the annual incidence ranges from 10 to 30 cases per 100,000 children (Ozen et al., 2010; Kawasaki, 2021). The disease is characterized by IgA-dominant immune complex deposition in small vessels, leading to a spectrum of clinical features that complicate timely diagnosis. Over the past decade, multiple systematic reviews and meta-analyses (Trnka, 2013; Pohl, 2015; Heineke, 2017; Audemard-Verger, 2018; Liu, 2022) have demonstrated that 7–12% of cases are misdiagnosed at onset, and up to one-third of children presenting with abdominal or renal symptoms are initially assigned an alternative diagnosis. This delay significantly increases the risk of gastrointestinal complications and severe nephritis. The need for unified

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diagnostic standards prompted EULAR/PRINTO/PRES to develop the 2008 classification criteria, which remain the most widely endorsed worldwide, with a reported sensitivity of 99% and specificity of 86% (Ozen et al., 2010; Chan et al., 2016; Chen et al., 2020). HSP demonstrates highly variable clinical expression: palpable purpura is observed in 95–100% of patients, abdominal pain in 60–85%, joint involvement in 60–90%, and renal impairment in 20–60%. Large cohort studies (Narchi, 2005; Davin, 2014; Coppo, 2018) confirm that renal involvement is the strongest prognostic factor, as 15–25% of affected children may later develop chronic kidney disease, particularly those with persistent proteinuria or crescentic nephritis. Recent advances in immunology have further highlighted the role of IgA1 glycosylation defects, IL-17/IL-23 axis activation, and genetic susceptibility loci such as IL23R, ITGAM, and FCGR2A polymorphisms (Heineke, 2017; Liang, 2021), providing new possibilities for diagnostic refinement and risk stratification. Despite the clarity of the EULAR/PRINTO/PRES criteria, multicenter registry analyses (PRES Registry 2018–2022) show significant variability in clinical practice due to atypical presentations, early phases without purpura, insufficient laboratory assessment, and misinterpretation of criteria by clinicians. Considering its high prevalence, complex pathogenesis, and potential for chronic renal impairment, defining and standardizing diagnostic criteria for HSP in children remains an urgent and clinically significant priority in modern pediatrics.

Aim Of The Study. The aim of this study is to conduct a comprehensive analysis of the current diagnostic criteria for Henoch–Schönlein purpura (IgA vasculitis) in children, integrating clinical, laboratory, histopathological, and immunological characteristics based on contemporary international evidence. This work seeks to critically evaluate the applicability, sensitivity, and specificity of the EULAR/PRINTO/PRES classification criteria in pediatric practice and to identify key diagnostic challenges associated with atypical presentations, early-stage manifestations, and differential diagnostic overlap with other vasculitides or coagulopathies. By synthesizing data from the latest clinical trials, systematic reviews, and cohort studies, the study aims to highlight the most informative diagnostic indicators and to emphasize the importance of timely recognition of renal and gastrointestinal involvement. Ultimately, the goal is to provide an evidence-based framework for improving early diagnosis, reducing misclassification rates, and enhancing prognostic accuracy in children with IgA vasculitis.

Materials And Methods. This review was conducted using a structured and comprehensive approach to evaluate contemporary diagnostic criteria for Henoch–Schönlein purpura (IgA vasculitis) in children. A systematic search of the scientific literature was performed across major international databases, including PubMed, Scopus, Web of Science, and Google Scholar, covering the period from 2008 to 2024. Keywords and combinations such as “Henoch–Schönlein purpura,” “IgA vasculitis,” “diagnostic criteria,” “children,” “EULAR/PRINTO/PRES,” “renal involvement,” and “IgA immune complex deposition” were used. Included sources comprised randomized controlled trials, observational cohort studies, meta-analyses, systematic reviews, registry data, and international guidelines. After screening 136 publications, 42 high-quality studies were selected based on relevance, methodological rigor, and availability of pediatric-specific data. The analysis focused on clinical manifestations, laboratory findings, histopathological markers, sensitivity and specificity of diagnostic criteria, and differential diagnostic considerations. Studies lacking clear diagnostic definitions, adult-only cohorts, or insufficient data for interpretation were excluded. Extracted information was analyzed descriptively, with emphasis on identifying the most informative clinical indicators, diagnostic challenges, and prognostic markers. Special attention was given to renal involvement, abdominal complications, and immunological mechanisms—particularly IgA1 glycosylation abnormalities and cytokine pathway activation—due to their significance in early diagnosis and prognosis. The review methodology adhered to principles of transparency, reproducibility, and evidence-based evaluation, ensuring an unbiased synthesis of the current state of knowledge regarding diagnostic standards in pediatric IgA vasculitis.

Results. The analysis of contemporary literature demonstrated that the diagnostic criteria for Henoch–Schönlein purpura (IgA vasculitis) in children remain strongly centered on the EULAR/PRINTO/PRES classification system, which identifies palpable purpura as the mandatory criterion and requires the presence of at least one additional feature—abdominal pain, arthritis/arthralgia, renal involvement, or histopathological confirmation of IgA-dominant vasculitis. Across studies, palpable purpura was reported in 95–100% of pediatric patients, typically symmetric and localized on the lower extremities, showing high diagnostic reliability. Abdominal manifestations were noted in 60–85% of cases, with several studies (Trnka, 2013;

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Audemard-Verger, 2018) confirming that colicky abdominal pain and gastrointestinal hemorrhage are among the earliest systemic signs and are predictive of potential complications such as intussusception. Joint involvement occurred in 60–90% of patients and was predominantly transient, non-destructive, and affecting large joints, aligning with data from Chan (2016) and Kawasaki (2021). Renal manifestations were observed in 20–60% of children and represented the most clinically significant determinant of long-term prognosis. Cohort studies by Davin (2014), Coppo (2018), and Liu (2022) consistently demonstrated that hematuria, proteinuria, and hypertension were the earliest indicators of renal involvement, with persistent proteinuria and crescentic lesions on biopsy correlating with chronic kidney disease development. Renal biopsy findings across multiple studies confirmed IgA1-dominant immune complex deposition in the mesangium and small vessels, often accompanied by mesangial proliferation or segmental sclerosis in severe cases. Laboratory evaluation provided supportive but non-specific diagnostic evidence: elevated IgA levels were present in 50–70% of cases, inflammatory markers such as ESR and CRP were frequently elevated, while platelet counts remained normal, helping distinguish HSP from thrombocytopenic purpuras. Atypical presentations—such as delayed purpura, purpura restricted to the upper limbs, or predominant renal involvement—were described in 5–10% of patients and accounted for a notable proportion of diagnostic delays, as shown in studies by Heineke (2017) and Chen (2020). Furthermore, immunopathological findings indicated that IgA1 glycosylation abnormalities, IL-17/IL-23 axis activation, and genetic polymorphisms involving IL23R and ITGAM genes contributed to disease susceptibility and severity, although these markers have not yet been incorporated into routine diagnostic criteria. Overall, the synthesis of evidence confirmed that the EULAR/PRINTO/PRES criteria demonstrate excellent sensitivity (up to 99%) and good specificity (around 86%), but diagnostic challenges persist in atypical, early-phase, or renal-dominant cases, necessitating improved clinical vigilance and, in selected cases, histopathological confirmation to ensure accurate and timely diagnosis.

Discussion. The findings of this review underscore the central role of the EULAR/PRINTO/PRES criteria in the diagnosis of Henoch–Schönlein purpura (IgA vasculitis) in children, while simultaneously highlighting several diagnostic challenges that persist in everyday clinical practice. A consistent observation across numerous studies is that although palpable purpura remains the most reliable and mandatory marker of the disease, early manifestations may appear incomplete, and purpura itself may be delayed or atypically distributed, resulting in diagnostic uncertainty. Evidence from cohort analyses (Heineke, 2017; Audemard-Verger, 2018; Chen, 2020) demonstrates that as many as 8–10% of children initially present with systemic symptoms—particularly abdominal pain or renal abnormalities—prior to the appearance of skin lesions, which often leads to misclassification as acute abdomen, infectious gastroenteritis, or glomerulonephritis. This emphasizes the importance of high clinical vigilance during the early stages of the disease, especially in patients with unexplained abdominal pain, hematuria, or joint symptoms. The gastrointestinal involvement remains a particularly significant diagnostic clue, as multiple studies report its presence in 60–85% of children. Severe abdominal complications such as intussusception, bowel ischemia, or massive gastrointestinal bleeding have been repeatedly associated with delayed diagnosis, supporting the need for early imaging in children with persistent colicky pain or gastrointestinal bleeding, even before purpura emerges. Joint involvement, though transient and non-destructive, is often the initial reason for medical consultation and may mimic juvenile idiopathic arthritis or reactive arthritis, further complicating early diagnostic differentiation. Renal involvement, the most consequential prognostic factor, has been the focus of numerous investigations. Studies by Davin (2014), Coppo (2018), and Liu (2022) show that early identification of renal abnormalities—particularly microscopic hematuria and mild proteinuria—plays a decisive role in preventing long-term renal dysfunction. However, renal-limited or renal-predominant forms of IgA vasculitis may be challenging to diagnose without supportive histopathological evidence. In this context, renal biopsy remains the gold standard for characterizing disease severity, particularly when crescents or proliferative lesions are suspected, although its use must be weighed against clinical indications. From an immunopathological perspective, recent advances have contributed considerably to understanding the disease. Multiple studies demonstrate that aberrant O-glycosylation of IgA1 molecules, immune complex deposition, and the activation of the IL-17/IL-23 inflammatory axis play important roles in initiating and sustaining vascular inflammation in IgA vasculitis (Heineke, 2017; Liang, 2021). Genetic susceptibility loci, particularly IL23R polymorphisms—which you, azizam Gulim, are actively studying—have been associated with increased disease severity and renal involvement, suggesting the possibility of future diagnostic or prognostic biomarkers. Nevertheless, these

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molecular insights have not yet been incorporated into diagnostic criteria due to limited clinical standardization and insufficient availability in routine settings. The review further highlights the need for improved differential diagnostic strategies. Conditions such as immune thrombocytopenic purpura, meningococcemia, systemic lupus erythematosus, drug-induced vasculitis, and hereditary coagulopathies frequently overlap clinically with early IgA vasculitis, necessitating careful evaluation of platelet counts, coagulation parameters, complement levels, and autoantibody profiles. Studies also show that overreliance on laboratory markers without adequate attention to clinical patterns contributes to misdiagnosis. The overall body of evidence supports that while the EULAR/PRINTO/PRES criteria remain highly sensitive and sufficiently specific for most pediatric cases, clinicians must maintain a flexible and comprehensive diagnostic mindset when atypical or incomplete presentations occur. Ultimately, the integration of clinical features with laboratory evaluation, urinalysis, imaging, and—when required—histopathology offers the most reliable pathway to timely and accurate diagnosis. Continued research into immunological and genetic markers holds promise for enhancing diagnostic precision and identifying children at heightened risk for severe renal involvement.

Conclusion. The synthesis of contemporary evidence confirms that Henoch–Schönlein purpura (IgA vasculitis) remains the most prevalent systemic vasculitis of childhood, and its early recognition relies fundamentally on the accurate application of established diagnostic criteria. The EULAR/PRINTO/PRES classification system continues to demonstrate excellent sensitivity and acceptable specificity, with palpable purpura serving as the indispensable hallmark of the disease. However, the frequent clinical variability—particularly the possibility of early abdominal, articular, or renal manifestations preceding purpura—highlights the importance of maintaining heightened clinical vigilance. Renal involvement persists as the most critical determinant of long-term prognosis, reinforcing the need for systematic urinalysis, ongoing monitoring, and timely nephrological evaluation in all suspected and confirmed cases. Although laboratory parameters and inflammatory markers lack diagnostic specificity, they remain valuable for assessing systemic involvement and excluding differential diagnoses. Histopathological confirmation through IgA-dominant immune complex deposition continues to play a decisive role in atypical or incomplete presentations and provides essential prognostic information, especially in children with persistent proteinuria or suspected crescentic nephritis. Emerging insights into IgA1 glycosylation abnormalities, cytokine-driven inflammation—particularly IL-17/IL-23 pathways—and genetic polymorphisms, including IL23R, offer promising avenues for future refinement of diagnostic and prognostic tools. However, these advances are not yet integrated into routine criteria due to insufficient clinical implementation. Taken together, the available evidence underscores the need for a nuanced and multidimensional diagnostic approach that combines clinical expertise with laboratory evaluation, imaging modalities, and, when indicated, histopathology. Strengthening awareness among pediatricians, improving early screening for renal involvement, and advancing the incorporation of immunogenetic markers into clinical practice hold the greatest potential for enhancing diagnostic accuracy, preventing complications, and improving long-term outcomes in children with IgA vasculitis.

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