

Quiz 3 review | September 21, 2016



Quiz 3 Review

- Autoimmune diseases
- Immunodeficiencies
- Transfusion
- Transplantation

Quiz 3 Review

- Autoimmune diseases

Lupus

Things You Must Know

- Typical patient: young woman with butterfly rash
- Symptoms unpredictable (relapsing/remitting)
- Multisystem (skin, kidneys, joints, heart)
- Antinuclear antibodies

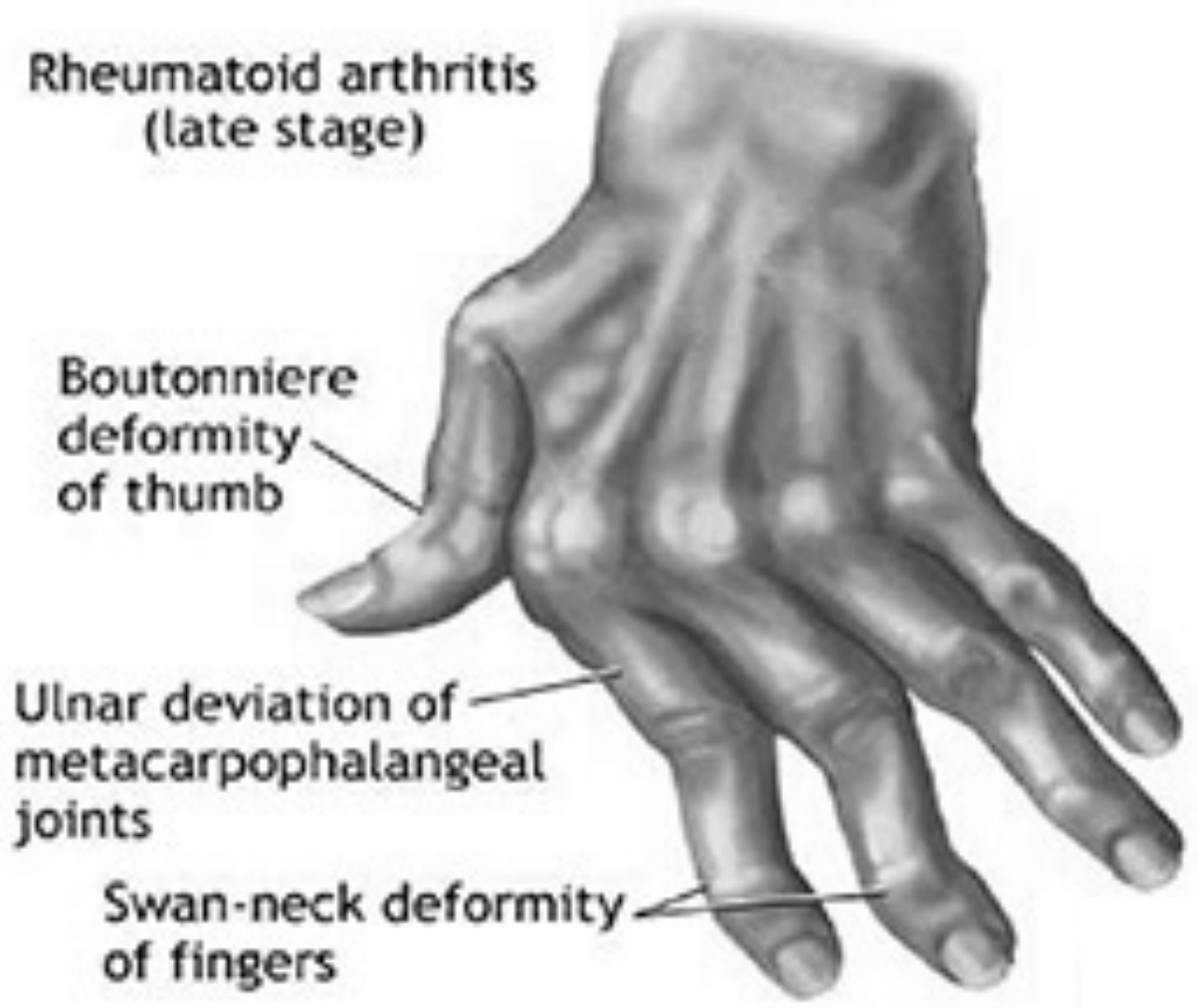


Lupus: butterfly rash

Rheumatoid Arthritis

Things You Must Know

- Symmetric, mostly small-joint arthritis
- Systemic symptoms (skin, heart, vessels, lungs)
- Rheumatoid factor
- Cytokines (especially TNF) cause damage



Rheumatoid arthritis
(late stage)

Boutonniere
deformity
of thumb

Ulnar deviation of
metacarpophalangeal
joints

Swan-neck deformity
of fingers

Rheumatoid arthritis joint deformities

Sjögren Syndrome

Things You Must Know

- Inflammatory disease of salivary and lacrimal glands
- Dry eyes, dry mouth
- T cells react against some Ag in gland; gland gets destroyed
- Increased risk of lymphoma



Sjögren syndrome: salivary gland enlargement

Scleroderma (Systemic Sclerosis)

Things You Must Know

- Excessive fibrosis throughout body: skin, viscera
- Claw hands, mask-like face
- Microvascular disease also present
- Diffuse and limited types



Systemic sclerosis: claw hands

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Immune Diseases Outline

Disease	Transmission	Defect	Clinical stuff
XLA	X-linked	No mature B cells; no Ig	Infant with recurrent bacterial infections
CVID	non x-linked	Defective Ab production	Infections, AI diseases, lymphoma
IgA deficiency	non x-linked	↓ IgA	Most common. Asx. AI diseases
Hyper-IgM	X-linked	↓ IgG, A, E T cell defect	Bacterial and fungal infections
DiGeorge	non x-linked	No thymus	Viral, fungal infections
SCID	half are x-linked	Many B and T cell defects	Bacterial and other infections

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Genotype	Antigens	Blood type
AA	A	A
AO		
BB	B	B
BO		
AB	A and B	AB
OO	None	O

Genotype	Antigens	Blood type
DD	D	Rh +
Dd	D	Rh +
dd	none	Rh -

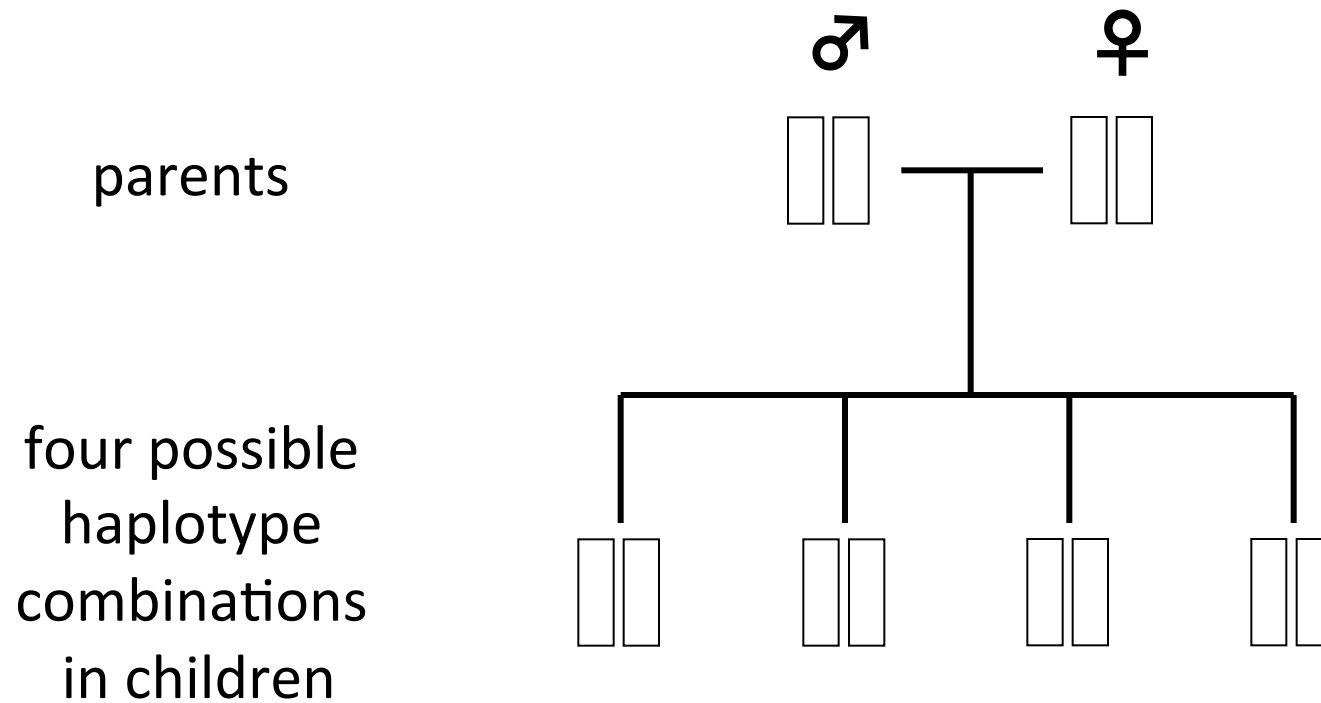
What can go wrong?

- Transfusion reactions
 - hemolytic
 - non-hemolytic
- Other complications
 - infections
 - iron overload

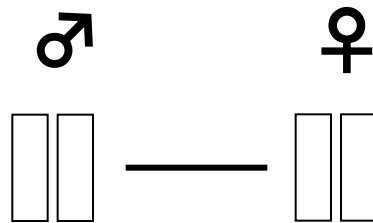
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HLA genes are inherited as sets (haplotypes)

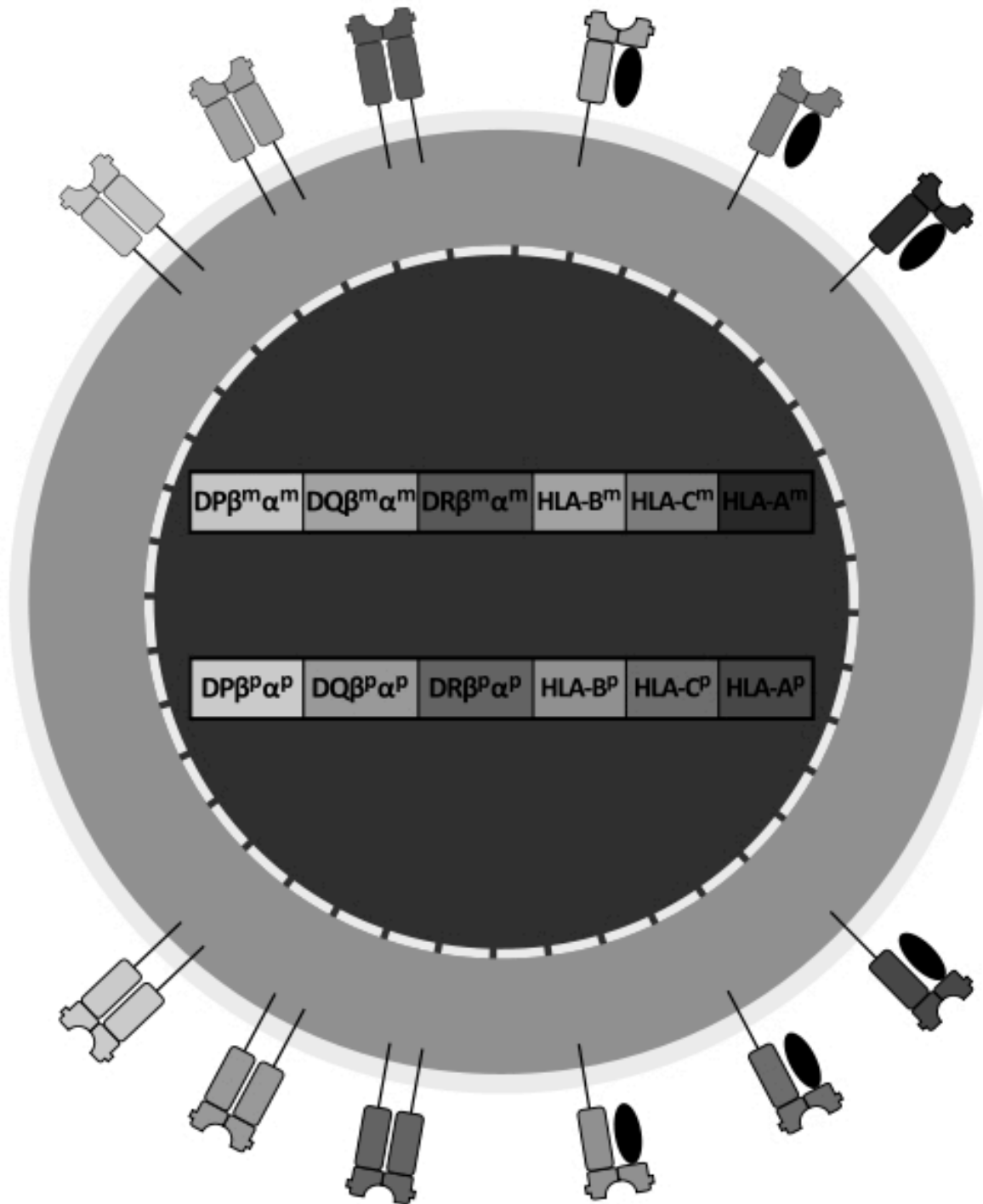


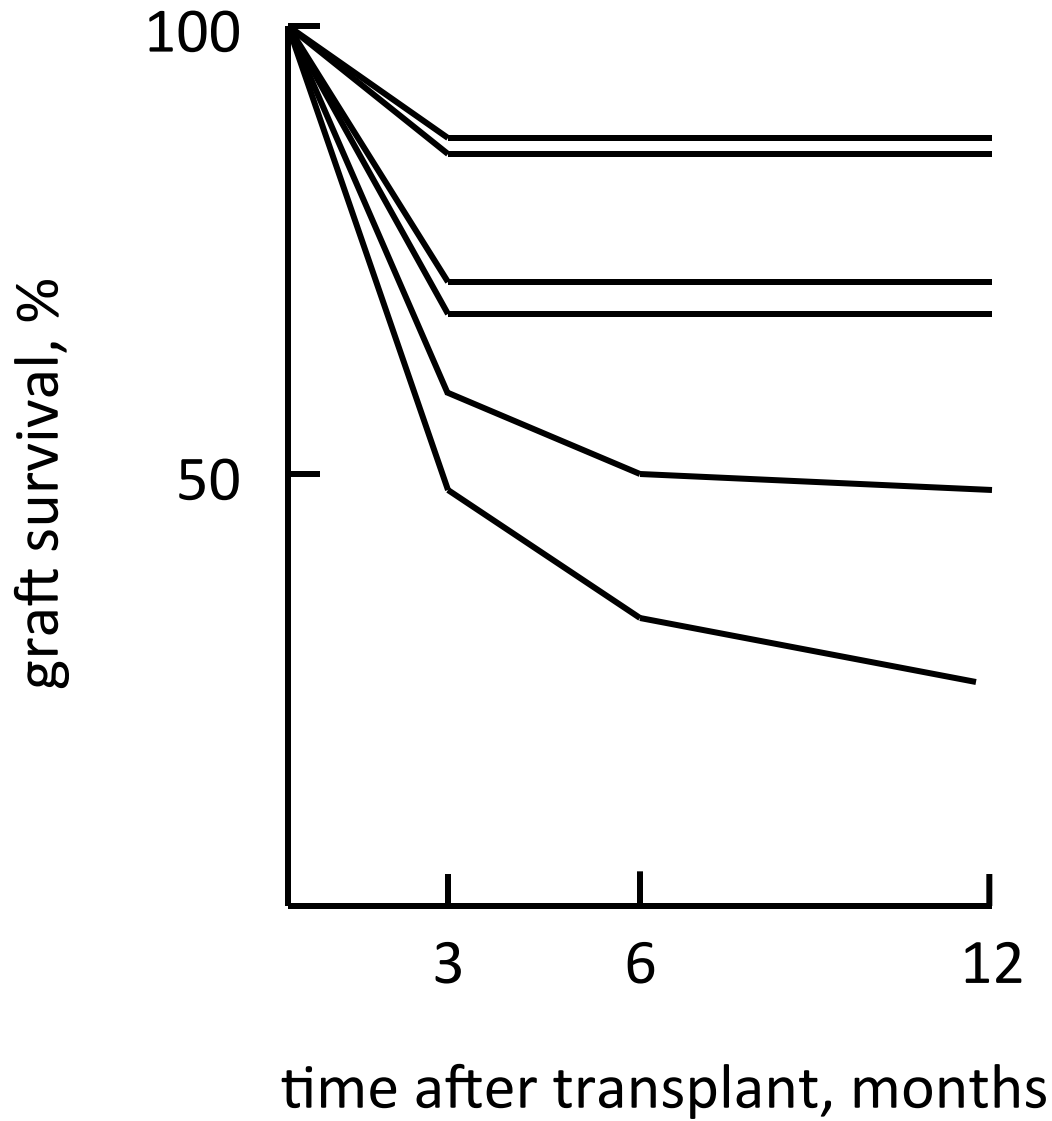
HLA genes are polymorphic



	A	B	C	DR	DQ	DP
Dad's HLA genes	1	7	w3	2	1	1
	2	8	w2	3	2	2
Mom's HLA genes	3	44	w4	4	1	3
	11	35	w1	7	3	4

HLA genes are
codominantly
expressed





number of mismatches

Class I	Class II
0	0
1 or 2	0
3 or 4	0
0	1 or 2
1 or 2	1 or 2
3 or 4	1 or 2