# LOS NUEVOS CRITERIOS CLASIFICATORIOS DEL LES

**SLICC 2012** 







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# ES LUPUS ??

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- 1. Facial erythema (butterfly rash)
  Diffuse erythema, flat or raised, over the malar eminence(s) and/or bridge of the nose; may be unilateral.
- Discoid lupus
   Erythematous raised patches with adherent keratotic
   scaling and follicular plugging; atrophic scarring
   may occur in older lesions; may be present any where on the body.
- 3. Raynaud's phenomenon Requires a two-phase colour reaction, by patient's history or physician's observation.
- 4. Alopecia
  Rapid loss of a large amount of the scalp hair, by patient's history or physician's observation.
- 5. Photosensitivity
  Unusual skin reaction from exposure to sunlight,
  by patient's history or physician's observation.
- 6. Oral or nasopharyngeal ulceration
- 7. Arthritis without deformity
  One or more peripheral joints involved with any of
  the following in the absence of deformity;
  - (a) Pain on motion,
  - (b) Tenderness,
  - (c) Effusion or periarticular soft tissue swelling. (Peripheral joints are defined for this purpose as feet, ankles, knees, hips, shoulders, elbows, wrists, metacarpophalangeal, proximal interphalangeal, terminal interphalangeal, and temporomandibular joints)

#### 8. L.E. cells

Two or more classical L.E. cells seen on one occasion or one cell seen on two or more occasions, using an accepted published method.

- Chronic false-positive serological test for syphilis Known to be present for at least 6 months and confirmed by TPI or Reiter's tests.
- 10. Profuse proteinuria Greater than 3.5 g./day.
- 11. Cellular casts

May be red cell, haemoglobin, granular, tubular, or mixed.

- 12. One or both of the following:
  - (a) Pleuritis; good history of pleuritic pain; or rub heard by a physician; or x-ray evidence of both pleural thickening and fluid,
  - (b) Pericarditis, documented by EKG or rub.
- 13. One or both of the following:
  - (a) Psychosis.
  - (b) Convulsions, by patient's history or physician's observation in the absence of uraemia or offending drugs.
- 14. One or more of the following:
  - (a) Haemolytic anaemia,
  - (b) Leucopenia, WBC less than 4,000 per cu. mm. on two or more occasions,
  - (c) Thrombocytopenia, platelet count less than 100,000 per cu. mm.

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  eminence(s) and/or bridge of the nose; may be
  unilateral.
- Discoid lupus
   Erythematous raised patches with adherent keratotic scaling and follicular plugging; atrophic scarring may occur in older lesions; may be present anywhere on the body.
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Greater than 3.5 g./day.

11. Cellular casts

May be red cell, haemoglobin, granular, tubular, or mixed.

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## THE 1982 REVISED CRITERIA FOR CLASSIFICATION OF SYSTEMIC LUPUS ERYTHEMATOSUS

Criterion	Definition	
1. Malar rash	Fixed erythema, flat or raised, over the malar eminences, tending to spare the nasolabial folds	
2. Discoid rash	Erythematous raised patches with adherent keratotic scaling and follicular plugging; atrophic scarring may occur in older lesions	
3. Photosensitivity	Skin rash as a result of unusual reaction to sunlight, by patient history or physician observation	
4. Oral ulcers	Oral or nasopharyngeal ulceration, usually painless, observed by physician	
5. Arthritis	Nonerosive arthritis involving 2 or more peripheral joints, characterized by tenderness, swelling, or effusion	
6. Serositis	a) Pleuritisconvincing history of pleuritic pain or rubbing heard by a physician or evidence of pleural effusion     OR	
	b) Pericarditisdocumented by ECG or rub or evidence of pericardial effusion	
7. Renal disorder	a) Persistent proteinuria greater than 0.5 grams per day or grater than 3+ if quantitation not performed OR	
	b) Cellular castsmay be red cell, hemoglobin, granular, tubular, or mixed	
8. Neurologic disorder	a) Seizuresin the absence of offending drugs or known metabolic derangements; e.g., uremia, ketoacidosis, or electrolyte imbalance OR	
	b) Psychosisin the absence of offending drugs or known metabolic derangements, e.g., uremia, ketoacidosis, or electrolyte imbalance	
Hematologic disorder	a) Hemolytic anemiawith reticulocytosis	
	b) Leukopenialess than 4,000/mm<>3<> total on 2 or more occasions  OR	
	c) Lyphopenialess than 1,500/mm<>3<> on 2 or more occasions OR	
	d) Thrombocytopenialess than 100,000/mm<>3<> in the absence of offending drugs	
10. Immunologic disorder	a) Positive LE cell preparation	
	OR b) Anti-DNA: antibody to native DNA in abnormal titer OR	
	c) Anti-Sm: presence of antibody to Sm nuclear antigen  OR	
	A) False positive serologic test for syphilis known to be positive for at least 6 months and confirmed by <i>Treponema pallidum</i> immobilization or fluorescent treponemal antibody absorption test	
11. Antinuclear antibody	An abnormal titer of antinuclear antibody by immunofluorescence or an equivalent assay at any point in time and in the absence of drugs known to be associated with "drug-induced lupus" syndrome	

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8. Neurologic disorder	a) Seizuresin the absence of offending drugs of known metabolic derangements; e.g., uremia, ketoacidosis, or electrolyte imbalance OR b) Psychosisin the absence of offending drugs of known metabolic derangements, e.g., uremia, ketoacidosis, or electrolyte imbalance
9. Hematologic disorder	a) Hemolytic anemiawith reticulocytosis OR b) Leukopenialess than 4,000/mm<>3<> total on 2 or more occasions OR c) Lyphopenialess than 1,500/mm<>3<> on 2 or more occasions OR d) Thrombocytopenialess than 100,000/mm<>3<> in the absence of offending drugs
10. Immunologic disorder	a) Positive LE cell preparation OR b) Anti-DNA: antibody to native DNA in abnormal titer OR c) Anti-Sm: presence of antibody to Sm nuclear antigen OR d) False positive serologic test for syphilis known to be positive for at least 6 months and confirmed by Treponema pallidum immobilization or fluorescent treponemal antibody absorption test
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#### ACR criteria for the classification of systemic lupus erythematosus

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Malar rash	Fixed erythema, flat or raised, over the malar eminences, tending to spare the nasolabial folds	
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Oral ulcers	Oral or nasopharyngeal ulceration, usually painless, observed by a physician	
Arthritis	Nonerosive arthritis involving 2 or more peripheral joints, characterized by tenderness, swelling, or effusion	
Serositis	Pleuritis - convincing history of pleuritic pain or rub heard by a physician or evidence of pleural effusion <b>OR</b>	
	Pericarditis - documented by EKG, rub or evidence of pericardial effusion	
Renal disorder	Persistent proteinuria greater than 0.5 grams per day or greater than 3+ if quantitation not performed <b>OR</b>	
	Cellular casts - may be red cell, hemoglobin, granular, tubular, or mixed	
Neurologic disorder	Seizures <b>OR</b> psychosis - in the absence of offending drugs or known metabolic derangements (uremia, ketoacidosis, or electrolyte imbalance)	
Hematologic disorder	gic Hemolytic anemia - with reticulocytosis <b>OR</b>	
disorder	Leukopenia - less than 4000/mm <sup>3</sup> total on two or more occasions <b>OR</b>	
Lymphopenia - less than 1500/mm <sup>3</sup> on two or more occasions <b>O</b>		
	Thrombocytopenia - less than 100,000/mm <sup>3</sup> in the absence of offending drugs	
Immunologic	Anti-DNA - antibody to native DNA in abnormal titer <b>OR</b>	
disorders	Anti-Sm - presence of antibody to Sm nuclear antigen <b>OR</b>	
	Positive antiphospholipid antibody on:	
	1. an abnormal serum level of IgG or IgM anticardiolipin antibodies, or	
	a positive test result for lupus anticoagulant using a standard method, or	
	3. a false-positive test result for at least 6 months confirmed by Treponema pallidum immobilization or fluorescent treponemal antibody absorption test	
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Serositis	Pleuritis - convincing history of pleuritic pain or rub heard by a physician or evidence of pleural effusion <b>OR</b>	
	Pericarditis - documented by EKG. rub or evidence of pericardial effusion	

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	Lymphopenia - less than $1500 / \text{mm}^3$ on two or more occasions $\textbf{OR}$	
	Thrombocytopenia - less than 100,000/mm $^{\rm 3}$ in the absence of offending drugs	
Immunologic disorders	Anti-DNA - antibody to native DNA in abnormal titer <b>OR</b>	
disorders	Anti-Sm - presence of antibody to Sm nuclear antigen <b>OR</b>	
	Positive antiphospholipid antibody on:  1. an abnormal serum level of IgG or IgM anticardiolipin antibodies, or  2. a positive test result for lupus anticoagulant using a standard method, or  3. a false-positive test result for at least 6 months confirmed by Treponema pallidum immobilization or fluorescent treponemal antibody absorption test	
Antinuclear antibody	An abnormal titer of antinuclear antibody by immunofluorescence or an equivalent assay at any point in time and in the absence of drugs known to be associated with "drug-induced lupus" syndrome	

# PROBLEMAS DE LOS CRITERIOS ACR

Excesivo peso de la piel (3/11)

No es imprescindible la inmunología

Nefritis lúpica aislada

**Complemento** 



# Derivation and Validation of the Systemic Lupus International Collaborating Clinics Classification Criteria for Systemic Lupus Erythematosus

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#### ARTHRITIS & RHEUMATISM

## Deriva Systemic Lupus Classification Crite

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#### Table 3. Clinical and immunologic criteria used in the SLICC classification system\*

#### Clinical criteria

1. Acute cutaneous lupus, including:

Lupus malar rash (do not count if malar discoid)

Bullous lupus

Toxic epidermal necrolysis variant of SLE

Maculopapular lupus rash

Photosensitive lupus rash

in the absence of dermatomyositis

OR subacute cutaneous lupus (nonindurated psoriaform and/or annular polycyclic lesions that resolve without scarring, although occasionally with postinflammatory dyspigmentation or telangiectasias)

2. Chronic cutaneous lupus, including:

#### Classic discoid rash

Localized (above the neck)

Generalized (above and below the neck)

Hypertrophic (verrucous) lupus

Lupus panniculitis (profundus)

Mucosal lupus

Lupus erythematosus tumidus

Chillblains lupus

Discoid lupus/lichen planus overlap

3. Oral ulcers

Dalata

Buccal

Tongue

OR nasal ulcers

in the absence of other causes, such as vasculitis, Behçet's disease, infection (herpesvirus), inflammatory bowel disease, reactive arthritis, and acidic foods

4. Nonscarring alopecia (diffuse thinning or hair fragility with visible broken hairs)

in the absence of other causes such as alopecia areata, drugs, iron deficiency, and androgenic alopecia

5. Synovitis involving 2 or more joints, characterized by swelling or effusion

OR tenderness in 2 or more joints and at least 30 minutes of morning stiffness

Serositis

Typical pleurisy for more than 1 day

OR pleural effusions

OR pleural rub

Typical pericardial pain (pain with recumbency improved by sitting forward) for more than 1 day

OR pericardial effusion

OR pericardial rub

OR pericarditis by electrocardiography

in the absence of other causes, such as infection, uremia, and Dressler's pericarditis

7. Rena

Urine protein-to-creatinine ratio (or 24-hour urine protein) representing 500 mg protein/24 hours

OR red blood cell casts

 Neurologio Seizures

Psychosis

Sychosis

Mononeuritis multiplex

in the absence of other known causes such as primary vasculitis

Myelitis

Peripheral or cranial neuropathy

in the absence of other known causes such as primary vasculitis, infection, and diabetes mellitus

Acute confusional state

in the absence of other causes, including toxic/metabolic, uremia, drugs

9. Hemolytic anemia

10. Leukopenia (<4,000/mm3 at least once)

in the absence of other known causes such as Felty's syndrome, drugs, and portal hypertension

OR

Lymphopenia (<1,000/mm3 at least once)

in the absence of other known causes such as corticosteroids, drugs, and infection

11. Thrombocytopenia (<100,000/mm3) at least once

in the absence of other known causes such as drugs, portal hypertension, and thrombotic thrombocytopenic purpura Immunologic criteria

1. ANA level above laboratory reference range

2. Anti-dsDNA antibody level above laboratory reference range (or >2-fold the reference range if tested by ELISA)

3. Anti-Sm: presence of antibody to Sm nuclear antigen

4. Antiphospholipid antibody positivity as determined by any of the following:

Positive test result for lupus anticoagulant

False-positive test result for rapid plasma reagin

Medium- or high-titer anticardiolipin antibody level (IgA, IgG, or IgM)

Positive test result for anti-β<sub>2</sub>-glycoprotein I (IgA, IgG, or IgM)

5. Low complement

Low C3

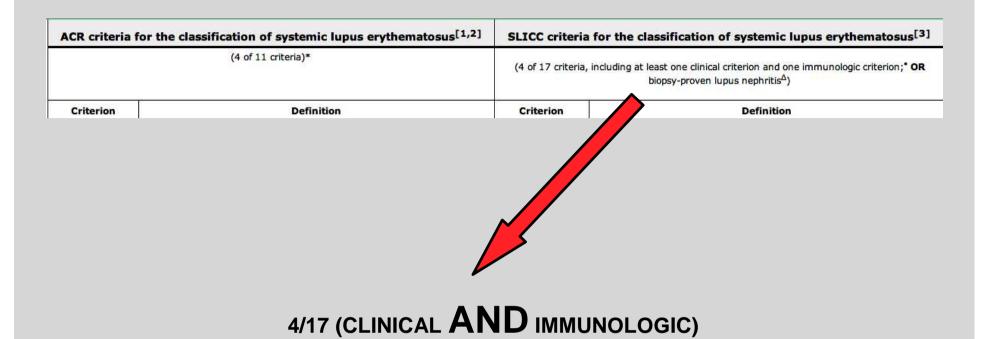
Low C4 Low CH50

6. Direct Coombs' test in the absence of hemolytic anemia

ACR criteria for the classification of systemic lupus erythematosus <sup>[1,2]</sup>		SLICC criteria for the classification of systemic lupus erythematosus <sup>[3]</sup>		
(4 of 11 criteria)*		(4 of 17 criteria, including at least one clinical criterion and one immunologic criterion;* <b>OR</b> biopsy-proven lupus nephritis <sup>Δ</sup> )		
Criterion Definition		Criterion Definition		
Malar rash	Fixed erythema, flat or raised, over the malar eminences, tending to spare the nasolabial folds	Acute cutaneous lupus	Lupus malar rash (do not count if malar discoid); bullous lupus; toxic epidermal necrolysis variant of SLE; maculopapular lupus rash;	
Photosensitivity	Skin rash as a result of unusual reaction to sunlight, by patient history or physician observation		photosensitive lupus rash (in the absence of dermatomyositis); <b>OR</b> subacute cutaneous lupus (nonindurated psoriaform and/or annular polycyclic lesions that resolve without scarring, although occasionally with postinflammatory dyspigmentation or telangiectasias)	
Discoid rash	Erythematosus raised patches with adherent keratotic scaling and follicular plugging; atrophic scarring may occur in older lesions	Chronic cutaneous lupus	Classic discoid rash; localized (above the neck); generalized (above and below the neck); hypertrophic (verrucous) lupus; lupus panniculitis (profundus); mucosal lupus; lupus erythematosus tumidus; chilblains lupus; <b>OR</b> discoid lupus/lichen planus overlap	
		Nonscarring alopecia	Diffuse thinning or hair fragility with visible broken hairs (in the absence of other causes such as alopecia areata, drugs, iron deficiency, and androgenic alopecia)	
Oral ulcers	Oral or nasopharyngeal ulceration, usually painless, observed by a physician	Oral or nasal ulcers	Palate, buccal, tongue, <b>OR</b> nasal ulcers (in the absence of other causes, such as vasculitis, Behçet's disease, infection (herpesvirus), inflammatory bowel disease, reactive arthritis, and acidic foods)	
Arthritis	Nonerosive arthritis involving 2 or more peripheral joints, characterized by tenderness, swelling, or effusion	Joint disease	Synovitis involving 2 or more joints, characterized by swelling or effusion <b>OR</b>	
			Tenderness in 2 or more joints and at least 30 minutes of morning stiffness	

ACR criteria for the classification of systemic lupus erythematosus <sup>[1,2]</sup>		SLICC criteria for the classification of systemic lupus erythematosus <sup>[3]</sup>		
(4 of 11 criteria)*		(4 of 17 criteria, including at least one clinical criterion and one immunologic criterion;* <b>OR</b> biopsy-proven lupus nephritis <sup>△</sup> )		
Criterion Definition		Criterion	Definition	
Serositis	Pleuritis - convincing history of pleuritic pain or rubbing heard by a physician or evidence of pleural effusion <b>OR</b>	Serositis	Typical pleurisy for more than 1 day, pleural effusions, or pleural rub, <b>OR</b>	
	Pericarditis - documented by EKG, rub, or evidence of pericardial effusion		Typical pericardial pain (pain with recumbency improved by sitting forward) for more than 1 day, pericardial effusion, pericardial rub, or pericarditis by electrocardiography in the absence of other causes, such as infection, uremia, and Dressler's syndrome	
Renal disorder	Persistent proteinuria greater than 500 mg/24 hours or greater than 3+ if quantitation not performed <b>OR</b>	Renal	Urine protein-to-creatinine ratio (or 24-hour urine protein) representing 500 mg protein/24 hours, <b>OR</b>	
	Cellular casts - may be red cell, hemoglobin, granular, tubular, or mixed		Red blood cell casts	
Neurologic disorder	Seizures <b>OR</b> psychosis - in the absence of offending drugs or known metabolic derangements (uremia, ketoacidosis, or electrolyte imbalance)	Neurologic	Seizures; psychosis; mononeuritis multiplex (in the absence of other knowledges such as primary vasculitis); myelitis; peripheral or cranial neuropathy (in the absence of other known causes such as primary vasculitis, infection, and diabetes mellitus); <b>OR</b> acute confusional state (in the absence of other causes, including toxic/metabolic, uremia, drugs)	
Hematologic	Hemolytic anemia - with reticulocytosis OR	Hemolytic anemia	Hemolytic anemia	
disorder	Leukopenia - less than 4000/mm <sup>3</sup> total on two or more occasions <b>OR</b> Lymphopenia - less than 1500/mm <sup>3</sup> on two or more occasions <b>OR</b>	Leukopenia or lymphopenia	Leukopenia (<4000/mm³ at least once) (in the absence of other known causes such as Felty's syndrome, drugs, and portal hypertension), <b>OR</b>	
	Thrombocytopenia - less than 100,000/mm³ (in the absence of offending drugs)		Lymphopenia (<1000/mm³ at least once) (in the absence of other known causes such as corticosteroids, drugs, and infection)	
		Thrombocytopenia	Thrombocytopenia (<100,000/mm³) at least once in the absence of other known causes such as drugs, portal hypertension, and thrombotic thrombocytopenic purpura	

ACR criteria for the classification of systemic lupus erythematosus <sup>[1,2]</sup> (4 of 11 criteria)*		SLICC criteria	a for the classification of systemic lupus erythematosus <sup>[3]</sup>
		(4 of 17 criteria, including at least one clinical criterion and one immunologic criterion; OR biopsy-proven lupus nephritis (4)	
Criterion Definition		Criterion	Definition
		,	Immunologic criteria
ANA	An abnormal titer of antinuclear antibody by immunofluorescence or an equivalent assay at any point in time and in the absence of drugs known to be associated with "drug-induced lupus" syndrome	ANA	ANA level above laboratory reference range
Immunologic disorders	Anti-DNA - antibody to native DNA in abnormal titer <b>OR</b>	Anti-dsDNA	Anti-dsDNA antibody level above laboratory reference range (or >twofold the reference range if tested by ELISA)
	Anti-Sm - presence of antibody to Sm nuclear antigen <b>OR</b>	Anti-Sm	Presence of antibody to Sm nuclear antigen
	Positive finding of antiphospholipid antibody based on an abnormal seru- level of IgG or IgM anticardiolipin antibodies, on a positive test result for lupus anticoagulant using a standard method, or on a false positive sero test for syphilis known to be positive for at least six months and confirm by Treponema pallidum immobilization or fluorescent treponemal antibo- absorption test	Antiphospholipid	Antiphospholipid antibody positivity as determined by any of the following: positive test result for lupus anticoagulant; false-positive test result for rapid plasma reagin; medium- or high-titer anticardiolipin antibody level (IgA, IgG, or IgM); or positive test result for anti-β2-glycoprotein I (IgA, IgG, or IgM)
		Low complement	Low C3; low C4; OR low CH50
		Direct Coombs' test	Direct Coombs' test in the absence of hemolytic anemia



OR

**BIOPSY-PROVEN LUPUS NEPHRITIS PLUS ANA / ANTI-DNA** 

#### 2003: Revisión de posibles variables

#### **DERIVATION SET: 716 PACIENTES**

SLE (n 293), RA (n 119), myositis (n 55), chronic cutaneous lupus (n 50), undifferentiated connective tissue disease (n 44), vasculitis (n 37), primary APS (n 33), scleroderma (n 28), fibromyalgia (n 25), Sjogren's syndrome (n 15), rosacea (n 8), psoriasis (n 7), sarcoidosis (n 1), and JIA (n 1).

#### **VALIDATION SET: 690 PACIENTES**

SLE (n 337), RA (n 118), undifferentiated connective tissue disease (n 89), primary APS (n 30), vasculitis (n 29), chronic cutaneous lupus (n 24), scleroderma (n 20), Sjogren's syndrome (n 15), myositis (n 14), psoriasis (n 8), fibromyalgia (n 4), alopecia areata (n 1), and sarcoidosis (n 1).

Identificación de variables y discusión del set final: n=17

## **Demographics**

AroseID:

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Date of Birth	08/10/1956	Gender	• Female	O Male	
Race	Black or African Amer	ican			

#### Paragraph describing all pertinent history and laboratory tests:

This Afro-Caribbean woman developed extensive vasculitic skin rash on legs, arms and abdomen and one year later pleuro-pericardial chest pains on inspiration. She has also suffered from recurrent anterior uveitis requiring steroid eye drops and arthralgias. Skin biopsy revealed a nodular vasculitis with septal panniculitis. She developed increasing arthritis in the wrists and proximal IP joints with tenderness and warmth. Subsequent flares of vasculitis rash were associated with arthritis (tender, swollen joints) affecting the knees and tenosynovitis in the hands. She did not do well with steroids, NSAIDs and dapsone so the latter was stopped and she was given azathioprine with little benefit and had a flare of pericarditis soon after this was started. She later became hypertensive. Urinalysis and renal function normal. She improved on MMF.

ANA, DNA, ENA, ANCA, anti-cardiolipin all negative. ACE, biochemistry, full blood count all normal. Lupus anticoagulant negative. Immunoglobulins normal.

C3 & C4 normal. CRP <5g/l.

Current ACR criteria: arthritis, serositis.

SLiCC clinical: synovitis, serositis

SLiCC immunologic: nil

#### 2003: Revisión de posibles variables

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SLE (n 293), RA (n 119), myositis (n 55), chronic cutaneous lupus (n 50), undifferentiated connective tissue disease (n 44), vasculitis (n 37), primary APS (n 33), scleroderma (n 28), fibromyalgia (n 25), Sjogren's syndrome (n 15), rosacea (n 8), psoriasis (n 7), sarcoidosis (n 1), and JIA (n 1).

Identificación de variables y discusión del set final: n=17

#### **VALIDATION SET: 690 PACIENTES**

SLE (n 337), RA (n 118), undifferentiated connective tissue disease (n 89), primary APS (n 30), vasculitis (n 29), chronic cutaneous lupus (n 24), scleroderma (n 20), Sjogren's syndrome (n 15), myositis (n 14), psoriasis (n 8), fibromyalgia (n 4), alopecia areata (n 1), and sarcoidosis (n 1).

590 (86%) con 80% de acuerdo en primera vuelta (12 miembros) 100 (14%) a segunda vuelta (33 miembros)

Table 4. Performance of the proposed SLICC criteria compared with the current ACR criteria in the derivation sample (n = 702 scenarios)\*

	Rule		
	1997 ACR criteria	SLICC criteria	
Sensitivity†	267/310 (86)	292/310 (94)	
Specificity:	365/392 (93)	361/392 (92)	
Misclassified cases	70	49	

Table 6. Performance of the proposed SLICC criteria compared with the current ACR criteria in the validation sample (n=690 scenarios)\*

	Rul	e
	1997 ACR criteria	SLICC criteria
Sensitivity†	290/349 (83)	340/349 (97)
Specificity:	326/341 (96)	288/341 (84)
к	0.79	0.82
Misclassified cases§	74	62











# Now What ?!

