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Review Article

# Overview of childhood vasculitis

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Pediatric vasculitis and adult vasculitis differ in several aspects. While both involve inflammation of blood vessels, pediatric vasculitis tends to present with distinct clinical features and may involve different types of blood vessels compared to adult vasculitis. Despite its relatively rare occurrence compared to adult vasculitis, pediatric vasculitis warrants careful attention due to its potential for profound and diverse clinical manifestations, ranging from mild cutaneous symptoms to life-threatening systemic complications. Childhood vasculitis should be suspected in children who present symptoms attributable to systemic inflammation and complications arising from multi-organ dysfunction. However, achieving a diagnosis necessitates thorough exclusion of alternative conditions manifesting similar symptoms and findings. Hence, children suspected of vasculitis should undergo meticulous history-taking, comprehensive physical examination, and requisite laboratory investigations, imaging studies, and sometimes tissue biopsies to elucidate the diagnosis. Early detection and treatment of childhood vasculitis are crucial, as the condition can affect various organs and potentially lead to life-threatening complications or long-term sequelae in adulthood if left untreated. This review aimed to provide an exhaustive overview of childhood vasculitis, outlining its epidemiology, classification, clinical presentation, diagnostic modalities, therapeutic strategies and outcome.

**Keywords:** Vasculitis, Child, Classification, Mucocutaneous lymph node syndrome, IgA vasculitis

## INTRODUCTION

Vasculitis encompasses a heterogeneous group of diseases characterized by inflammation and damage to the blood vessel walls. The primary etiology of vasculitis is often unknown; however, it can manifest as a secondary complication triggered by several factors, including infections, drug reactions, hypersensitivity reactions, and systemic rheumatic conditions such as systemic lupus erythematosus. The clinical presentation and severity of vasculitis depend on the size and location of affected blood vessels, the extent of vascular damage, and underlying histopathological characteristics. Although primary vasculitis is less prevalent in pediatric populations than in adults, conditions such as Henoch-Schönlein purpura (HSP) and Kawasaki disease (KD), the most frequent forms of vasculitis in children, exhibit a higher incidence in this demographic [1,2]. Early identification and management of pediatric vasculitis are crucial, as prompt intervention can mitigate disease progression, prevent long-term organ damage, and improve overall outcomes for affected children. Furthermore, the unique considerations in managing pediatric patients, such as their growth and developmental stages, underscore the importance of a multidisciplinary approach tailored to meet their specific needs. Therefore, this review aimed to provide an overview of childhood vasculitis emphasizing its epidemiological characteristics, classification systems, diagnostic approaches, and therapeutic modalities. Special emphasis is placed on the importance of early recognition and comprehensive care in improving outcomes for affected children.

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**Table 1.** EULAR/PRINTO/PReS 2008 classification scheme of childhood vasculitis

| Category | Criteria |
|---|---|
| 1. Predominately large vessel | |
| 1.1. Takayasu arteritis | Angiographic abnormalities of the aorta or its branches and pulmonary arteries (aneurysm/dilatation, narrowing, occlusion, or arterial wall thickening not due to fibromuscular dysplasia) and at least one of the following:<br>• Pulse deficit (lost/decreased/unequal peripheral artery pulse[s]) and/or claudication induced by activity<br>• Systolic BP difference >10 mmHg between any limbs<br>• Bruits or thrills over the aorta and/or its major branches<br>• Hypertension<br>• Elevated acute-phase reactants (ESR or CRP) |
| 2. Predominately medium-sized vessel | |
| 2.1. Childhood polyarteritis nodosa | Abnormalities in a small- or medium-sized artery (necrotizing vasculitis on biopsy or aneurysms, stenoses, or occlusions not due to fibromuscular dysplasia by angiography) and at least one of the following systemic features:<br>• Skin involvement (livedo reticularis, tender subcutaneous nodules, superficial or deep skin infarctions)<br>• Myalgia or muscle tenderness<br>• Hypertension<br>• Peripheral neuropathy (sensory peripheral neuropathy or motor mononeuritis multiplex)<br>• Kidney involvement (proteinuria, hematuria, or red blood cell casts, or GFR <50% normal for age) (proteinuria, hematuria, or red blood cell casts, or glomerular filtration rate of less than 50 percent the normal value for age) |
| 2.2 Cutaneous polyarteritis nodosa | No classification criteria proposed |
| 2.3. Kawasaki disease | Fever for five or more days and at least four of the five following features:<br>• Bilateral bulbar conjunctival injection<br>• Oral mucous membrane changes<br>• Polymorphous exanthema<br>• Perineal or peripheral extremity changes<br>• Cervical lymphadenopathy<br>(Patients with fever and fewer than four of the above criteria can also be diagnosed with Kawasaki disease if coronary artery abnormalities are detected either by 2D echocardiography or coronary angiography) |
| 3. Predominately small vessel | |
| 3.1. Granulomatous | |
| 3.1.1. Granulomatosis with polyangiitis (Wegener's granulomatosis) | Requires three of the following six features:<br>• Granulomatous inflammation within the wall of an artery or in the perivascular or extravascular area on biopsy<br>• Upper airway involvement (chronic/recurrent purulent or bloody nasal discharge or crusting, recurrent epistaxis, nasal septum perforation/saddle nose deformity, or chronic/recurrent sinus inflammation)<br>• Laryngotracheobronchial stenosis<br>• Pulmonary involvement (chest radiograph, CT showing nodules, cavities, or fixed infiltrates)<br>• Kidney involvement (hematuria, proteinuria, or red blood cell casts, or necrotizing pauci-immune glomerulonephritis)<br>• P-ANCA or C-ANCA positivity |
| 3.1.2. Eosinophilic granulomatosis with polyangiitis (Churg-Strauss syndrome) | No classification criteria proposed |

**Table 1.** Continued

| Category | Criteria |
|---|---|
| 3.2. Nongranulomatous | |
| 3.2.1. Microscopic polyangiitis | No classification criteria proposed |
| 3.2.2. Henoch-Schönlein purpura (IgA vasculitis) | Purpura (commonly palpable and in crops) or petechiae (without thrombocytopenia) with lower limb predominance and one or more of the following:<br>•Diffuse abdominal pain<br>•Leukocytoclastic vasculitis or proliferative glomerulonephritis with predominant IgA deposition<br>•Arthritis or arthralgia<br>•Kidney involvement (hematuria, red blood cell casts, or proteinuria) |
| 3.2.3. Isolated cutaneous leukocytoclastic vasculitis | No classification criteria proposed |
| 3.2.4. Hypocomplementemic urticarial vasculitis | No classification criteria proposed |
| 4. Other vasculitides | |
| 4.1. Behçet disease | |
| 4.2. Vasculitis secondary to infection (including hepatitis B associated polyarteritis nodosa), malignancies, and drugs, including hypersensitivity | |
| 4.3. Vasculitis associated with connective tissue diseases | |
| 4.4. Isolated vasculitis of the central nervous system | |
| 4.5. Cogan syndrome | |
| 4.6. Unclassified | |

EULAR: European League against Rheumatism, PRINTO: Paediatric Rheumatology International Trials Organization, PReS: Pediatric Rheumatology European Society, BP: blood pressure, ESR: erythrocyte sedimentation rate, CRP: C-reactive protein, GFR: glomerular filtration rate, ANCA: Antineutrophil cytoplasmic antibody, IgA: immunoglobulin A.

## MAIN SUBJECTS

### Epidemiology

Pediatric vasculitis is uncommon in children, with incidence rates reported between 12 to 53 cases per 100,000 individuals under 17 years of age [1-5]. However, despite its rarity, pediatric vasculitis exhibits significant heterogeneity. Therefore, evaluating incidence rates by specific underlying cause, rather than overall rates, provides a more accurate picture. These rates also vary depending on the population studied. However, globally, HSP is the most prevalent primary vasculitis in children, while KD is more common in Korea, Japan, and other Asian regions. In Korea, the average incidence rate of KD among children under 5 years was 196.9 per 100,000 individuals during 2015 to 2017, ranking second globally after Japan [1]. However, interestingly, stringent nonpharmacological interventions implemented worldwide during the COVID-19 pandemic, including Korea, have been associated with a significant decrease in KD incidence [6]. In Korea, KD rates declined by 40% post-pandemic compared with pre-pandemic levels. This suggests a link between environmental factors, such as infections, and the occurrence of childhood vasculitis [7]. Regarding HSP, a Korean study utilizing the national insurance database reported an incidence rate of 55.9 per 100,000 children under 18 years, exceeding the 14~20 (29.9) per 100,000 incidence rate reported internationally [2]. In contrast, other vasculitis like polyarteritis nodosa (PAN), are extremely rare, with an incidence less than 1 per 100,000 individuals [8,9]. Similarly, Antineutrophil cytoplasmic antibody (ANCA)-associated vasculitis (AAV) and Takayasu arteritis (TA) are rarely observed in children, with reported incidence rates ranging from 1 to 6.3 per million and 1 to 2.6 per million-population, respectively [10-13].

### Classification systems

Similar to adults, the primary classification system for childhood vasculitis categorizes patients based on the size of the affected vessels. The most commonly used classification criteria for vasculitis, approved at the 1994 Chapel Hill Consensus Conference, endorsed by the American College of Rheumatology (ACR) [14], faced limitations. Designed primarily for adult vasculitis, these criteria excluded common childhood conditions such as KD and HSP. Consequently, many pediatric vasculitis

cases remained unclassified using these criteria. Therefore, in 2005, the European League against Rheumatism (EULAR) and the Pediatric Rheumatology European Society (PReS) proposed classification criteria specifically for primary vasculitis in children under 18 years [15]. These were further revised in 2008 through a collaboration with the EULAR, PReS, and the Paediatric Rheumatology International Trials Organization (PRINTO), improving sensitivity for four specific diseases (HSP, Wegener's granulomatosis, childhood PAN, and TA) (Table 1) [16].

The second Chapel Hill Consensus Conference held in 2012 provided the most recent nomenclature and extensive framework for vasculitis classification, including specific disease definitions [17]. The Diagnostic and Classification Criteria in Vasculitis Study (DCVAS) plan was established to collect data on patients with vasculitis and develop enhanced classification criteria for adults with vasculitis. This led to the revision, approval, and publication of ACR/EULAR criteria for AAV subtypes, including granulomatosis with polyangiitis (GPA), microscopic polyangiitis (MPA), and eosinophilic granulomatosis with polyangiitis (EGPA) in 2022 [18-20]. These MPA criteria mark the first categorical system for this disease; however, their applicability to children remains to be evaluated.

## Clinical manifestations and diagnosis

The diagnosis of primary vasculitis in children can be challenging due to its highly variable clinical manifestations. These variables depend on factors such as the location and severity of inflammation, extent of vascular damage, and resultant hemodynamic changes. Therefore, vasculitis should be considered as a differential diagnosis in children presenting with unexplained systemic inflammation or multi-organ involvement. Table 2 summarizes the features suggestive of vasculitis. Additionally, distinguishing between primary vasculitis and other conditions that exhibit similar symptoms, or secondary vasculitis, is necessary (Table 3) [21,22].

To diagnose suspected vasculitis, a detailed medical history, physical examination, and basic laboratory tests are essential. The medical history should include recent infections, medications, and detailed family history. Physical examination involves measuring blood pressure in the extremities, auscultating vascular bruits, and assessing peripheral pulses. Additionally, examining for tender nodules, purpura, ulcers, or livedo reticularis on the skin, and performing a neurological evaluation for peripheral neuropathy are warranted.

Laboratory investigations typically include a complete blood count, erythrocyte sedimentation rate, C-reactive protein level, liver function tests, blood urea nitrogen level, creatinine level, and urinalysis to assess renal and hepatic involvement. Depending on the suspected vasculitis type, autoimmune antibody tests (antinuclear antibodies, ANCA) and complement levels may be performed. While tissue biopsy or characteristic vascular abnormalities on imaging studies, such as CT angiography and magnetic resonance angiography, are gold standards for confirmation, particularly when medium- or large-vessel vasculitis is suspected, specific clinical features may be sufficient for diag-

**Table 2.** Clinical manifestations and laboratory findings suspected for vasculitis

| Clinical feature | Laboratory finding |
|---|---|
| • Constitutional symptoms<br>:fever, weight loss, unexplained fatigue<br>• Cutaneous lesions<br>: palpable purpura, petechiae, ulcerations, livedo reticularis<br>• Neurological symptoms<br>: headache, mononeuritis multiplex, peripheral neuropathy, stroke<br>• Musculoskeletal symptoms<br>: arthralgia or arthritis, myalgia or myositis, tenosynovitis<br>• Hypertension<br>• Renal manifestations<br>: nephritis and renal insufficiency<br>• Respiratory involvement<br>: pulmonary infiltrates and pulmonary hemorrhage<br>• Cardiovascular involvement<br>: cardiac ischemia and arrhythmias | • Elevated acute-phase reactants (ESR or CRP)<br>• Leukocytosis, anemia, thrombocytosis<br>• Eosinophilia<br>• Positive ANCA<br>• Increased von Willebrand factor<br>• Cryoglobulinemia<br>• Circulating immune complexes<br>• Hematuria |

ESR: erythrocyte sedimentation rate, CRP: C-reactive protein, ANCA: Antineutrophil cytoplasmic antibodies.

**Table 3.** Differential diagnosis for primary systemic vasculitis in children

| Category | Criteria |
|---|---|
| Infection | • Bacteria/Fungi: Acute bacterial endocarditis, Meningococcemia, Group A streptococcal infections<br>• Viruses: HIV, HBV, CMV, EBV, Parvovirus B19, HSV, VZV, SARS-CoV-2<br>• Others: Tuberculosis, Syphilis, Leishmaniasis, typhus, and rickettsialpox |
| Tumors | • Lymphoma<br>• Leukemia |
| Autoimmune/systemic inflammatory diseases | • Systemic lupus erythematosus<br>• Dermatomyositis<br>• Systemic sclerosis<br>• Sarcoidosis<br>• Inflammatory bowel disease |
| Drug and toxin exposure | • Leflunomide<br>• Anti-TNF agents<br>• Anti-thyroid agents<br>• Antimicrobials<br>• Diuretics<br>• Anticonvulsants<br>• Cocaine<br>• Marijuana |
| Non-inflammatory mimicking conditions | • Neurofibromatosis<br>• Thrombocytopenia<br>• Antiphospholipid syndrome<br>• Thrombosis<br>• Raynaud phenomenon<br>• Coarctation |

HIV: human immunodeficiency virus, HBV: hepatitis B virus, CMV: cytomegalovirus, EBV: Epstein–Barr virus, HSV: herpes simplex virus, VZV: Varicella-zoster virus, SARS-CoV-2: severe acute respiratory syndrome coronavirus 2, TNF: tumor necrosis factor.

nosing certain vasculitis syndromes, such as KD, HSP, and GPA (Table 1) [23].

## Management

The principles of treatment for primary vasculitis in children prioritizes distinguishing it from similar conditions (Table 3). Treatment plans are then tailored based on disease activity, severity, and a risk-benefit analysis. As pediatric vasculitis treatment adheres to specific guidelines for each vasculitis type, this review emphasizes KD and HSP management. KD and HSP, the most common causes of primary vasculitis in children, may exhibit self-limiting clinical courses. Effective treatment strategies have been proposed based on an understanding of the characteristic clinical courses and potential long-term complications.

For KD, intravenous immunoglobulin (IVIG) at a dose of 2 g/kg and aspirin (30~50 mg/kg/day or 80~100 mg/kg/day) during the acute phase are the primary treatments used to reduce the risk of cardiovascular complications, such as coronary artery aneurysms [24]. Persistent or recurrent fever after IVIG may necessitate re-administration of IVIG or steroids. Infliximab, a tumor necrosis factor (TNF) inhibitor, may be used for patients unresponsive to these treatments [24]. Following the febrile phase, low-dose aspirin (3~5 mg/kg/day) is administered, exerting an antiplatelet effect by inhibiting thromboxane A2 and prostacyclin synthesis. If echocardiography at around 6 to 8 weeks shows no coronary artery aneurysms, low-dose aspirin may be discontinued [25].

Most patients with HSP receive symptomatic treatment. Graduated therapy is commonly used based on the clinical presentation, with nonsteroidal anti-inflammatory drugs (NSAIDs) for patients with joint and/or abdominal pain. Steroids are used for patients with severe abdominal pain that interferes with their oral intake and those unresponsive to NSAID or complications like nephritis [26]. Patients with HSP nephritis who do not respond to glucocorticoids may be candidates for other immunosuppressive therapies, such as cyclophosphamide, mycophenolate mofetil, or rituximab [27].

For primary vasculitides other than KD and HSP, symptoms are often chronic or recurrent, necessitating the use of steroids as the mainstay of treatment to maintain remission. Although

steroids alone may be sufficient for mild cases, more severe cases often require combination therapy with cyclophosphamide. Patients with various pediatric systemic vasculitides refractory to conventional therapy or experiencing frequent relapses, consideration may benefit from other immunosuppressive agents, such as methotrexate and azathioprine, and biologics (e.g., TNF inhibitors, rituximab, and tocilizumab) [28].

### Outcomes

The prognosis of primary vasculitis in pediatric patients depends on several factors: the underlying cause, disease severity, involvement of major organs, treatment response, and treatment-related complications. KD and HSP, the predominant types, typically exhibit spontaneous recovery with a favorable prognosis as long-term complications are infrequent. Instead, for other forms of primary vasculitides, effective immunosuppressive therapies have improved survival and remission rates. In 2008, a systematic review encompassing both adult and pediatric populations reported 5-year survival rates for various types of systemic vasculitis: GPA (approximately 75%), MPA (45%~75%), EGPA (68%~100%), HSP (75% in adult-onset, higher in pediatric-onset), medium-vessel vasculitis including PAN (75%~80%), KD (>99% at 5 years), and TA (70%~93%) [29].

In KD, long-term morbidity depends on the severity of coronary artery involvement. Data analysis was conducted on demographic characteristics, clinical manifestations, treatment modalities, and coronary complications of KD between 2015 and 2017 [1]. The study identified 15,378 KD cases over 3 years, with 17.1% exhibiting coronary artery dilatation (18.2% in 2015, 17.7% in 2016, and 15.1% in 2017) and 1.7% developing coronary aneurysms. Among these, giant coronary aneurysms (internal diameter >8 mm) developed in 19 (0.17%) patients. Among patients who were unresponsive to initial IVIG therapy, the rate of coronary artery dilatation was 27.2%, with an incidence of giant aneurysms of 0.27%. Two fatalities were recorded during the study period: one due to multi-organ failure at 15 months and the other due to hepatic encephalopathy at 83 months.

HSP typically exhibits a favorable prognosis [30-33], with spontaneous resolution observed in the majority of cases within weeks to months. However, a minority of patients may experience complications such as renal involvement or disease recurrence, underscoring the importance of long-term monitoring and individualized management strategies to optimize patient outcomes. Recurrences of HSP occur in approximately one-third of patients and are notably more prevalent in slightly older children (aged over 8 years), particularly in those exhibiting nephritis, acute inflammation, or having undergone steroid treatment [26,31,33-35]. Kidney involvement, observed in around 20 to 54 percent of pediatric HSP cases [31,36,37], tends to be more prominent among older children and adults [38-40]. Short-term kidney outcomes for HSP are generally favorable, with complete recovery documented in 94 percent of children and 89 percent of adults, typically within an average period of 18 months [41]. However, long-term consequences of HSP nephritis can lead to chronic kidney disease, accounting for 1%~2% of end-stage kidney disease cases [42].

A meta-analysis of nine case-control studies involving 969 HSP nephritis patients identified clinical risk factors for CKD, including older age at disease onset, low glomerular filtration rate, and an initial presentation with nephrotic or nephritic-nephrotic syndrome, all of which were associated with unfavorable kidney outcomes [43]. Conversely, a presentation characterized by hematuria and mild proteinuria, with or without hematuria, tended to correlate with more favorable outcomes.

In a retrospective cohort study conducted in Italy involving 208 children aged 0 to 18 years who met the criteria for HSP [44], treatment outcomes were examined. NSAIDs were administered to sixty-seven percent of patients, while corticosteroids were given to 62 patients. The use of corticosteroids was primarily due to renal dysfunction (13% of the cohort), persistent skin lesions (9%), severe abdominal pain (5%), or scrotal involvement (2%). Only 2 patients required immunosuppressive therapy (azathioprine and cyclophosphamide), one due to persistent proteinuria and the other due to non-response to corticosteroid therapy. Both patients demonstrated a positive response to immunosuppressive drugs and experienced favorable outcomes. No severe infections or adverse reactions were reported during treatment.

## CONCLUSION

Childhood vasculitis presents a multifaceted challenge, requiring a nuanced understanding of its diverse manifestations and tailored management strategies. Early recognition, accurate diagnosis, and comprehensive care are crucial to prevent persistent complications in vital organs. Continued research efforts

and collaborative clinical approaches are essential for advancing our understanding of childhood vasculitis and refining therapeutic interventions to improve the long-term prognosis and quality of life for pediatric patients.

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## CONFLICT OF INTEREST

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