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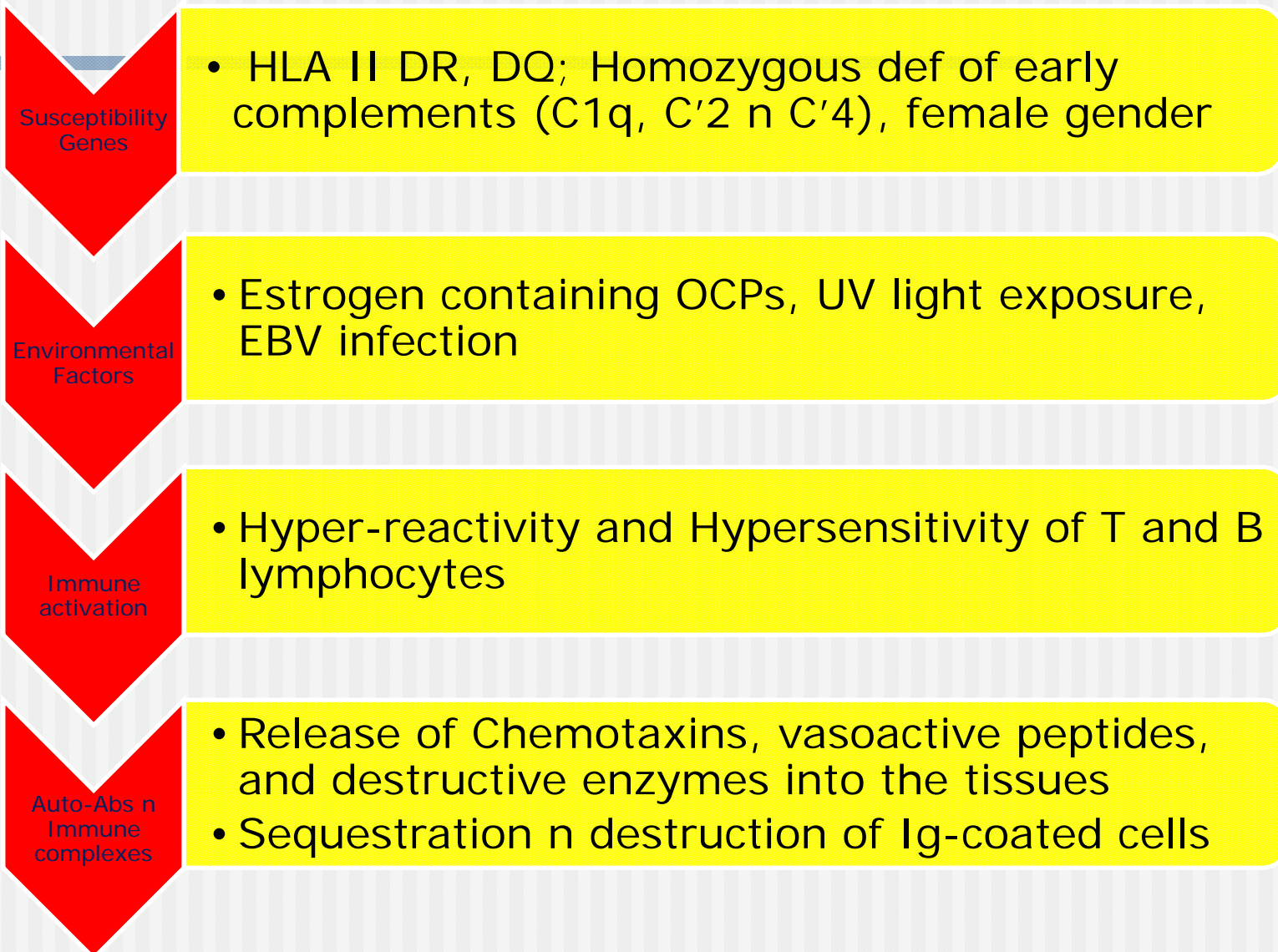
# Systemic Lupus Erythematosus

# SLE

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- Autoimmune disorder
- Cellular damage bcs of Auto-Abs n Immune-complexes

# SLE: Etio-pathogenesis



# SLE: Histopathology

- Skin Biopsy → T cell infiltrate at DEJ, perivascular and around appendages
  - Ig deposition at DEJ
- Renal Biopsy → Class I (MMLN)
  - Class II (MesPGN)
  - Class III (FLN): A, C, A/C
  - Class IV (DLN): S/G for A, C, A/C
  - Class V (MGP)
  - Class VI (DGS)

# Clinical Manifestations

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- Musculoskeletal:

- Non-erosive Polyarthrititis

If erosions....some alternative diagnosis (? RA)

If monoarthrititis...Ischemic bone necrosis  
(Steroids for SLE n not disease activity)

Hand deformity only in 10%

- Myalgia/ Myositis ....Also consider Steroid  
and Anti-malarial (HCQS) myopathy



# Clinical Manifestations

## ■ Muco-Cutaneous/Lupus Dermatitis:

i) DLE ii) SCLE iii) Others

DLE → circular, raised erythematous lesions with hyperpigmented rims & central atrophy

→ 5 % of pts with DLE....have SLE

→ 20% Of pts with SLE....have DLE

Photosensitivity → V-region of neck, upper back, extensor surfaces of arms

Malar Rash & recurrent oral ulcers

# Clinical Manifestations



# Clinical Manifestations

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- Renal/ Lupus Nephritis:

All pts to have urine analysis

Kidney biopsy if active urinary sediments/  
proteinuria

Histologic Class I-VI

Class IV → ESRD in 2 years (if untreated)

Better prognosis for Nephrotic synd as  
compared to Nephritic synd



# Clinical Manifestations

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- CNS/ PNS:

Unexplained Psychosis → At Presentation,  
during Acute flare-up n ...Steroid induced (1<sup>st</sup>  
week and > 40 mg/d of prednisolone)

Unexplained seizures

ATM

Mononeuritis Multiplex, Peripheral neuropathy

# Clinical Manifestations

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- Vascular occlusion:

TIA, CVA, ACS

CVA... Vasculitic, aPL-Ab(APLA synd),  
Libman-Sachs endocarditis asso.

- Resp.:

Pleuritis (with or without effusion)

ILD

Intra-alveolar hemorrhage

# Clinical Manifestations

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## ■ Cardiac:

Pericarditis (MC)

Myocarditis

Fibrinous endocarditis of Libman-Sachs

(Valvular insuff or embolic events)

## ■ Hemat:

NCNC Anemia of Chr ds, Hemolytic anemia

Leukopenia

TCP

# Clinical Manifestations

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## ■ GIT:

Autoimm Peritonitis

Hypertransaminasemia

Mesenteric vasculitis..... Ischemia, bleeding,  
perforation, sepsis

## ■ Ocular:

Sicca synd & non-sp. Conjunctivitis

Retinal vasculitis

Optic neuritis



# Diagnostic Criteria: ACR (4/11)

1. Malar rash
2. Discoid rash
3. Photosensitivity
4. Oral ulcers
5. Non-erosive arthritis ( $\geq 2$  joints)
6. Serositis
7. Renal...  $> 0.5$  g/d proteinuria or cellular cast
8. Neurologic...unexplained psychosis/seizures
9. Hemat...hemolytic anemia, leukopenia, TCP
10. Immunologic...Anti-dsDNA, Anti-Sm, aPL Ab
11. ANA (absence of drugs known to induce)

# Lab tests

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- For diagnosis:
  1. ANA: + in 95% of cases
  2. Anti-dsDNA: specific for SLE, Lupus Nephritis, high titers during Ac. Flare-up
  3. aPL Ab: not specific for SLE, venous/arterial thrombosis, fetal loss, TCP
  4. Std (routine) lab tests

# Lab tests

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- For Ac. Flare-up
  1. Tests that indicate status of organ involved such as Hb, Pl count, KFT, Urine R/M, LFT
  2. Others → Anti-dsDNA titer, C<sup>3</sup> complement levels, Anticardiolipin Ig G

# Treatment

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- Non-life threatening events/ Potentially reversible organ damage

- Arthralgia/ Arthritis

NSAIDs: renal dysfun esp if asso lupus nephritis

Antimalarials: retinal toxicity, ototoxicity, Peripheral neuropathy

Systemic steroids (low dose-0.07-0.3mg/Kg)

Methotraxate: BM suppression, pulm fibrosis



# Treatment

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➤ Malar rash/ Discoid rash/ Photosensitivity/  
Oral ulcers

Topical sunscreen (SPF at least 15): contact dermatitis

Systemic retinoids: congenital fetal anomalies

Topical glucocorticoids

Methotrexate : BM suppression, Pulm fibrosis,  
hapatotoxic

# Treatment

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- Life-threatening SLE

- Proliferative forms of Lupus Nephritis  
(Class III, IV & V)

Methyl Pred. 1g iv q 24h X 3d

f/b Oral Pred. 0.5-1 mg/Kg qd X 4-6 weeks f/b

Oral Pred. 5-10 mg qd (low dose maintenance)

+

Cyclophosphamide 500mg/m<sup>2</sup> monthly X 6 mths

# Treatment

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- Side-effects of therapy:
  - Systemic glucocorticoids →
    1. Infections
    2. HTN & volume overload
    3. Psychosis
    4. Hyperglycemia
    5. Osteoporosis
    6. Myopathy

# Treatment

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- Side-effects:

- Cyclophosphamide →

1. BM suppression
2. Gonadal failure
3. Hemorrhagic cystitis
4. Ca urinary bladder
5. Alopecia



# Treatment

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- Preventive therapies: usually for S/E of steroids n cytotoxic agents
  1. H. Influenza n Pneumococcal vaccine
  2. T/t for Osteoporosis, HTN n Dyslipidemia
  3. Prevention of UTI (adeq fluid intake n local hygiene)

# Prognostic factors

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- Serum Creatinine > 1.4 mg%
  - HTN
  - Nephritic synd >> Nephrotic synd
  - a PL Ab + status
- ❖ Leading causes of death → systemic disease activity, CKD, Infections

# SLE n Special Situations

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## ■ Pregnancy n Lupus →

1. Recurrent fetal losses: LMWH increases chances of fetal survival
2. High chances in aPL Ab+ status or Nephritis
3. Higher steroid requirement as placental dehydrogenase deactivates prednisolone
4. Neonatal lupus (skin rash + heart blocks)
5. S/E of prednisolone on fetus: LBW, CNS developmental anomalies, adult metab synd

# En Special Situations

Wolfe n APLA synd →

1/0 venous or arterial thrombosis and/or recurrent fetal losses & aPL + at least on two separate occasions

2/1x : long-term Anticoag, Target INR= 3.0

3/0 Microvascular thrombotic crisis (TTP/HUS)

4/0 Tissue damage in Brain n Kidney, high mortality, usually young pts with Nephritis

5/0 1/t : Plasma exchange/Plasmapheresis ; No

# Drug induced Lupus

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ANA + with fever, arthritis, rash n serositis

diff<sup>ed</sup> from SLE as....dsDNA only rarely +

....less female predilection

....resolves over weeks

after offending drug withdrawn



# Drug induced Lupus

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Drugs....

Anticonvulsants → Carbamazepine, Phenytoin

Antipsychotics → Lithium

Antithyroid → PTU

AntiHTN → ACEI, Thiazides,  $\beta$ -blockers,  
Hydralazine

Anti-arrhythmics → Procainamide, Propafenone

# Experimental therapies for SLE

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Against T cell-B cell interaction:

Anti CD40L Ab, Anti CTLA4-Ig fusion prot.

Anti CD20 Ab

Against Complement syst:

Anti C'5 Ab

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ARE YOU DONE WITH  
SLE(EPING) ???

ANY QUESTIONS ?

