Glomerulonephritis associated with systemic disease

A- Lupus Nephritis

B- Henoch-Schonlein Purpura

A- Lupus Nephritis :- Lupus is an autoimmune disease that triggers your immune system to attack your tissues. In addition to your kidneys, lupus can damage your brain, heart, joints, skin and other parts of your body.

Stages of Lupus Nephritis

Based on the kidney biopsy, will know the stages or classification of lupus nephritis. **The six stages, or classes, are based on:**

a- Cell changes in the glomeruli as seen under the microscope

b- Immune deposits seen on immunofluorescence

C- Electronic microscopy

Stage 1: Minimal mesangial glomerulonephritis

- Minor kidney damage
- No obvious other signs or symptoms

Stage 2: Mesangial proliferative glomerulonephritis

- Some clear damage to the kidney
- Extra blood or protein in your urine that your health care team can detect with lab tests

Stage 3 : Focal glomerulonephritis

- More damage that amounts to *less than* 50% of important blood vessels in your kidney
- Higher amounts of blood or protein in urine
- Possible high blood pressure

Stage 4: Diffuse proliferative nephritis

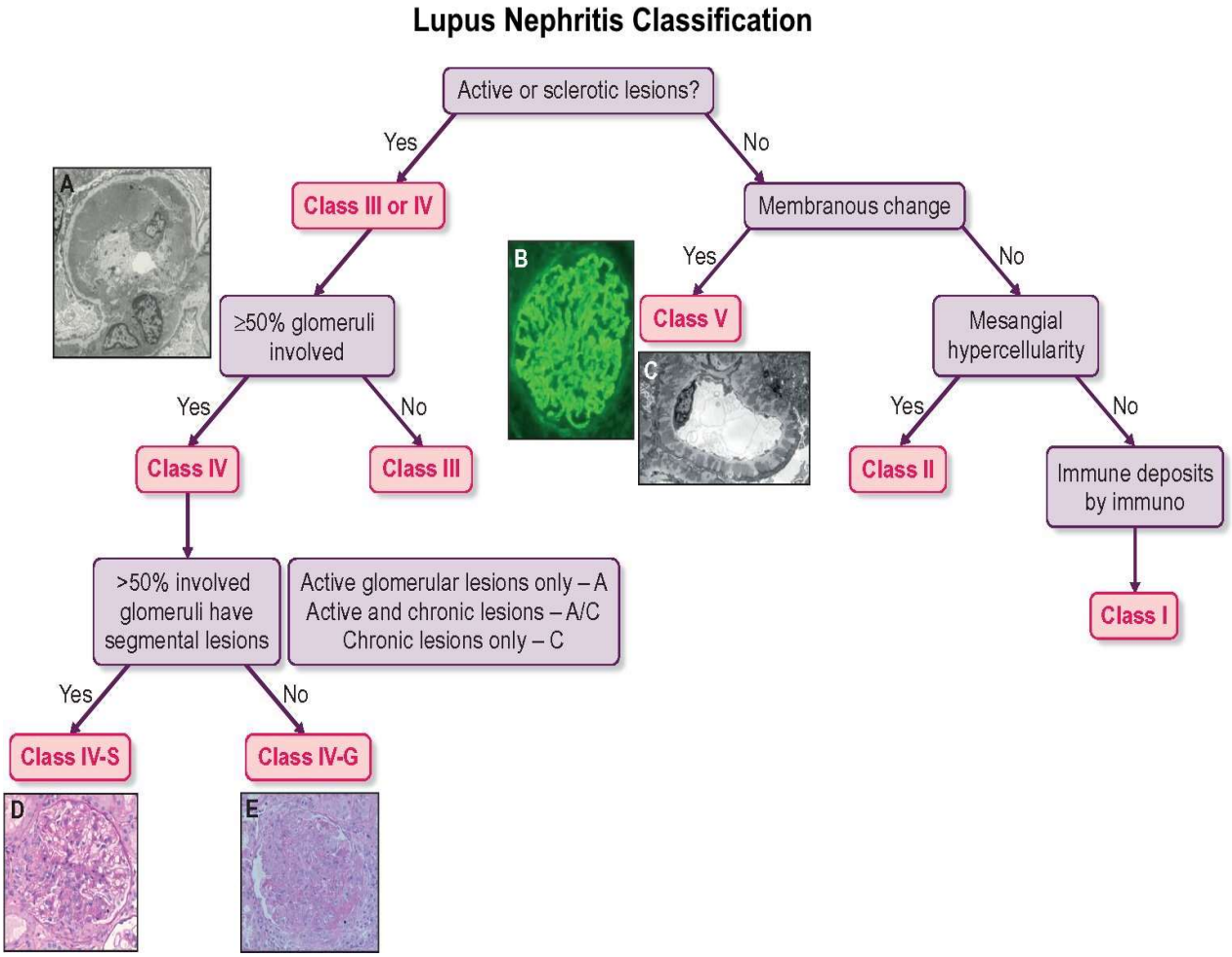
- Damage that amounts to *more than* 50% of important blood vessels in the kidney
- Blood or protein in urine
- Possible high blood pressure
- Possible need for dialysis as kidneys stop working properly

Stage 5: Membranous glomerulonephritis

- Thickening of important parts of the kidney
- Blood or protein in urine
- Possible high blood pressure
- Dialysis or possible kidney transplant

Stage 6: Advanced sclerotic

- Damage to more than 90% of important kidney blood vessels
- When treatment is possible, may need dialysis or kidney transplant
- Plus other signs: Blood or protein in urine and high blood pressure



Symptoms of lupus nephritis

Symptoms of lupus nephritis tend to develop about five years after lupus symptoms first appear. But lupus nephritis can be the first — and sometimes the only — manifestation of systemic lupus erythematosus (SLE). Lupus nephritis can cause:

1. Edema (swelling due to fluid buildup) in your lower body or around your eyes.
2. Fever with no known cause.
3. Hematuria (blood in the urine).

4. High blood pressure.
5. Increased urination, especially at night.
6. Joint pain or swelling.
7. Muscle pain.
8. Proteinuria (protein in the urine), which often causes your urine to look foamy.
9. Red skin rash on the face.
10. Weight gain due to excess fluid in your body.

Causes lupus nephritis

No one knows why some people with SLE develop lupus nephritis. Your family background and ancestry, medical conditions, and environmental factors such as exposure to chemicals or pollutants may all play a role in causing the disease.

Lupus nephritis usually gets worse over time, which can lead to kidney failure. The cause of lupus in most cases, however, is unknown.

Pathophysiology

Autoimmunity plays a major role in the pathogenesis of lupus nephritis. The immunologic mechanisms include production of autoantibodies directed against nuclear elements. The characteristics of the nephritogenic autoantibodies associated with lupus nephritis are as follows :

1. **Antigen specificity** directed against nucleosome or double-stranded DNA (dsDNA) - Some anti-dsDNA antibodies cross-react with the glomerular basement membrane

2. **Higher-affinity autoantibodies** may form intravascular immune complexes, which are deposited in glomeruli

3. **Cationic autoantibodies** have a higher affinity for the anionic glomerular basement membrane

4. **Autoantibodies of certain isotypes** (immunoglobulin [Ig] G 1 and IgG 3) readily Activate complement

These autoantibodies form pathogenic immune complexes intravascularly, which are deposited in glomeruli. Alternatively, autoantibodies may bind to antigens already located in the glomerular basement membrane, forming immune complexes in situ. Immune complexes promote an inflammatory response by activating complement and attracting inflammatory cells, including lymphocytes, macrophages, and neutrophils.

Diagnosis of lupus nephritis

The diagnosis of lupus nephritis depends on

1- blood tests .

2- urinalysis :- a nephritic picture (i.e symptoms of nephritic) is found and red blood cell casts, red blood cells and proteinuria is found.

3- X-rays .

4- ultrasound scans of the kidneys .

5- kidney biopsy.

Treatment of lupus nephritis

1- **Corticosteroids** :- These strong anti-inflammatory drugs can decrease inflammation .

2- **Immunosuppressive drugs :-** These drugs, which are related to the ones used to treat cancer or prevent the rejection of transplanted organs, work by suppressing immune system activity that damages the kidneys. They include azathioprine (Imuran), cyclophosphamide (Cytoxan), voclosporin (Lupkynis) and mycophenolate (Cellcept).

3- Medications to prevent blood clots or lower blood pressure if needed .

The goals of treatment for lupus nephritis are to:

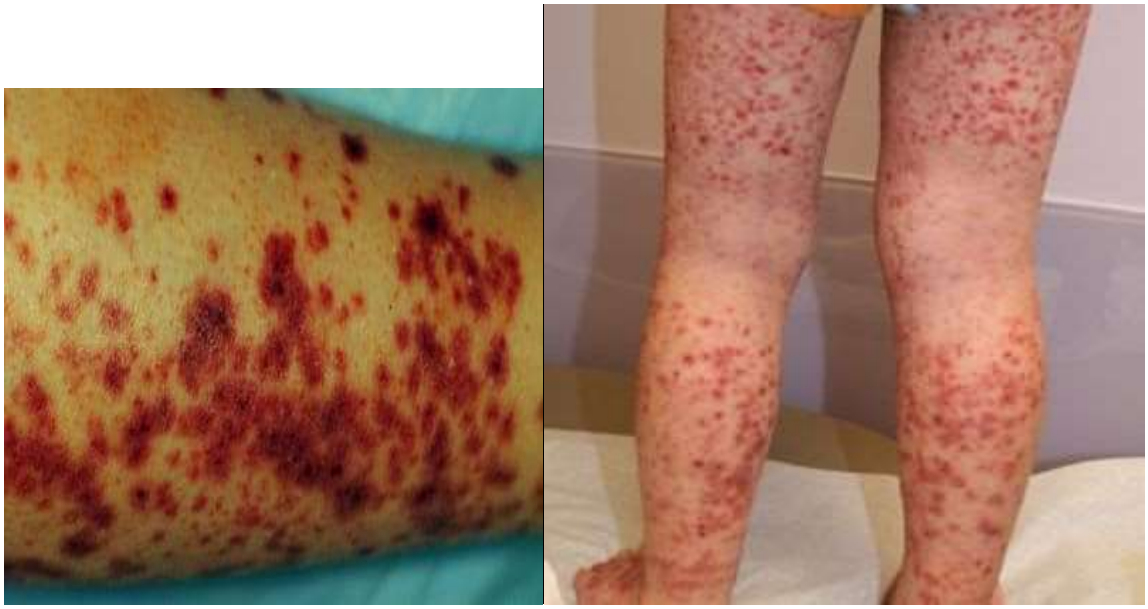
1- Reduce inflammation in your kidneys .

2- **Decrease** immune system activity .

3- Block your body's immune cells from attacking the kidneys directly or making antibodies that attack the kidneys .

B- Henoch-Schonlein Purpura

(HSP) is an acute immunoglobulin A (IgA)–mediated disorder characterized by a generalized vasculitis involving the small vessels of the skin, the gastrointestinal (GI) tract, the kidneys, the joints, and, rarely, the lungs and the central nervous system (CNS). It most commonly occurs in children. It most commonly occurs in children.



Symptoms of Henoch-Schonlein Purpura

1- The main symptom is a spotty rash

with numerous small bruises rash

2- joint pain and swelling

3- Abdominal pain

4- Blood in urine.

Before these symptoms begin, patients may have two to three weeks of fever, headache, and muscular aches and pains. Rarely, other organs, such as the brain, lungs, or spinal cord may be affected .

Causes and Risk Factors for Henoch-Schonlein Purpura

The exact cause of HSP is not known. The body's immune system is believed to play a role in targeting the blood vessels involved. An abnormal immune response to an infection may be a factor in many cases. Approximately two-

thirds of the cases of HSP occur days after symptoms of an upper respiratory tract infection develop.

Some causes of HSP have been linked to

1- vaccinations for typhoid, cholera, yellow fever, measles, or hepatitis B

2- foods

3 - Drugs

4- Chemicals

5- Insect bites.

6-Some experts also say that HSP is associated with the colder weather of fall and winter.

Diagnosis of Henoch-Schonlein Purpura

There is no specific test to diagnose HSP. It is diagnosed based on recognition of the classic symptoms, and exclusion of other conditions that can cause a similar rash. In many children with a classic rash, minimal testing is needed to establish a diagnosis of HSP. The rash is necessary for the diagnosis of HSP but is not always the first symptom to appear. When joint pain, swelling, or abdominal pain start before appearance of the rash, it can cause diagnosis can be challenging.

Tests in children with suspected HSP depend on the patient, but might include the following:

- Platelet count and coagulation studies to looks for other causes of bleeding.
- Laboratory tests to rule out other causes of vasculitis.
- Evaluation of kidney function by blood pressure check, creatinine level, electrolytes, and urine sample.

- In some patients, a biopsy may be taken of the skin, kidney or other tissue. Biopsies in patients with HSP often show high levels of a specific type of immune protein, called immunoglobulin A (IgA).

- Imaging of the bowels may be performed if abdominal pain is severe.

Treatment of Henoch-Schönlein purpura

Most of the time, Henoch-Schönlein purpura improves on its own without treatment. Medical care is more likely to be needed if HSP involves the kidneys.

To help the children to feel better, the doctor may recommend medicines such as:

- antibiotics, if an infection is causing the HSP .

- pain relievers (such as acetaminophen) .

- anti-inflammatory medicines (such as ibuprofen) to relieve joint pain and inflammation .

- corticosteroids (such as prednisone) for severe belly pain or kidney disease