CTD Antibody and HLA Associations

by Todd Mollet, MD, and Adrienne Lam, MD

CONNECTIVE TISSUE DISEASE-SPECIFIC ANTIBODIES	
Lupus Erythematosus	
ANTIBODY	**ASSOCIATION**
C1q	Severe LE; urticarial vasculitis
dsDNA	Also referred to as nDNA (native DNA); High levels confirm diagnosis of SLE; Low levels seen in RA, Hashimoto's, Grave's, Waldenstrom's macroglobulinemia, MCT, SSc, liver disease, SjS; correlates with disease activity
Histone	Drug-induced LE if negative for other antiodies
Phospholipid	Primary APA syndrome; SLE (50% of patients); Drugs (cocaine, interferon α, procainamide, hydralazine, phenothiazines, quinine, quinidine, fansidar, phenytoin); chronic infections (syphilis, mononucleosis, TB, leprosy, leptospirosis, malaria, typhus, trypanosomiasis, schistosomiasis, filariasis, CMV, HIV, HCV)
Ro	SCLE; Neonatal LE
rRNP	CNS disease
Sm	Most specific antibody for SLE
ssDNA	Low diagnostic value; SLE in DLE patients; Linear morphea in children
U1RNP	MCTD; SLE
Dermatomyositis	
ANTIBODY	**ASSOCIATION**
155 kd/Se	Amyopathic DM; cancer
Jo-1 (anti-histidyl tRNA synthetase)	Interstitial lung disease; anti-synthetase syndrome; mechanic's hands
Mi-2	Skin involvement only; good prognosis
SRP	Fulminant DM; Cardiac involvement
Sjogren's Syndrome	
ANTIBODY	**ASSOCIATION**
αFodrin	
La (SS-B)	
Ro (SS-A)	Annular erythema of SjS
Systemic Sclerosis	
ANTIBODY	**ASSOCIATION**
Centromere	CREST
Fibrillin-1	Localized SSc
Scl-70 (anti-topoisomerase-I)	Diffuse SSc

DISEASE	ANTIBODY	TARGET
Wegener's granulomatosis	cANCA	Proteinase 3
Microangiopathic vasculitis	pANCA	Myeloperoxidase

ANA patterns		Target
SLE	Peripheral	DNA
SLE	Homogenous	DNA, histones
SSc, SLE	Nucleolar	RNA
CREST	Centromere	Kinetochore
MCTD, SLE, SSc, SjS	Speckled	Ribonucleoproteins

HLA ASSOCIATIONS	
DISEASE	**ASSOCIATED HLA(s)**
Abacavir induced hypersensitivity syndrome	- B*5701
Actinic prurigo	- DR4 (DRB1*0401), -DRB1*0407
Acute generalized erythematous pustulosis	- B5, -DR11 and -DQ3
Allopurinol induced SJS/TEN	
- Han Chinese	- B*5801
Alopecia Areata	
- all types	- HLA-DQB1*0301 (DQ7), HLA-DQB1*03 (DQ3), and HLA-DRB1*1104 (DR11)
- severe alopecia totalis/universalis	- DRB1*0401 (DR4) and HLA-DQB1*0301 (DQ7)
Behçet's disease	- B51
Bullous pemphigoid	
- Caucasians	- DQB1*0301
- Japanese	- DRB1*04, DRB1*1101 and DQB1*0302

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HLA ASSOCIATIONS	
DISEASE	**ASSOCIATED HLA(s)**
Carbamazepine induced SJS/TEN	
- Asians and East Indians	- B*1502
- Europeans	- A*3101
Chronic urticaria	- DR4, -DQ8
Dermatitis herpetiformis	- DQ2, -B8
Dermatomyositis	
- Juvenile	- DR3, -B8
- with anti-JO antibodies	- DR52
- with anti-Mi-2 antibodies	- DR7, -DRw53
- adults with dermatomyositis overlap	- B14, -B40
- Japanese with juvenile dermatomyositis	- DRB1*15021
Epidermolysis bullosa aquisita	
- Caucasians and African Americans	- DRB1*1501, -DR5
- Koreans	- DRB1*13
Erythema dyschromium perstans	
- Mexican patients	- DR4
Erythema multiforme	- DQw3, DRw53, and Aw33
Generalized granuloma annulare	- Bw35
Henoch-Schonlein Purpura	
- With renal disease	- B35
Juvenile idiopathic arthritis	
- Type II Oligo/pauciarticular arthritis	- B27
- Enthesitis-related arthritis	- B27
Leprosy	
- Lepromatous form	- DQ1
- Tuberculoid form	- DR2, -DR3
Lichen planus	
- Oral and cutaneous	- DR1
- Oral	
- English patients	- B27, -B51, -Bw57
- Japanese and Chinese patients	- DR9
- HCV Patients	- DR6
Lichen sclerosus	- DQ7, -DRB1*12
Mixed connective tissue disease	- DR4, -DR1, -DR2
Mucous membrane pemphigoid	- DQw7
Pemphigoid gestationis	- DR3, -DR4
Pemphigus vulgaris	
- Caucasians	- DRB1*0402, DRB1*1401 and DQB1*0302
- Japanese	- DRB1*14 and DQB1*0503
Psoriasis	
- Early onset	- Cw6 (also in late-onset), -DRB1*0701/2
Relapsing polychondritis	- DR4
- Negatively associated w/ organ involvement	- DR6
Rheumatoid Arthritis	- DR1, -DR4, -DRB1
Sacroiliitis	- B27
- Psoriasis	
- Crohns	
- UC	
- SAPHO	
- Reactive arthritis	
Sarcoidosis	- 1, -B8, -DR3, -DRB1, DQB1
Stevens- Johnson Syndrome	
- With ocular complications	- DQB1*0601
Still's disease, Adult-onset	- B14, -B17, -B18, -B35, -Bw35, -Cw4, -DR2, -DR7, -DR4, -Dw6
Subacute cutaneous lupus erythematosus	- B8, -DR3
Systemic lupus erythematosus	- A1, B8, DR3
Wegener's granulomatosis	- DPB1*0401

References:

1. Bolognia JL, Jorizzo JL, Schaffer JV. *Dermatology.* 3rd Edition. Elsevier; 2012.

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