

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 antibodies
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
α Fodrin	
La (SS-B)	
Ro (SS-A)	Annular erythema of SJS

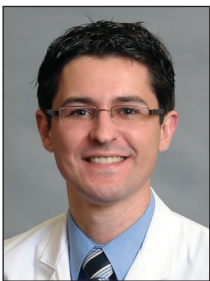
Systemic Sclerosis

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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CTD Antibody and HLA Associations (continued)

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HLA ASSOCIATIONS	
DISEASE	ASSOCIATED HLA(s)
Carbamazepine induced SJS/TEN - Asians and East Indians - Europeans	- B*1502 - A*3101
Chronic urticaria	- DR4, -DQ8
Dermatitis herpetiformis	- DQ2, -B8
Dermatomyositis - Juvenile - with anti-JO antibodies - with anti-Mi-2 antibodies - adults with dermatomyositis overlap - Japanese with juvenile dermatomyositis	- DR3, -B8 - DR52 - DR7, -DRw53 - B14, -B40 - DRB1*15021
Epidermolysis bullosa aquisita - Caucasians and African Americans - Koreans	- DRB1*1501, -DR5 - 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 - Enthesitis-related arthritis	- B27 - B27
Leprosy - Lepromatous form - Tuberculoid form	- DQ1 - DR2, -DR3
Lichen planus - Oral and cutaneous - Oral - English patients - Japanese and Chinese patients - HCV Patients	- DR1 - B27, -B51, -Bw57 - DR9 - DR6
Lichen sclerosus	- DQ7, -DRB1*12
Mixed connective tissue disease	- DR4, -DR1, -DR2
Mucous membrane pemphigoid	- DQw7
Pemphigoid gestationis	- DR3, -DR4
Pemphigus vulgaris - Caucasians - Japanese	- DRB1*0402, DRB1*1401 and DQB1*0302 - DRB1*14 and DQB1*0503
Psoriasis - Early onset	- Cw6 (also in late-onset), -DRB1*0701/2
Relapsing polychondritis - Negatively associated w/ organ involvement	- DR4 - DR6
Rheumatoid Arthritis	- DR1, -DR4, -DRB1
Sacroiliitis - Psoriasis - Crohns - UC - SAPHO - Reactive arthritis	- B27
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

Note:

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References:

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