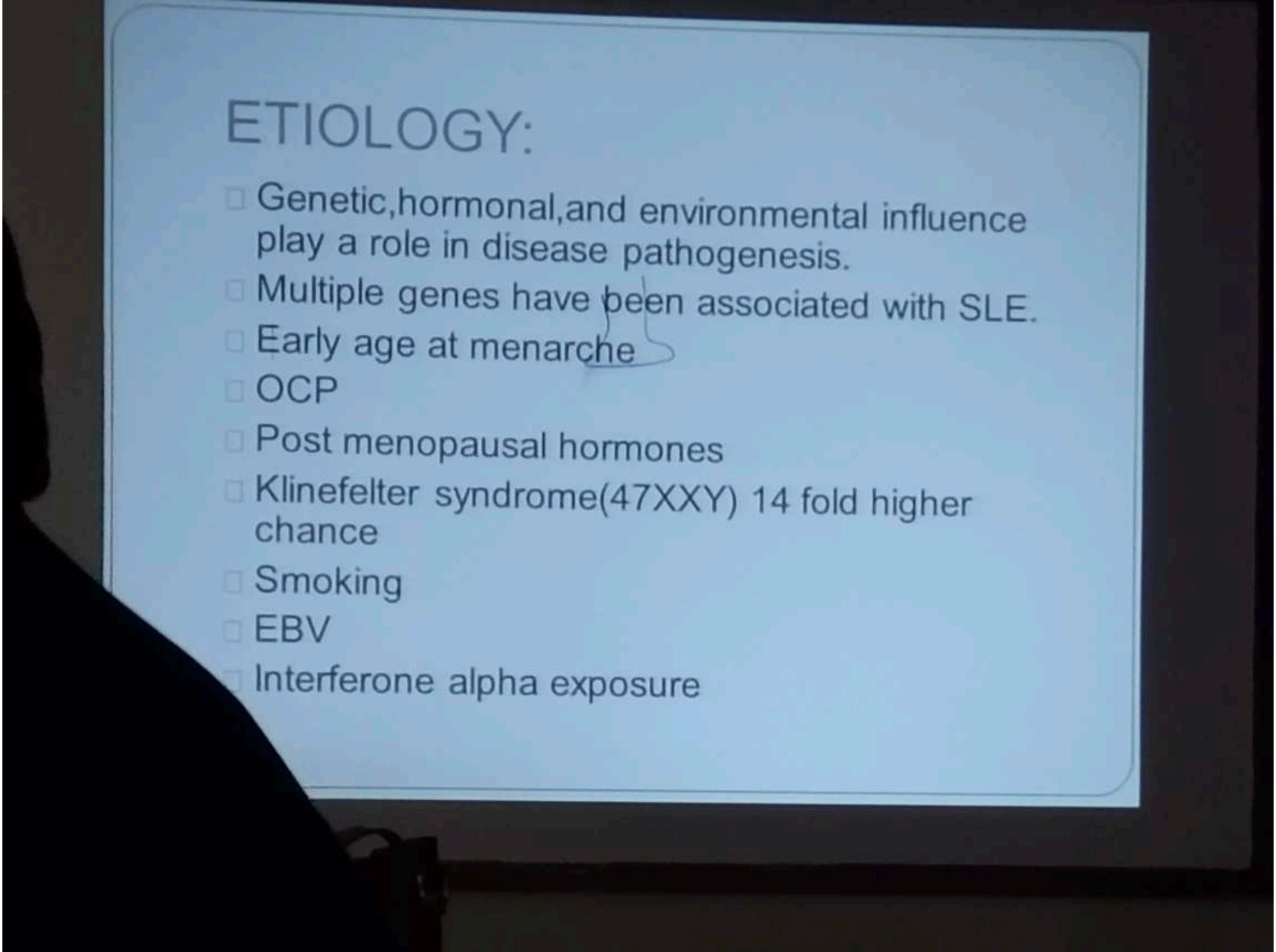
DEFINITION:

- A chronic inflammatory condition caused by an autoimmune disease. An autoimmune disease occurs when the body's tissues are attacked by its own immune system. Patients with lupus have unusual antibodies in their blood that are targeted against their own body tissues

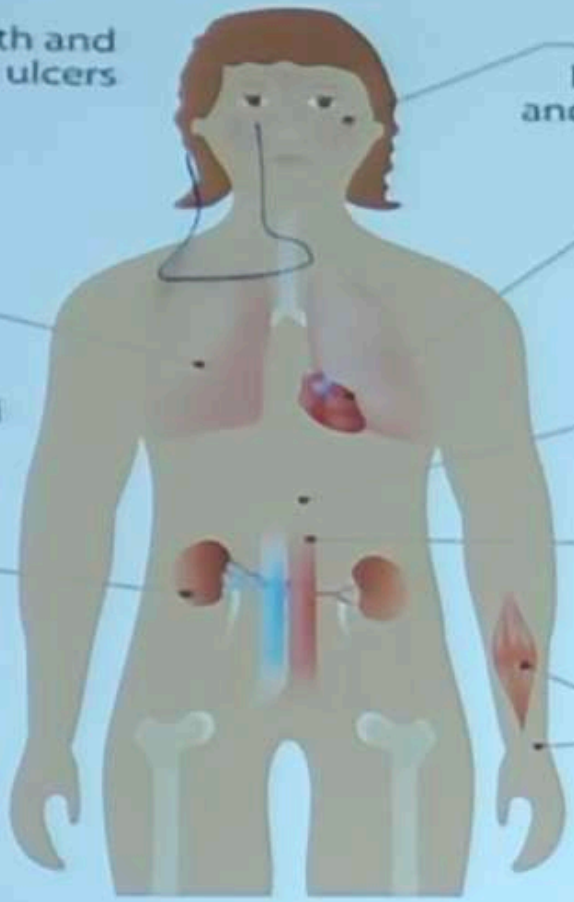

ETIOLOGY:
Genetic,hormonal,and environmental influence play a role in disease pathogenesis.
Multiple genes have been associated with SLE.
Early age at menarche
OCP
Post menopausal hormones
Klinefelter syndrome(47XXY) 14 fold higher chance
Smoking
EBV
Interferone alpha exposure

SYMPTOMS AND SIGNS:

- CONSTITUTIONAL:
  - fever
  - fatigue
  - weight changes

Systemic lupus erythematosus

Mouth and nose ulcers



Skin
butterfly rash
and red patches

Heart
- endocarditis
- atherosclerosis
- inflammation of the fibrous sac

Lungs
- pleuritis
- pneumonitis
- pulmonary emboli
- pulmonary hemorrhage

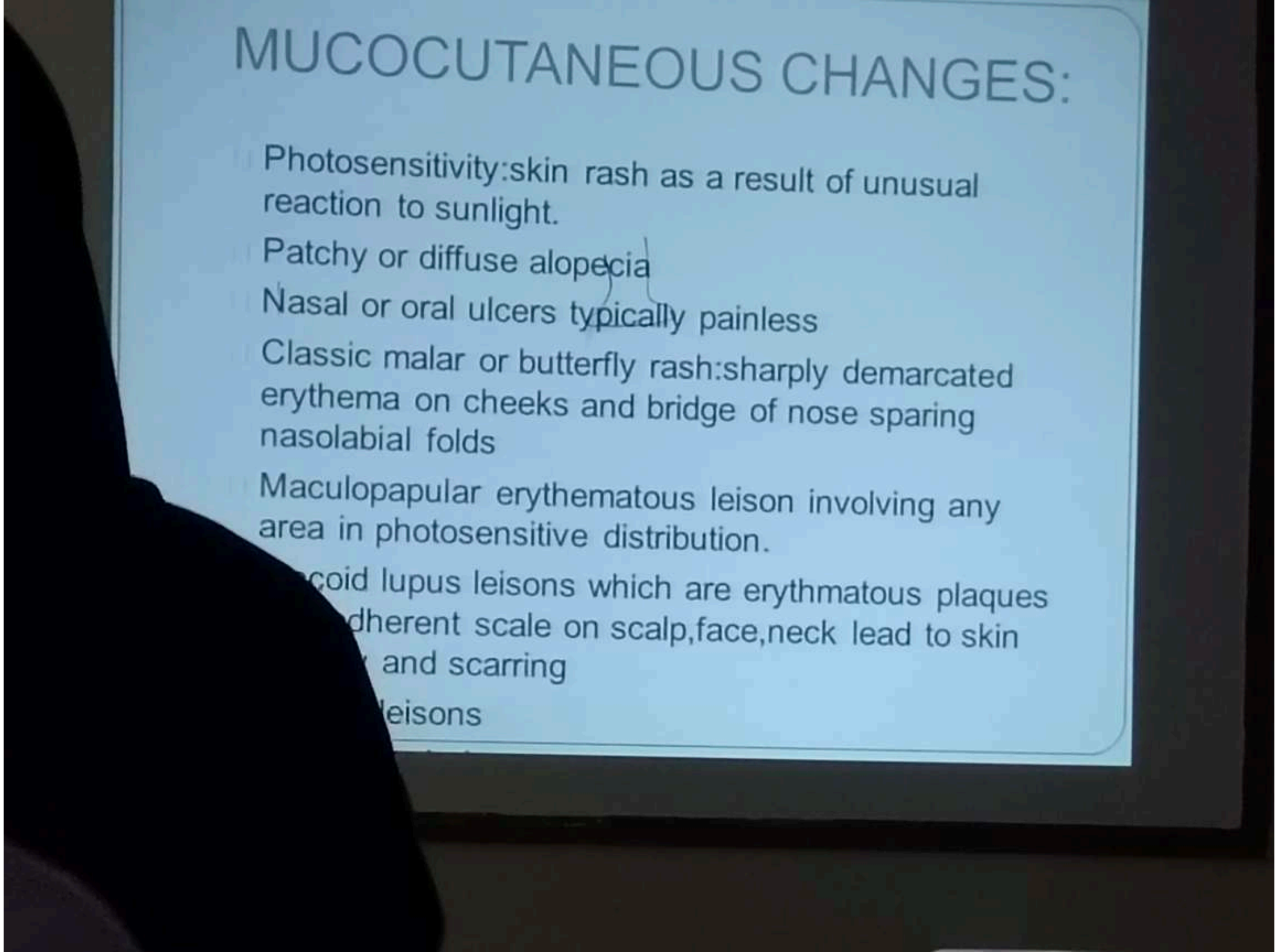
Severe abdominal pain

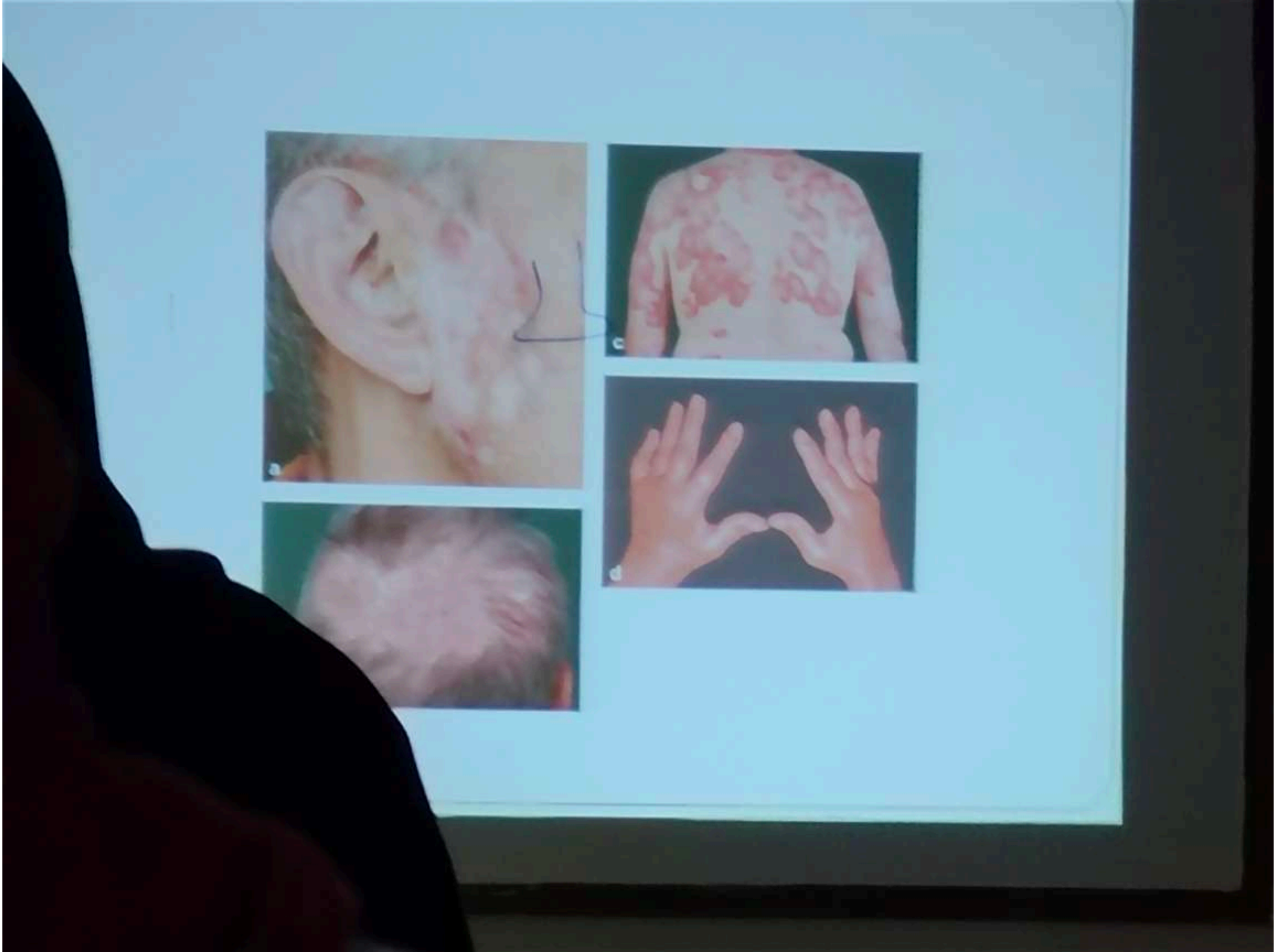
Kidneys
- blood in the urine

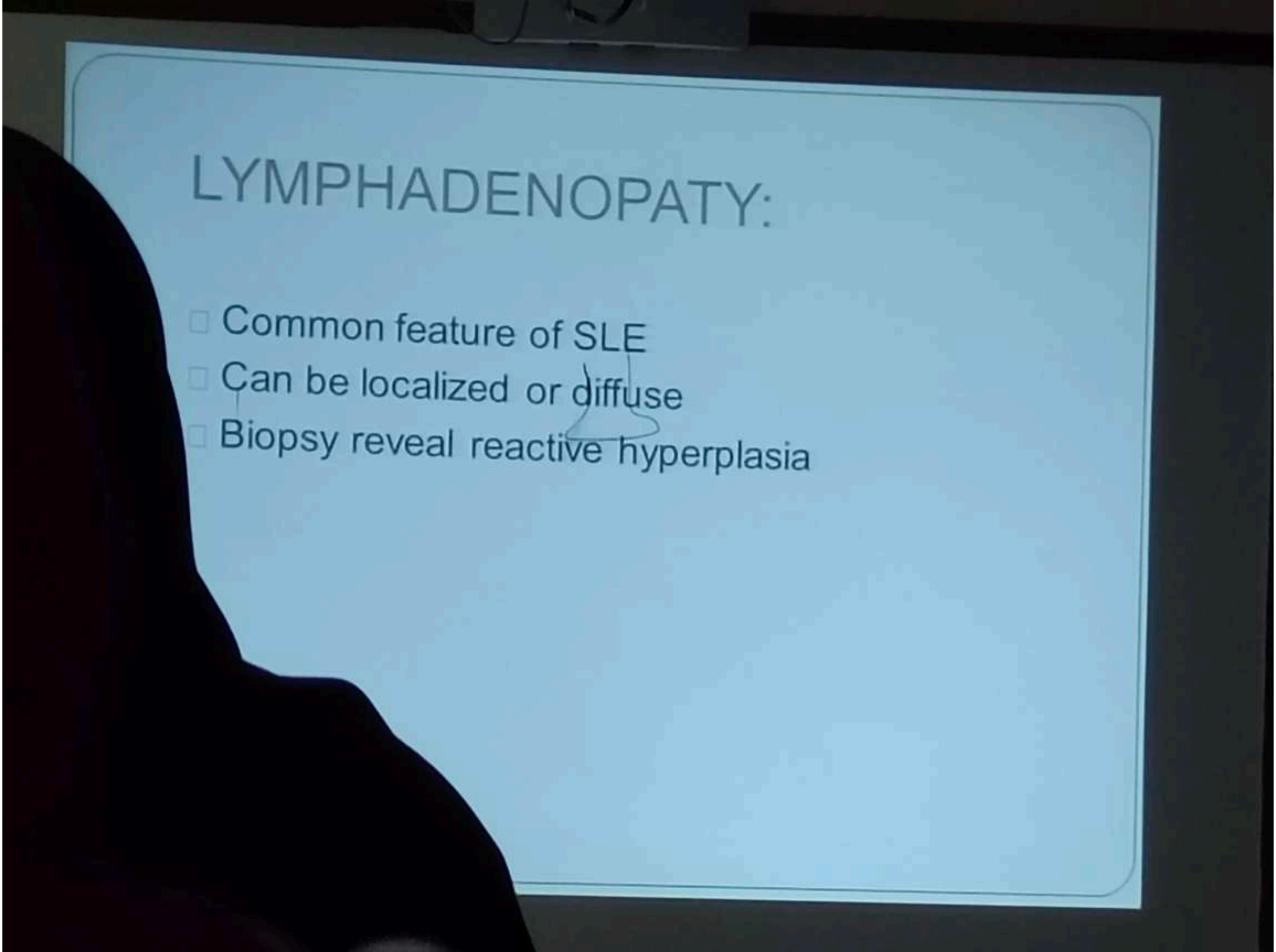
Blood
- anemia
- high blood pressure

Hair loss
High fever
Abnormal headache

Muscle and Joints
- pain and
- arthritisaches
- swollen joints

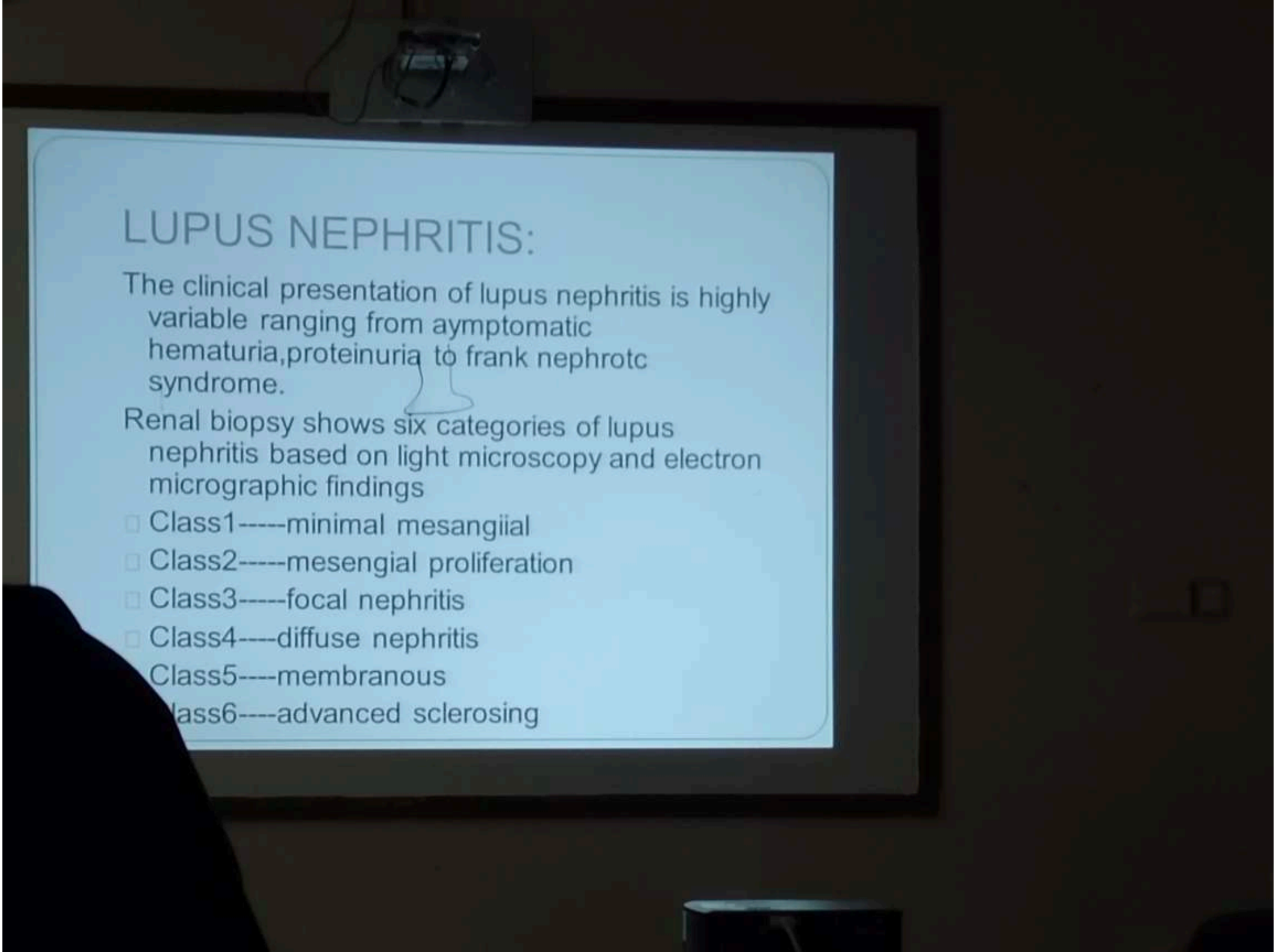

MUCOCUTANEOUS CHANGES:
Photosensitivity:skin rash as a result of unusual reaction to sunlight.
Patchy or diffuse alopecia
Nasal or oral ulcers typically painless
Classic malar or butterfly rash:sharply demarcated erythema on cheeks and bridge of nose sparing nasolabial folds
Maculopapular erythematous leison involving any area in photosensitive distribution.
coid lupus leisons which are erythmatous plaques
dherent scale on scalp,face,neck lead to skin
and scarring
leisons

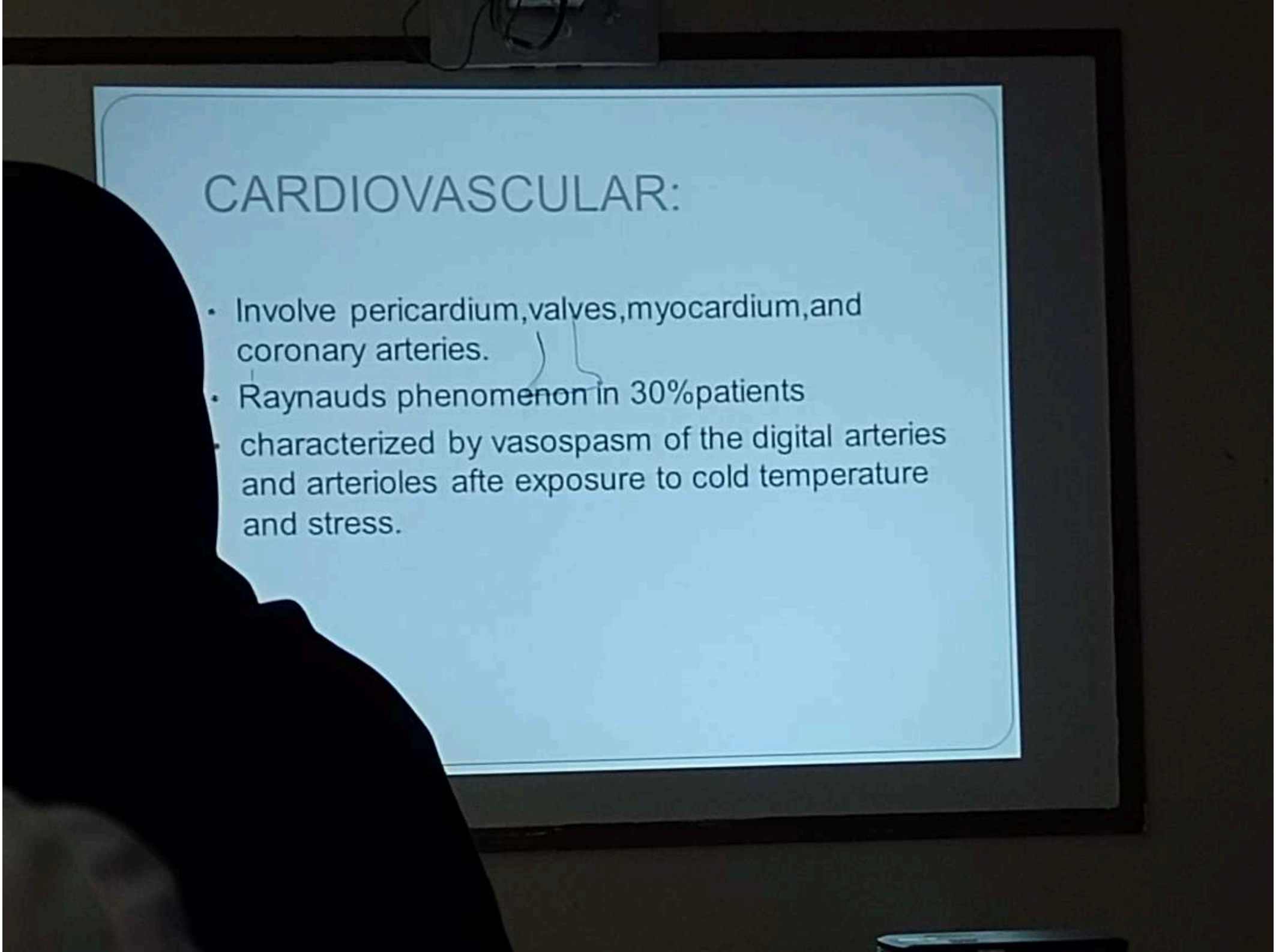



LYMPHADENOPATY:
Common feature of SLE
Can be localized or diffuse
Biopsy reveal reactive hyperplasia

MUSCULOSKELTAL:

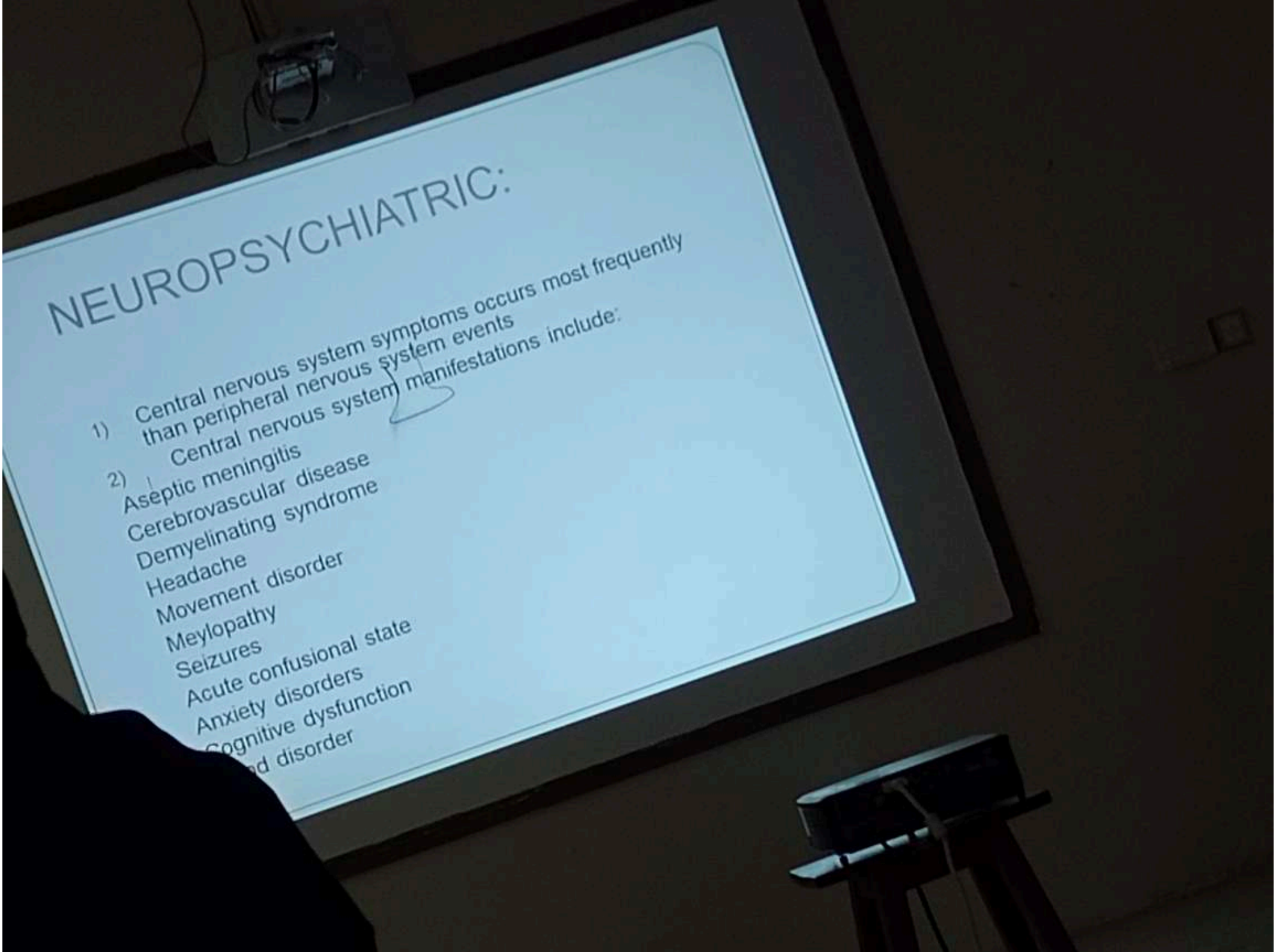
- Arthralgias ad arthritis in 95%
- Frequently involve wrists and small joints of hands
- Bony erosions rarely occur in SLE

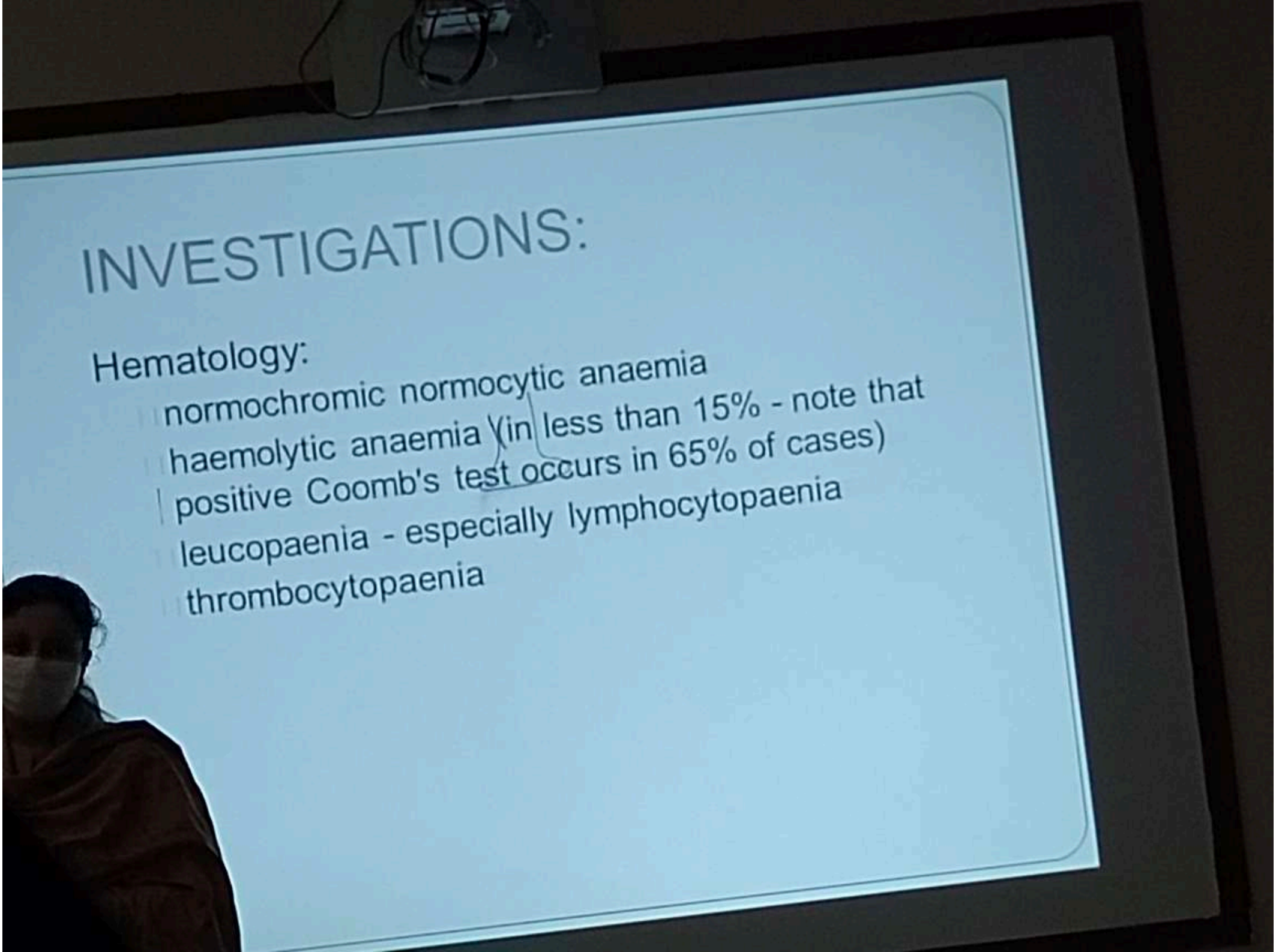

LUPUS NEPHRITIS:
The clinical presentation of lupus nephritis is highly variable ranging from aymptomatic hematuria,proteinuria to frank nephrotc syndrome.
Renal biopsy shows six categories of lupus nephritis based on light microscopy and electron micrographic findings
Class1-----minimal mesangiial
Class2-----mesengial proliferation
Class3-----focal nephritis
Class4----diffuse nephritis
Class5----membranous
lass6----advanced sclerosing

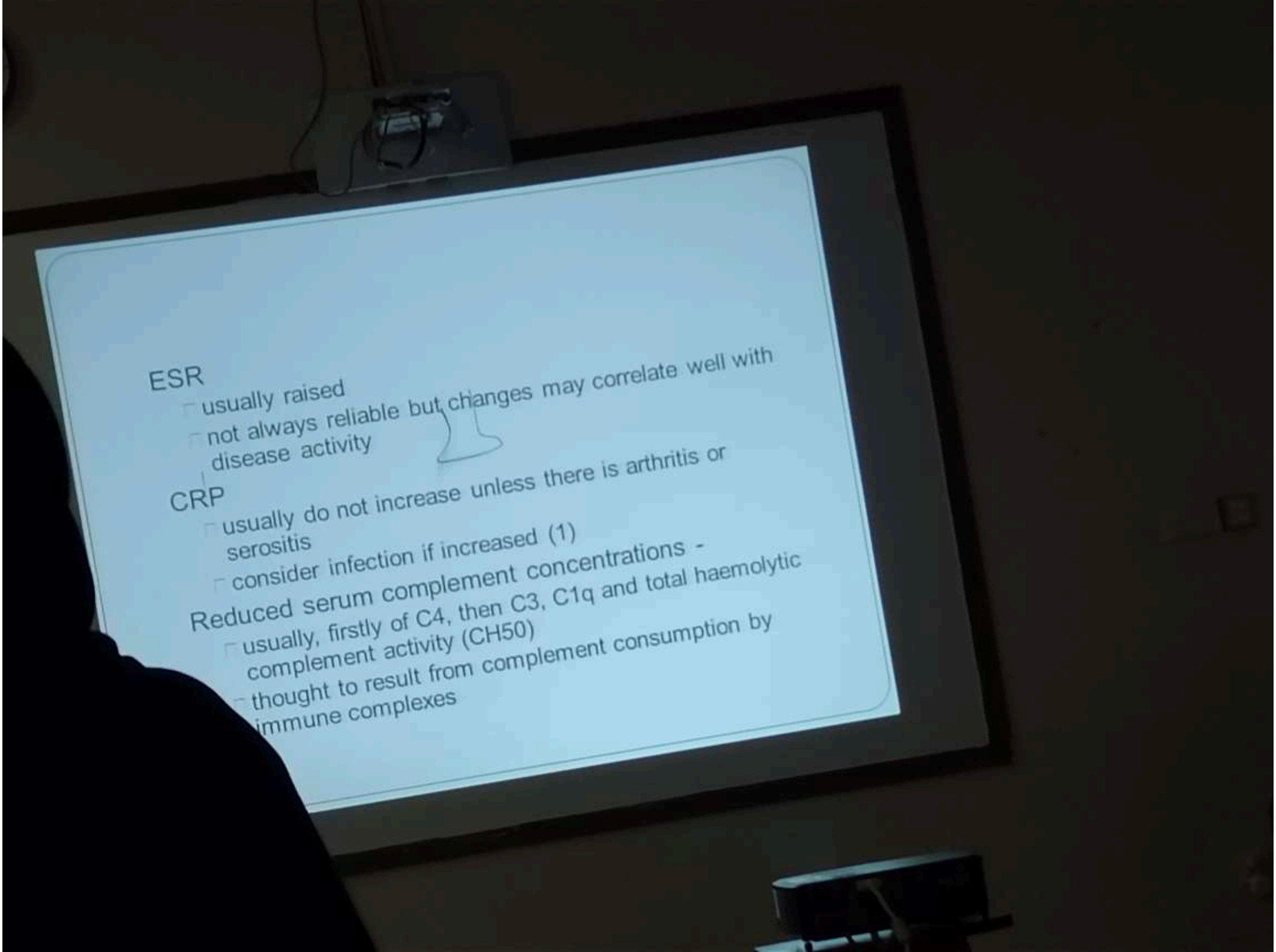

CARDIOVASCULAR:
• Involve pericardium,valves,myocardium,and coronary arteries.
• Raynauds phenomenon in 30%patients
• characterized by vasospasm of the digital arteries and arterioles afte exposure to cold temperature and stress.

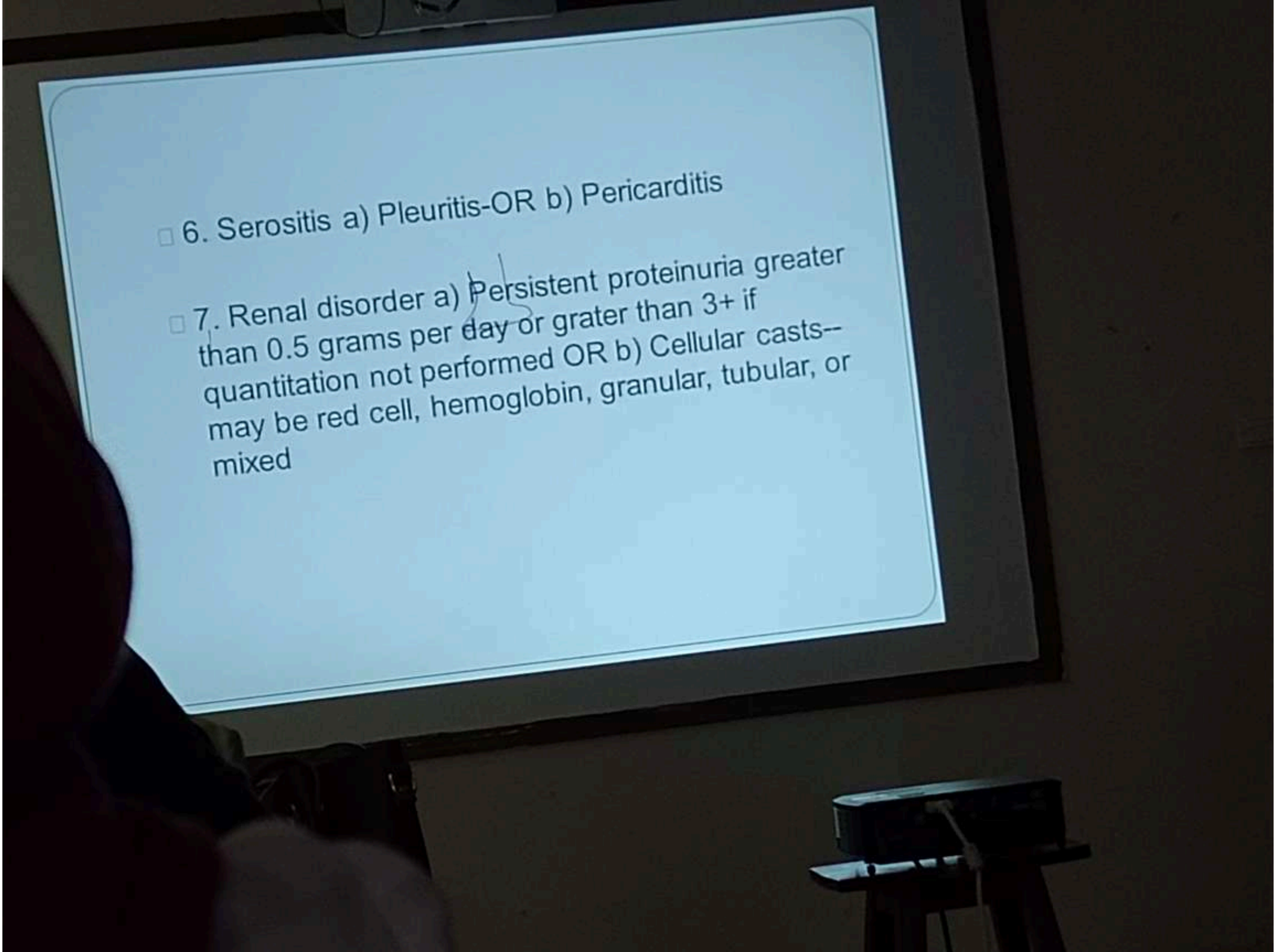
GASTROINTESTINAL:

- Perotonitis
- Pancreatitis
- Mesenteric vasculitis
- Intestinal pseudobstruction
- Lupus hepatitis


NEUROPSYCHIATRIC:
1) Central nervous system symptoms occurs most frequently than peripheral nervous system events
2) Central nervous system manifestations include:
Aseptic meningitis
Cerebrovascular disease
Demyelinating syndrome
Headache
Movement disorder
Meylopathy
Seizures
Acute confusional state
Anxiety disorders
Cognitive dysfunction
d disorder


INVESTIGATIONS:
Hematology:
normochromic normocytic anaemia
haemolytic anaemia (in less than 15% - note that positive Coomb's test occurs in 65% of cases)
leucopaenia - especially lymphocytopaenia
thrombocytopaenia


ESR
usually raised
not always reliable but changes may correlate well with disease activity
CRP
usually do not increase unless there is arthritis or serositis
consider infection if increased (1)
Reduced serum complement concentrations -
usually, firstly of C4, then C3, C1q and total haemolytic complement activity (CH50)
thought to result from complement consumption by immune complexes


6. Serositis a) Pleuritis-OR b) Pericarditis
7. Renal disorder a) Persistent proteinuria greater than 0.5 grams per day or grater than 3+ if quantitation not performed OR b) Cellular casts-- may be red cell, hemoglobin, granular, tubular, or mixed

10. Immunologic disorder
11. An abnormal titer of antinuclear antibody

4 out of 11 favours diagnosis of SLE

SKIN PROTECTION:

- Sun protection is a cornerstone of management of cutaneous lupus.
- Sun screen with SPF-30
- Topical glucocorticoids(hydrocortisone,triamicnolone,betamethasone,clobetasol)
- Topical tacrolimus and pimecrolimus is second line treatment.

Overview of management of SLE

Establish diagnosis

↓

Determine disease activity

↓

Assess severity and organ involvement

↓

Lifestyle (sun avoidance)
Topical agents
Symptomatic agents
nage co-morbidities

No major organ involvement
- Antimalarials
- Low-dose steroids
- Azathioprine/methotrexate

Major organ involvement
- Cyclophosphamide (IV)
- Mycophenolate mofetil
- Calcineurin inhibitors (CyA/tarcolimus)
- Biologics (rituximab/belimumab) **or**
- Enroll in a clinical trial

management of systemic lupus erythematosus -John A Reynolds, Ian N Bruce
views Series 7] Spring 2013

Treatment of Lupus Nephritis base on biopsy Class

Clasas	Treatment
I	Non specific
II	for 1 -3 months 20–40
III	Prednisolone 1mg/kg/d for 1 month then taper depending on the clinical response to 5-10 mg for 2-2.5 years or Pulses for 3 days in severly ill add immunosupressive when indicated, adjust immunosupressine with cbc
IV	
V	Prednisone for 1-3 months followed by low dose for 1-2 years Azathioprin , chlorambucil, cyclophosphamide or MMF may decrease proteinuria
VI	Prepare for hemo which is better than peritoneal Arrange for renal Tx, treat extra-renal lupus

SEROLOGY:AUTOANTIBODIES

ANA: - seen in 98% of patients

the specificity is low since ANAs are found in many other conditions such as scleroderma, polymyositis, dermatomyositis, rheumatoid arthritis

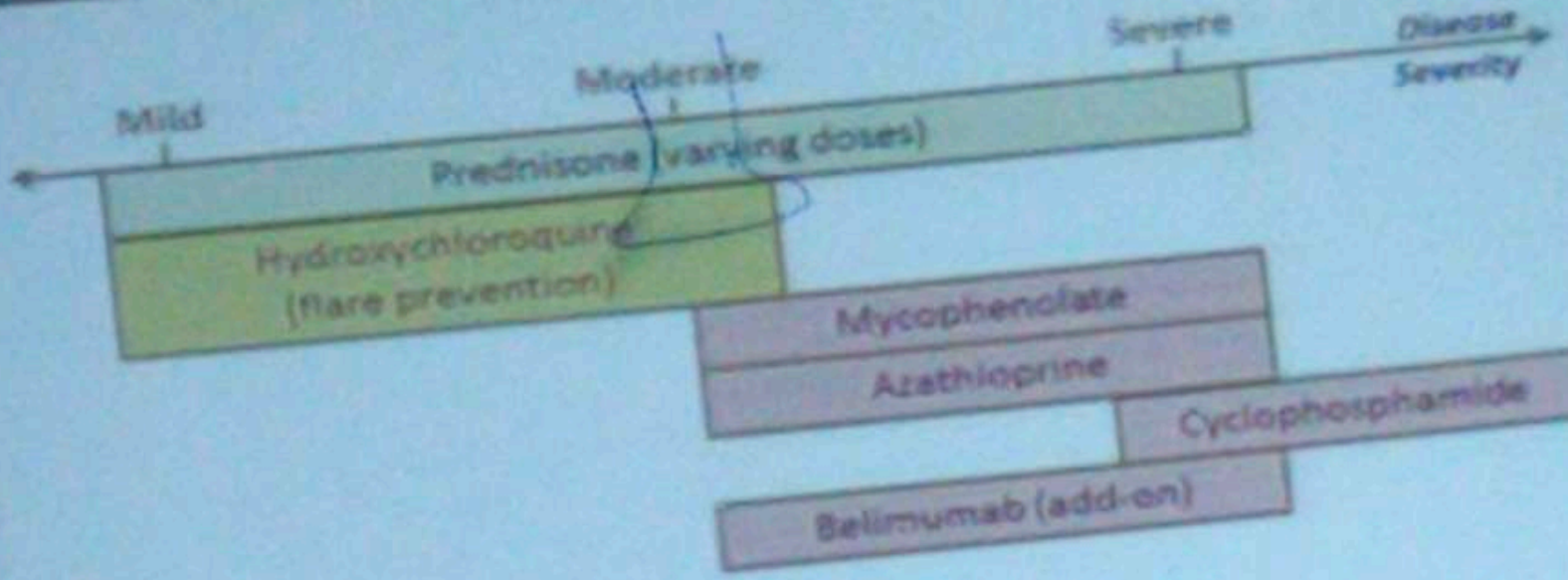
anti-double-stranded DNA (dsDNA) - highly specific for SLE, only present in around 70% of patients

other autoantibodies include - anti-Smith, anti-ribosomal P and anti-proliferating cell nuclear antigen (PCNA)

TREATMENT:

knowmedge

SLE Treatment



Hydroxychloroquine use requires annual ophthalmology visits due to risk of retinopathy