# Nephropathy in the Course of Henoch-Schönlein Purpura

***Essay***
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بسم الله الرحمن الرحيم

# (وَفَوْقَ كُلِّ ذِي عِلْمٍ عَلِيمٌ)

صدق الله العظيم

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# Abstract

Henoch-Schönlein purpura (HSP) is a non thrombocytopenic, purpuric, and systemic vasculitis that occurs most commonly in children. It is an acute immunoglobulin A (IgA) mediated leukocytoclastic vasculitis that affects primarily children.

It is frequent in the first decade of life; with a reported incidence of 13.5 per 100,000.The onset is often following an upper respiratory tract infection.

The dominant clinical features of HSP are cutaneous purpura, arthritis, abdominal pain, gastrointestinal bleeding, orchitis, and nephritis.

Although the prognosis for unselected children with Henoch-Schönlein purpura (HSP) is relatively good, severe nephritis remains the major cause of morbidity and mortality in patients with HSP.

Renal involvement can occur in 20 to 34 percent of children with HSP. Severe abdominal symptoms, an age of more than 4 years, and persistent purpura increased the risk of renal involvement.

**Keywords:**

**Vasculitis**, **Henoch-Schönlein Purpura**, **Nephropathy in HSP**

*To my Parents*

*To whom I owe all my success*

*To my loving Husband*

*The greatest support in my life*

*To my beloved Son*

*To whom this work took me at his expense*

# ***List of Abbreviations***

| | |
|---|---|
| **ACE** | Angiotensin I Converting enzyme. |
| **ACR** | American Collage Of Rheumatology. |
| **Agt** | Angiotensinogen. |
| **ANCAs** | Antineutrophil cytoplasm antibodies. |
| **ASO** | Antistreptolysin O. |
| **BUN** | Blood urea nitrogen. |
| **C5** | Complement 5. |
| **CCr** | Creatinine clearance. |
| **CH 50** | Total hemolytic complement. |
| **ChE** | Cholinesterase. |
| **CHO** | Carbohydrate. |
| **CNS** | Central nervous system. |
| **CRF** | Chronic renal failure. |
| **DIC** | Disseminated intravascular coagulation. |
| **ECP** | Eosinophil cationic protein. |
| **ESR** | Erythrocyte sedimentation rate. |
| **ESRD** | End stage renal disease. |
| **ETs** | Endothelines. |
| **FAB** | Antigen binding fragment. |
| **Fc** | Crystalizable fragment. |
| **GAS** | Group A beta hemolytic streptococcus. |
| **GIT** | Gastrointestinal tract. |
| **HSN** | Henoch-Schönlein nephritis. |
| **HSP** | Henoch-Schönlein purpura |
| **HSPN** | Henoch-Schönlein purpura nephritis. |
| **ICs** | Immune complexes. |

| | |
|---|---|
| **IgA** | Immunoglobuline A. |
| **IgAN** | Immunoglobuline A nephropathy. |
| **IL1RN-2** | Interleukin-1 receptor antagonist allele. |
| **ILs** | Interleukins. |
| **ITP** | Immune thrombocytopenic purpura. |
| **KD** | Kilo Dalton. |
| **MRI** | Magnetic resonant imaging. |
| **MUPC** | Methylprednisolone and Urokinase Pulse Therapy With Cyclophosphamide. |
| **MUPT** | Methylprednisolone and Urokinase Pulse Therapy. |
| **PP** | Plasmapharesis. |
| **RAS** | Renine angiotensin system. |
| **RPGN** | Rapidly progressive glomerulonephritis. |
| **SHS** | Schönlein-Henoch Syndrome |
| **SLE** | Systemic Lupus Erythematosis. |
| **TGF** | Transforming growth factor. |
| **TNF** | Tumor necrosis factor. |
| **UK** | Urokinase. |
| **WBCs** | White blood cells. |

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# *Introduction*

Henoch-Schönlein Purpura is the most common form of acute vasculitis primarily affecting children ***(Liu et al, 2004).*** It is a multisystem IgA-mediated vasculitis with self limited course, affecting the skin, joints, gastrointestinal tract and kidneys ***(Gedalia, 2004).***

Henoch-Schönlein Purpura nephritis (HSN) is diagnosed when hematuria and/or proteinuria were associated with a characteristic purpuric eruption and/or abdominal or joint pain (at least 2 of these 3 clinical signs) ***(Shin et al, 2005).*** Immunofluorescence microscopic techniques have demonstrated the presence of IgA deposits in the glumeruli and in the vessel wall ***(Ferrario & Rastaldi, 2005).***

Although the prognosis of unselected children with HSP is relatively good, severe nephritis remains the major cause of morbidity and mortality in patients with HSP ***(Halling et al, 2005).*** It is thus recommended that patients with HSP Nephritis are followed for longer periods of time ***(Chang et al, 2005).***

Even patients with mild renal symptoms at onset of HSP carry a risk for severe long term complications ***(Ronkainen et al, 2002).***

# *Aim of the essay*

The aim of this study is to focus that patients with HSP should be followed up with urine analysis and assessment of renal functions for at least 6 monthes.

We also aim to clarify the beneficial effect of several immunosuppressive agent and other new lines in treating severe HSP.

# *Vasculitis*

## ***Definitions:***

The term **vasculitis** indicates the presence of inflammation in a blood vessel wall. The inflammatory infiltrate may be one that is predominantly neutrophilic, eosinophilic or mononuclear ***(James and Ross, 2001)***.

**Perivasculitis** describes inflammation around the blood vessel wall but without involvement of the mural structure itself ***(Jennette et al., 1994).***

**Vasculopathy** a broader term indicates an abnormality of blood vessels that may be inflammatory but may also be degenerative or may result from intimal proliferation ***(James and Ross, 2001).***

## ***Classification:***

**Table( 1-1): Chapel Hill Consenus Conference on Nomenclature of Systemic Vasculitis:**

| | |
|---|---|
| **Large-Vessel Vasculitis** | |
| Giant cell (temporal) arteritis | Granulomatous arteritis of the aorta and its major branches, with a predilection for the extracranial branches of the carotid artery. *It often involves the temporal artery. It usually occurs in patients older than 50 years and is often associated with polymyalgia rheumatica.* |
| Takayasu's arteritis | Granulomatous inflammation of the aorta and its major branches .*It usually occurs in patients younger than 50 years.* |
| **Medium-Vessel Vasculitis** | |
| Polyarteritis nodosa | Necrotizing inflammation of medium-sized or small arteries without glomerulonephritis or vasculitis in arterioles, capillaries, or venules. |
| Kawasaki disease | Arteritis involving large, medium-sized, and small arteries and associated with mucocutaneous lymph node syndrome. *Coronary arteries are often involved. Aorta and veins may be involved .It usually occurs in children.* |
| **Small-Vessel vasculitis** | |
| Wegener's granulomatosis | Granulomatous inflammation involving the respiratory tract and necrotizing vasculitis affecting small to medium-sized vessels. *Necrotizing glomerulonephritis is common.* |

| | |
|---|---|
| Churg-Strauss syndrome | Eosinophil-rich and Granulomatous inflammation involving the respiratory tract and necrotizing vasculitis affecting small to medium-sized vessels and associated with asthma and eosinophilia . |
| Microscopic polyangiitis | Necrotizing vasculitis with few or no immune deposits, affecting small vessels. *Necrotizing arteritis involving small-and medium-sized arteries may be present. Necrotizing glomerulonephritis is very common. Pulmonary capillaritis often occurs.* |
| Henoch-Schonlein Purpura | Vasculitis, with IgA -dominant immune deposits affecting small vessels. *Typically involves skin, gut, and glomeruli and is associated with arthralgias or arthritis.* |
| Essential cryoglobulinemic vasculitis | Vasculitis with cryoglobulin immune deposits affecting small vessels and associated with cryoglobulins in serum. *Skin and glomeruli are often involved.* |
| Cutaneous leukocytoclastic angiitis | Isolated cutaneous leukocytoclastic angiitis without systemic vasculitis or glomerulonephritis. |

*Large vessels : aorta ,and larger branches directed toward major body regions.
*Medium vessels: renal, hepatic , coronary, and mesenteric arteries.
*small vessels : venules , capillaries arterioles, and intraparenchymal distal arteries that connect with arterioles.
**Essential component are shown in normal type, italicized type represent usual, but not essential, component. ***(Jennette et al., 1994).***