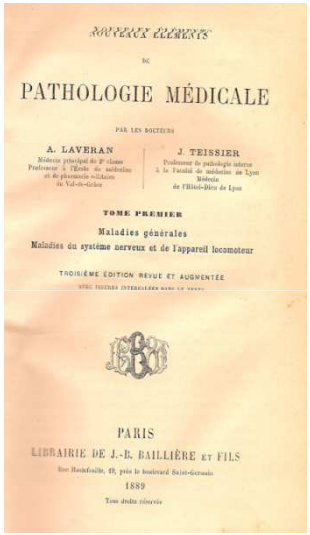
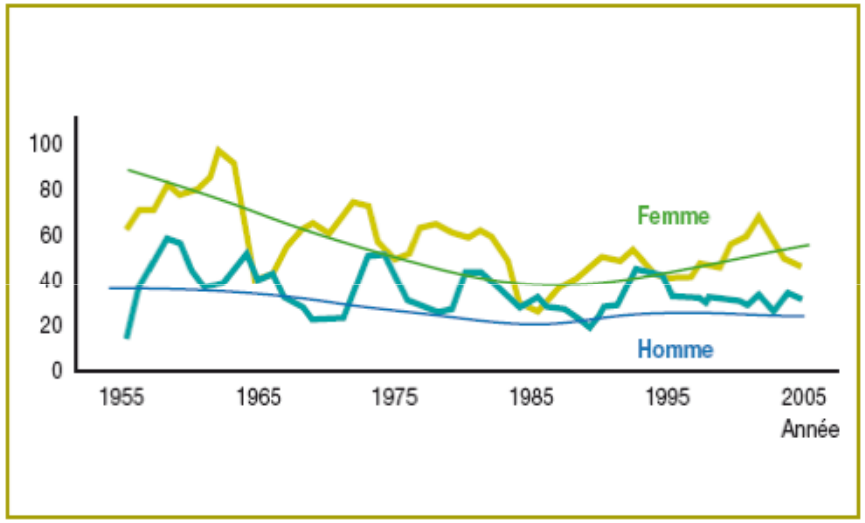
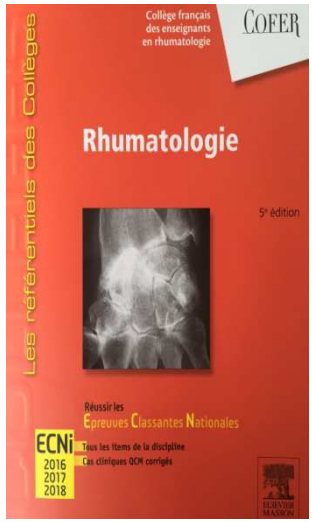
Changing patterns of inflammatory rheumatic diseases







Pr A Saraux, EA2216, INSERM ESPRI ERI29,
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CHU Brest

~~Breast~~

Brest

IRD encompass various concepts and the meaning of each subgroup is not the same everywere and everytime

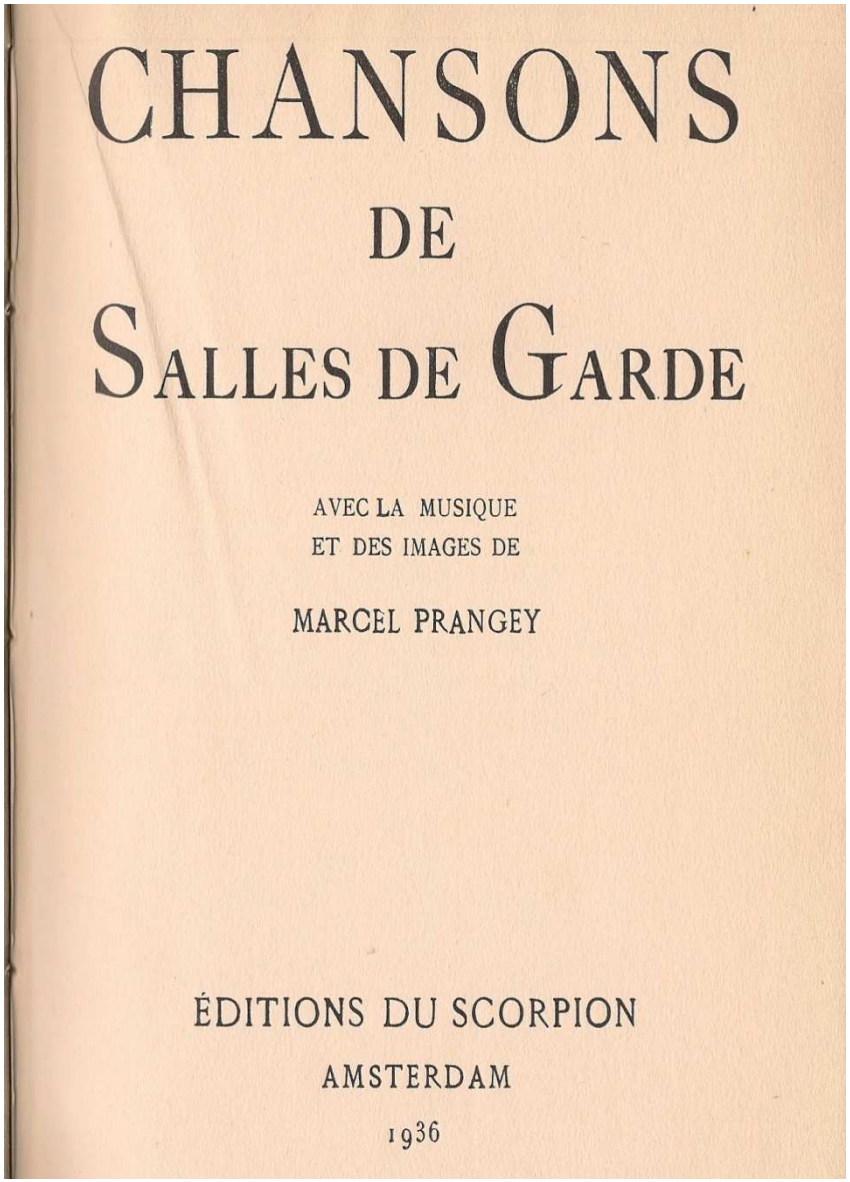
and both their names or their outlines are moving….

Ancestors medical doctors

Books on which they learnt

very nice dirty songs

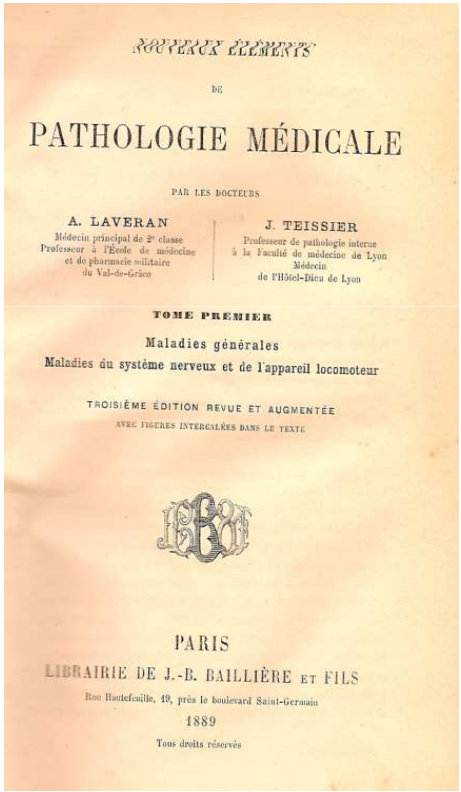
and drinking songs



What is an Inflammatory Rheumatic Disease? 1889-1891

- According to the books of the XIXth century used by the grand father of my grand mother, IRD were separated in three groups

  - Acute rheumatism
    - Rheumatic fever
    - Septic arthritis
    - Tuberculosis
  - Chronic rheumatism
  - nodosa rheumatism: small joints, elbow, shoulder
  - Simple chronic rheumatism : small joints, without deformity
  - Heberden's nodosity
  - Gout


NOUVEAUX ÉLÉMENTS

DE

PATHOLOGIE MÉDICALE

PAR LES DOCTEURS

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Médecin principal de 2e classe
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TOME PREMIER

Maladies générales
Maladies du système nerveux et de l'appareil locomoteur

TROISIÈME ÉDITION REVUE ET AUGMENTÉE
AVEC FIGURES INTERCALÉES DANS LE TEXTE

PARIS
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1889
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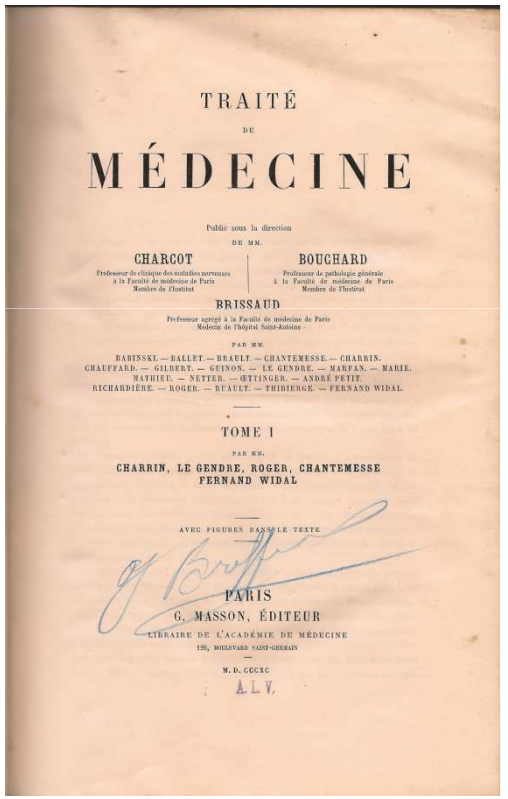
TRAITÉ

DE

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Publié sous la direction
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PAR MM.

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CHAUFFARD. — GILBERT. — GUINON. — LE GENDRE. — MARFAN. — MARIE.
MATHIEU. — NETTER. — OETTINGER. — ANDRÉ PETIT.
RICHARDIÈRE. — ROGER. — RUAULT. — THIBIERGE. — FERNAND WIDAL.

TOME I

PAR MM.

CHARRIN, LE GENDRE, ROGER, CHANTEMESSE
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AVEC FIGURES DANS LE TEXTE

PARIS
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120, BOULEVARD SAINT-GERMAIN
M. D. CCCXC

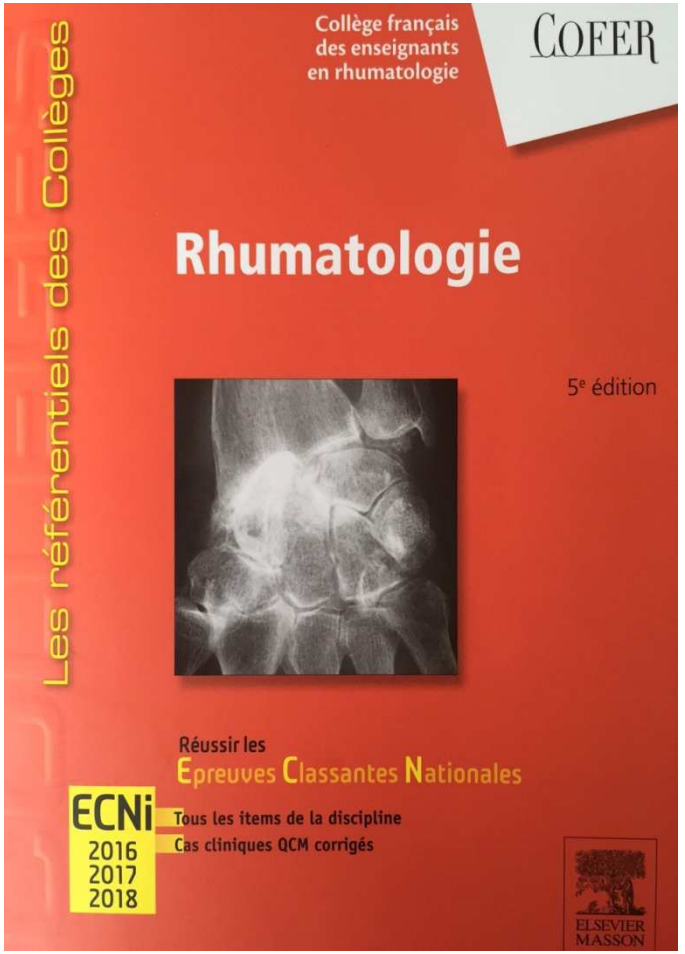
What is an Inflammatory Rheumatic Disease? 2016

It is really difficult to understand
how to connect our current concept

Infection
Crystal
Systemic
- Connective tissue diseases
- Vasculitis
- Other (autoinflammatory periodic fever, Still, sarcoidosis)

Primitive
- Rheumatoid arthritis
- Spondyloarthritis
- Polymyalgia rheumatica
- Idiopathic Juvenile Arthritis

but the identification of the etiology
Germs, crystal, autoantibodies
explain our better nosologic approach



The Nosology-Taxonomy of Recent-Onset Arthritis: The Experience of Early-Arthritis Clinics

Jean M. Berthelot, Alain Saraux, Yves Maugars, Alain Prost, and Paul Le Goff

Objective: To compare the conclusions of studies addressing the outcome of early-arthritis cohorts.

Methods: The methodologies of previous reports on early-arthritis cohorts were examined, and their results and conclusions were compared.

Results: Thirty-four reports on 23 cohorts of early arthritis were found. The methodology was poor in most studies, with numerous inclusion and exclusion biases, frequently short follow-up periods, and a lack of precision about the rationale for diagnosis. However, similar conclusions were reached on several points: a large number of cases of early arthritis remained undifferentiated and/or resolved spontaneously, about 80% of cases initially classified as undifferentiated or rheumatoid arthritis retained this diagnosis during follow-up, and the incidence of psoriatic arthritis in most studies was similar (2% to 4%). Conversely, there were striking discrepancies among studies concerning the frequency of crystal arthropathies (0% to 18%), spondyloarthropathy (1% to 33%) and rheumatoid arthritis (15% to 47%).

Conclusions: There appears to be a lack of agreement among researchers about the nosology and/or taxonomy of many cases of mild arthritis, despite the existence of classification criteria.

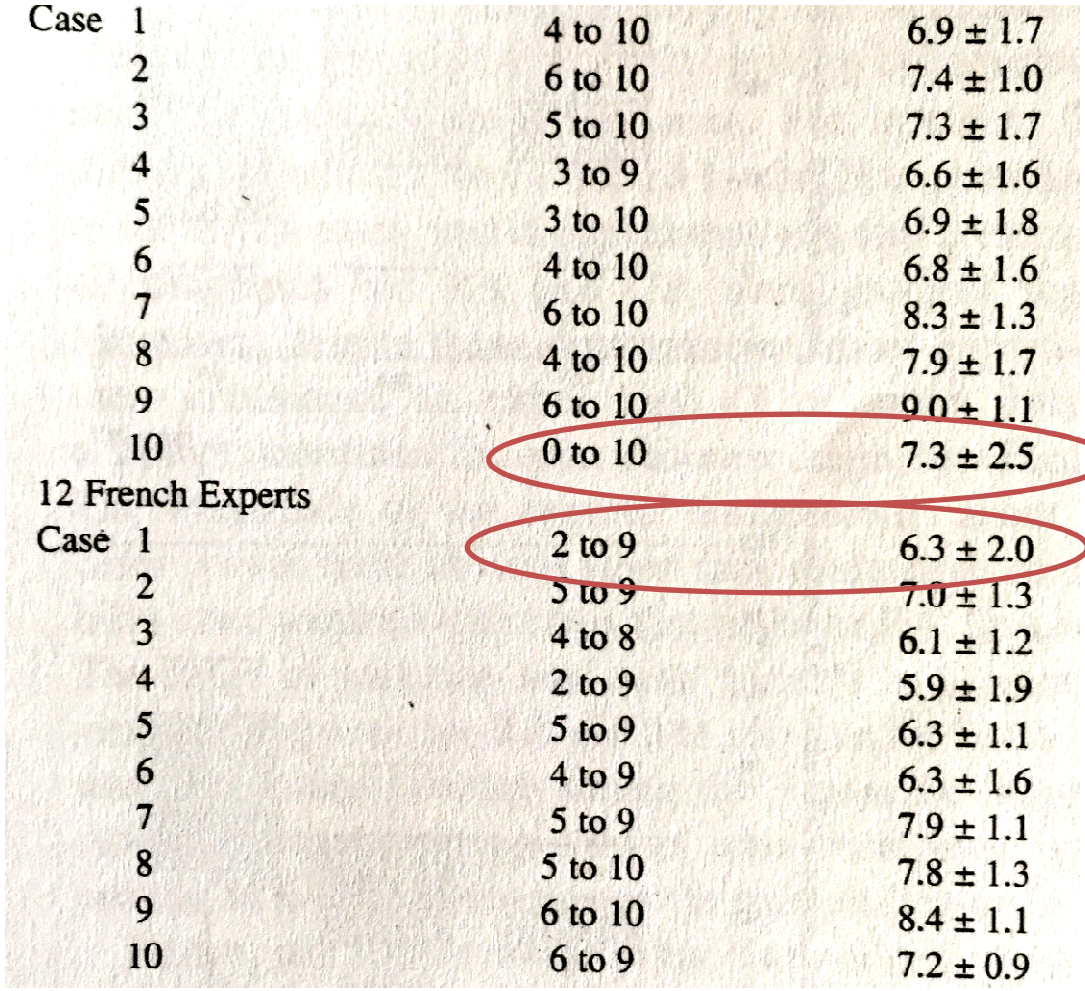
Relevance: Recognition of cultural bias in the diagnosis of early arthritis could be a prerequisite for the optimization of new sets of criteria for the diagnosis of early rheumatoid arthritis and spondyloarthropathy.

Semin Arthritis Rheum 30:354-365.

Certainty in diagnosis

To verify this fact, fifteen years ago we sent paper cases to panels of both international and french experts

We observed very important discordance in their confidence in the diagnosis



	Confidence	range	mean
Case	1	4 to 10	6.9 ± 1.7
	2	6 to 10	7.4 ± 1.0
	3	5 to 10	7.3 ± 1.7
	4	3 to 9	6.6 ± 1.6
	5	3 to 10	6.9 ± 1.8
	6	4 to 10	6.8 ± 1.6
	7	6 to 10	8.3 ± 1.3
	8	4 to 10	7.9 ± 1.7
	9	6 to 10	9.0 ± 1.1
	10	0 to 10	7.3 ± 2.5
12 French Experts			
Case	1	2 to 9	6.3 ± 2.0
	2	5 to 9	7.0 ± 1.3
	3	4 to 8	6.1 ± 1.2
	4	2 to 9	5.9 ± 1.9
	5	5 to 9	6.3 ± 1.1
	6	4 to 9	6.3 ± 1.6
	7	5 to 9	7.9 ± 1.1
	8	5 to 10	7.8 ± 1.3
	9	6 to 10	8.4 ± 1.1
	10	6 to 9	7.2 ± 0.9

Berthelot JM, J Rheumatol 2001, 28:975-81

Difference in understanding and application of 1987 ACR criteria for RA and 1991 ESSG criteria for SpA

Table I. Replies to questions about 1987 ACR criteria for RA and the 1991 ESSG criteria for SP.

	International	French	Total
1987 ACR criteria for RA			
Used the tree format in publications	5/16	5/16	10/32
Use the tree format in routine practice	2/16	4/16	6/32
Use the list format in routine practice	12/16	9/16	21/32
Criteria must be fulfilled at final examination	4/16	1/16	5/32
Criteria must have been present simultaneously	4/16	6/16	10/32
Criteria can be validated cumulatively	8/16	9/16	17/32
Duration of morning stiffness: until no more stiffness	2/16	4/16	6/32
Duration of morning stiffness: until maximal improvement	14/16	12/16	26/32
Tenosynovitis applies for soft tissue swelling	7/16	11/16	18/32
Bursitis applies for soft tissue swelling	0/16	3/16	3/32
Sausage-like swelling of toes applies for soft tissue swelling	7/16	7/16	14/32
Symmetrical involvement of one group of joints is sufficient	8/15	5/16	13/31
Symmetrical involvement of two groups of joints is required	7/15	4/16	11/31
Symmetrical involvement of three groups of joints is required	0/15	3/16	3/31
Symmetrical involvement of all groups of joints	0/15	4/16	4/31
Chondrolysis of wrist or fingers is considered as a criterion	3/16	10/16	13/32
Deviation of wrists or fingers is considered as a criterion	0/16	1/16	1/32
Erosion of other joints is considered as a criterion	12/16	13/16	25/32
Need for more precise definition of criteria	9/15	10/16	19/31
Need for precise recommendation for addition of criteria	6/15	9/16	15/31
Need for more subtle classification than RA or 'not-RA'	5/14	8/16	13/30
Need for removal of some criteria	5/15	9/16	14/31
Need for addition of new biological criteria	2/13	14/16	16/29
Need for new radiological criteria	4/14	15/16	19/30
Need for exclusion criteria	7/13	13/16	20/29

One explanation is that the use of criteria is not similar between experts

For example in RA, using the 1987 ACR criteria, some experts consider or not:
-criteria cumulatively and other simultaneously,
-tenosynovitis as soft tissue swelling
-that exclusion criteria are needed.

Berthelot JM, Clin Exp Rheumatol 2001, 28:975-81

Table 1. Office-based rheumatologist diagnoses at the last visit in 270 patients with arthritis*

Diagnosis	No. of patients
Rheumatoid arthritis	94
Rheumatoid arthritis plus spondylarthropathy	1
Fibromyalgia	1
Algodystrophy	1
Hemochromatosis	1
Osteoarthritis	3
Gouty arthritis	4
Chondrocalcinosis	7
Hydroxyapatite crystal–induced arthritis	4
Erythema nodosum–sarcoidosis	5
Septic arthritis	
Lyme disease	1
Neisseria gonorrhoeae	1
Viral infection	3
Spondylarthropathy	56
Still's disease	2
Sjögren's syndrome	6
Scleroderma	2
Polymyositis	4
Systemic lupus erythematosus	5
Remitting seronegative symmetrical synovitis with pitting edema	1
Giant cell arteritis–polymyalgia rheumatica	3
Polymyalgia rheumatica or rheumatoid arthritis (?)	1
Vasculitis	2
Lymphoma	1
Undifferentiated	61

* The office-based rheumatologists considered 19 of the 61 cases of undifferentiated arthritis to be "unlikely to be RA." ? = uncertain.

- in a cohort of early arthritis
- without diagnosis at inclusion

- all etiologies (infection, crystal, connective tissue disease and vasculitis) are possible but

- explaining that rheumatologist consider as useful a large panel of test for diagnosis, particularly in case without extra articular signs.

- RA, SpA, and undifferentiated arthritis are the 3 most common groups

Saraux A, Arthritis Rheum, 2001 Saraux A, arthritis Rheum 2002

Disease with and whitout criteria

Infection	Crystal	Systemic	Primitif
Acute rheumatic fever	gout	LED	RA
Septic arthritis	Calcium Pyrophosphate	pSS	SpA
	Hydroxyapatitis	SSc	PMR
		IIM	JIA
		Vasculitis	

•Criteria are necessary
- when we do not have tool to confirm the diagnosis with both sensitivity and specificity at 100% in routine practice
-to minimize discordance between clinician

• Finally we have criteria for a large majority of IRD but not for some other for which they are also needed

Do criteria change and why?

But these criteria change over the time for various reasons:

	Infection	*Crystal*	*Systemic*	*Primitif*
new mechanism				***Auto-inflammatory***
new tool	*Acute Rheumatic fever*	*Gout*		
discordance			*Sjogren*	
new treatment			*SLE*	
All these reasons				*PMR* *Vasculitis* *RA* *SpA*

The most amazing example is autoinflammatory periodic fever (familial Mediterranean fever; mevalonate kinase deficiency; TNF receptor-associated periodic fever syndrome; cryopyrin-associated periodic syndromes) for which we have the perfect gold standard (genetic mutation) using molecular analysis

Evidence-based provisional clinical classification criteria for autoinflammatory periodic fevers

Silvia Federici,[1] Maria Pia Sormani,[2] Seza Ozen,[3] Helen J Lachmann,[4] Gayane Amaryan,[5] Patricia Woo,[6] Isabelle Koné-Paut,[7] Natacha Dewarrat,[8] Luca Cantarini,[9] Antonella Insalaco,[10] Yosef Uziel,[11] Donato Rigante,[12] Pierre Quartier,[13] Erkan Demirkaya,[14] Troels Herlin,[15] Antonella Meini,[16] Giovanna Fabio,[17] Tilmann Kallinich,[18] Silvana Martino,[19] Aviel Yonatan Butbul,[20] Alma Olivieri,[21] Jasmin Kuemmerle-Deschner,[22] Benedicte Neven,[13] Anna Simon,[23] Huri Ozdogan,[24] Isabelle Touitou,[25] Joost Frenkel,[26] Michael Hofer,[8] Alberto Martini,[27] Nicolino Ruperto,[1] Marco Gattorno,[1] for the Paediatric Rheumatology International Trials Organisation (PRINTO) and Eurofever Project

The reference 'gold standard' group includes patients with FMF, TRAPS, CAPS or MKD with a confirmatory molecular analysis[14] defined as follows:

- FMF: two *MEFV* mutations, of which at least one is in exon 10[23];
- MKD: two *MVK* mutations with the exclusion of variants with an uncertain pathological role (such as S52N P165L, H20Q) (http://fmf.igh.cnrs.fr/infevers/)[23];
- TRAPS: heterozygous *TNFRSF1A* mutations with the exclusion of low-penetrance (such as R92Q or P46L) or uncertain mutations (http://fmf.igh.cnrs.fr/infevers/)[23];
- CAPS: heterozygous *NLRP3* mutations with the exclusion of low-penetrance variants (V198M), functional polymorphisms (Q703K) or variants with uncertain pathological role (http://fmf.igh.cnrs.fr/infevers/).[23]

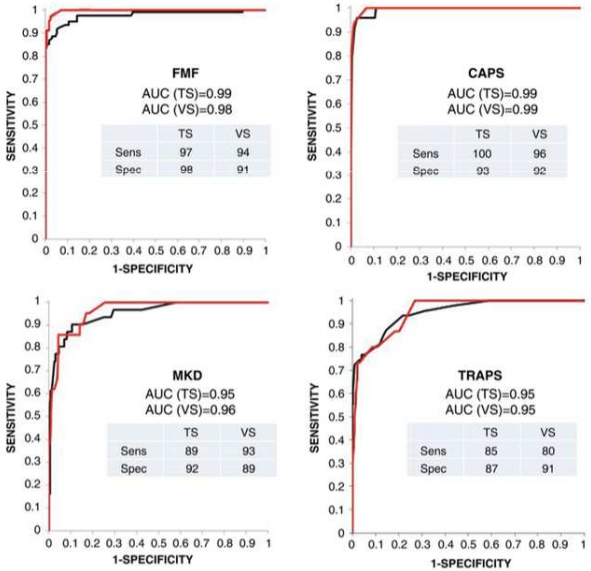


Figure 1 Receiver operating characteristic curves obtained for training (TS) and validation (VS) sets of gold standard patients, and the sensitivity (Sens) and specificity (Spec) of each classification criterion. AUC, area under the curve; CAPS, cryopyrin-associated periodic syndromes; FMF, familial Mediterranean fever; MKD, mevalonate kinase deficiency; TRAPS, receptor-associated periodic fever syndrome.

Federici S, *et al. Ann Rheum Dis* 2015;**74**:799–805.

Leading to these four classification criteria, with a scoring for presence or absence

Table 3 The Eurofever clinical diagnostic/classification criteria*

FMF		MKD		CAPS		TRAPS	
Presence	**Score**	**Presence**	**Score**	**Presence**	**Score**	**Presence**	**Score**
Duration of episodes < 2 days	9	Age at onset <2 years	10	Urticarial rash	25	Periorbital oedema	21
Chest pain	13	Aphthous stomatitis	11	Neurosensorial hearing loss	25	Duration of episodes >6 days	19
Abdominal pain	9	Generalised enlargement of lymph nodes or splenomegaly	8	Conjunctivitis	10	Migratory rash†	18
Eastern Mediterranean‡ ethnicity	22	Painful lymph nodes	13			Myalgia	6
North Mediterranean‡ ethnicity	7	Diarrhoea (sometimes/often)	20			Relatives affected	7
		Diarrhoea (always)	37				
Absence		**Absence**		**Absence**		**Absence**	
Aphthous stomatitis	9	Chest pain	11	Exudative pharyngitis	25	Vomiting	14
Urticarial rash	15			Abdominal pain	15	Aphthous stomatitis	15
Enlarged cervical lymph nodes	10						
Duration of episodes >6 days	13						
Cut-off	≥60	Cut-off	≥42	Cut-off	≥52	Cut-off	≥43

*The clinical features should be related to the typical fever episodes (ie, exclusion of intercurrent infection or other comorbidities). †Centrifugal migratory, erythematous patches most typically overlying a local area of myalgia, usually on the limbs or trunk.
‡Eastern Mediterranean: Turkish, Armenian, non-Ashkenazi Jewish, Arab. North Mediterranean: Italian, Spanish, Greek.
CAPS, cryopyrin-associated periodic syndromes; FMF, familial Mediterranean fever; MKD, mevalonate kinase deficiency; TRAPS, receptor-associated periodic fever syndrome.

Federici S, *et al*. *Ann Rheum Dis* 2015;**74**:799–805.

Do criteria change and why?

The identification of new tools to detect signs may also justify to build new criteria.

	Infection	*Crystal*	*Systemic*	*Primitif*
new mechanism				*Autoinflammatory*
new tool	***Acute Rheumatic fever***	***Gout***		
discordance			*Sjogren*	
new treatment			*SLE*	
All these reasons				*PMR* *RA* *SpA* *Vasculitis*

Acute Rheumatic Fever

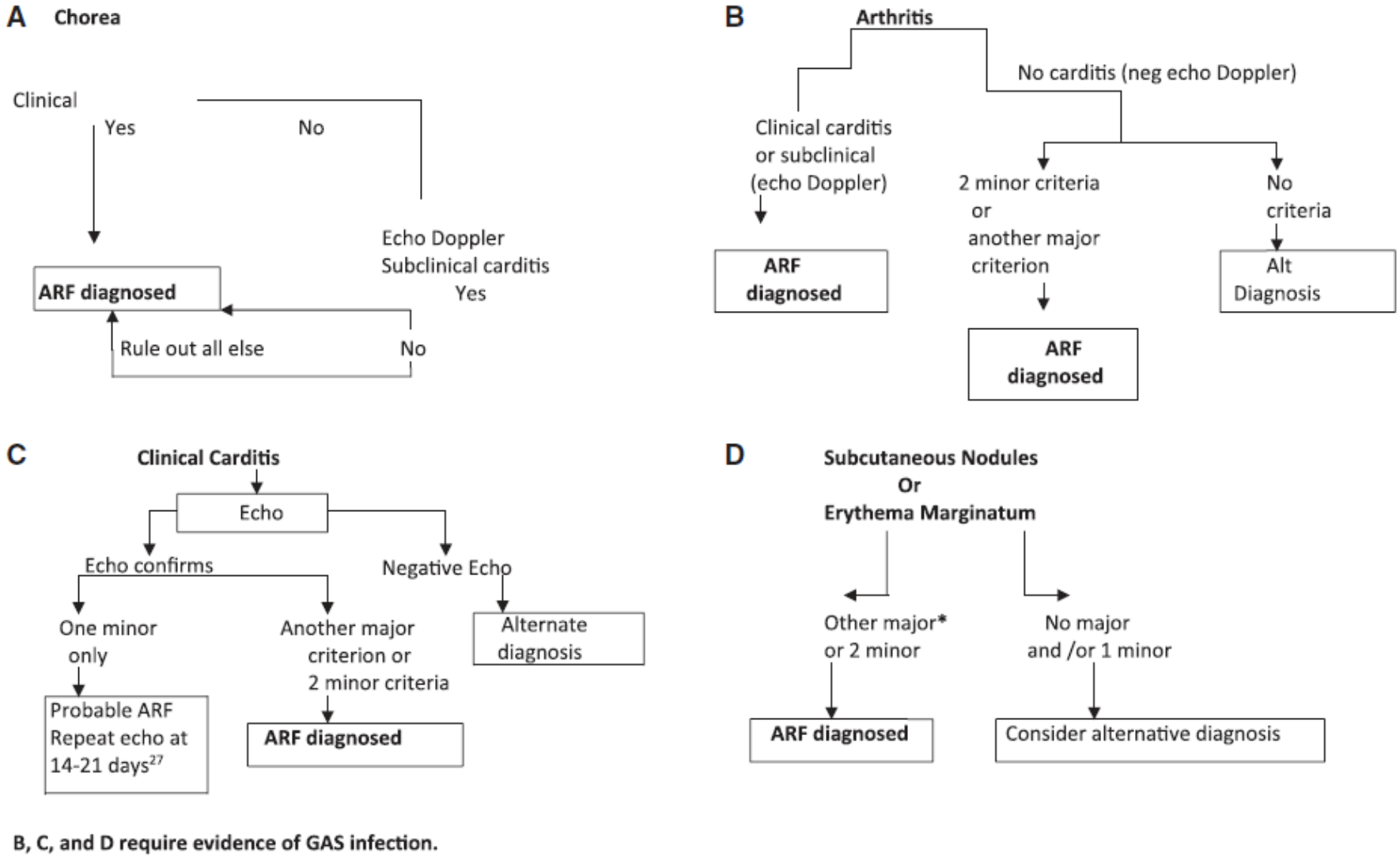
For example, in acute rheumatic fever, Jones criteria are may be improved

Table 7. Revised Jones Criteria

A. For all patient populations with evidence of preceding GAS infection	
Diagnosis: initial ARF	2 Major manifestations or 1 major plus 2 minor manifestations
Diagnosis: recurrent ARF	2 Major or 1 major and 2 minor or 3 minor
B. Major criteria	
Low-risk populations*	Moderate- and high-risk populations
Carditis† • Clinical and/or subclinical	Carditis • Clinical and/or subclinical
Arthritis • Polyarthritis only	Arthritis • Monoarthritis or polyarthritis • Polyarthralgia‡
Chorea	Chorea
Erythema marginatum	Erythema marginatum
Subcutaneous nodules	Subcutaneous nodules
C. Minor criteria	
Low-risk populations*	Moderate- and high-risk populations
Polyarthralgia	Monoarthralgia
Fever (≥38.5°C)	Fever (≥38°C)
ESR ≥60 mm in the first hour and/or CRP ≥3.0 mg/dL§	ESR ≥30 mm/h and/or CRP ≥3.0 mg/dL§
Prolonged PR interval, after accounting for age variability (unless carditis is a major criterion)	Prolonged PR interval, after accounting for age variability (unless carditis is a major criterion)

Circulation **May 19, 2015** **Gewitz et al** **Revised Jones Criteria for Acute Rheumatic Fever**

by adding echocardiography using algorythms according to the clinical pattern



B, C, and D require evidence of GAS infection.

Figure. Diagnosis strategy for acute rheumatic fever. *Subclinical carditis can be considered. Alt indicates alternative; ARF, acute rheumatic fever; echo, echocardiography; GAS, group A streptococcal; and neg, negative.

Another example is the classification of gout for which new criteria were published last year by the ACR/EULAR

2015 Gout classification criteria: an American College of Rheumatology/European League Against Rheumatism collaborative initiative

Tuhina Neogi,[1] Tim L Th A Jansen,[2,3] Nicola Dalbeth,[4] Jaap Fransen,[3] H Ralph Schumacher,[5] Dianne Berendsen,[3] Melanie Brown,[6] Hyon Choi,[1] N Lawrence Edwards,[7] Hein J E M Janssens,[3] Frédéric Lioté,[8] Raymond P Naden,[9] George Nuki,[10] Alexis Ogdie,[5] Fernando Perez-Ruiz,[11] Kenneth Saag,[12] Jasvinder A Singh,[13] John S Sundy,[14,15] Anne-Kathrin Tausche,[16] Janitzia Vaquez-Mellado,[17] Steven A Yarows,[18] William J Taylor[6]

Neogi T, *et al*. *Ann Rheum Dis* 2015;**74**:1789–1798.

Using new imaging features such as

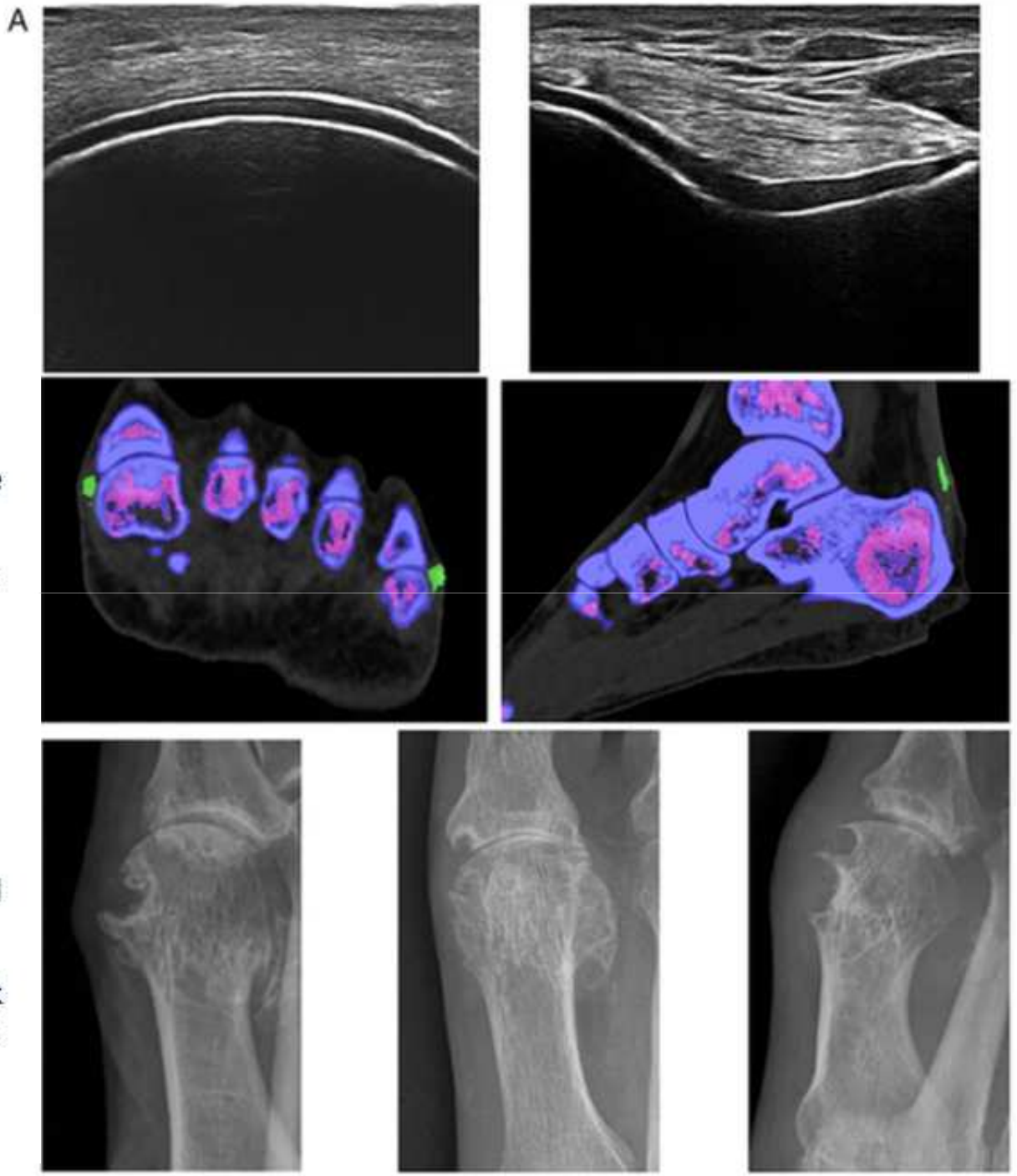


Figure 3 Examples of imaging features included in the classification criteria. (A) Double-contour sign seen on ultrasonography. Left panel shows a longitudinal ultrasound image of the femoral articular cartilage; right panel shows a transverse ultrasound image of the femoral articular cartilage. Both images show hyperechoic enhancement over the surface of the hyaline cartilage (images kindly provided by Dr Esperanza Naredo, Hospital Universitario Gregorio Marañon, Madrid, Spain). (B) Urate deposition seen on dual-energy CT. Left panel shows urate deposition at the first and fifth metatarsophalangeal joints; right panel shows urate deposition within the Achilles tendon. (C) Erosion, defined as a cortical break with sclerotic margin and overhanging edge, seen on conventional radiography of the first metatarsophalangeal joint.

Which may help clinicians when monosodic urate crystals are not detected in a symptomatic joint, bursa or tophus.

Table 2 The ACR/EULAR gout classification criteria*

	Categories	Score
Step 1: Entry criterion (only apply criteria below to those meeting this entry criterion)	At least 1 episode of swelling, pain, or tenderness in a peripheral joint or bursa	
Step 2: Sufficient criterion (if met, can classify as gout without applying criteria below)	Presence of MSU crystals in a symptomatic joint or bursa (ie, in synovial fluid) or tophus	
Step 3: Criteria (to be used if sufficient criterion not met)		
Clinical		
Pattern of joint/bursa involvement during symptomatic episode(s) ever	Ankle *or* mid-foot (as part of monoarticular or oligoarticular episode without involvement of the first metatarsophalangeal joint	1
	Involvement of the first metatarsophalangeal joint (as part of monoarticular or oligoarticular episode)	2
Characteristics of symptomatic episode(s) ever		
▶ Erythema overlying affected joint (patient-reported or physician-observed)	One characteristic	1
▶ Can't bear touch or pressure to affected joint	Two characteristics	2
▶ Great difficulty with walking or inability to use affected joint	Three characteristics	3
Time course of episode(s) ever		
Presence (ever) of ≥2, irrespective of anti-inflammatory treatment:		
▶ Time to maximal pain <24 h	One typical episode	1
▶ Resolution of symptoms in ≤14 days	Recurrent typical episodes	2
▶ Complete resolution (to baseline level) between symptomatic episodes		
Clinical evidence of tophus		
Draining or chalk-like subcutaneous nodule under transparent skin, often with overlying vascularity, located in typical locations: joints, ears, olecranon bursae, finger pads, tendons (eg, Achilles)	Present	4
Laboratory		
Serum urate: Measured by the uricase method. Ideally should be scored at a time when the patient was not receiving urate-lowering treatment and it was >4 weeks from the start of an episode (ie, during the intercritical period); *if* practicable, retest under those conditions. The highest value irrespective of timing should be scored	<4 mg/dL (<0.24 mmol/L)†	−4
	6–<8 mg/dL (0.36–<0.48 mmol/L)	2
	8–<10 mg/dL (0.48–<0.60 mmol/L)	3
	≥10 mg/dL (≥0.60 mmol/L)	4
Synovial fluid analysis of a symptomatic (ever) joint or bursa (should be assessed by a trained observer)‡	MSU negative	−2
Imaging§		
Imaging evidence of urate deposition in symptomatic (ever) joint or bursa: ultrasound evidence of double-contour sign¶ *or* DECT demonstrating urate deposition**	Present (either modality)	4
Imaging evidence of gout-related joint damage: conventional radiography of the hands and/or feet demonstrates at least 1 erosion††	Present	4

Do criteria change and why?

new criteria are also needed in case of discordance

	Infection	*Crystal*	*Systemic*	*Primitif*
new mechanism				*Autoinflammatory*
new tool	*Acute Rheumatic fever*	*Gout*		
discordance			***Sjogren***	
new treatment			*SLE*	
All these reasons				*PMR* *RA* *SpA* *Vasculitis*

Level of agreement between 2002 American–European Consensus Group and 2012 American College of Rheumatology classification criteria for Sjögren's syndrome and reasons for discrepancies

Divi Cornec[1,2], Alain Saraux[1,2], Béatrice Cochener[3], Jacques-Olivier Pers[2,4], Sandrine Jousse-Joulin[1,2], Yves Renaudineau[2,5], Thierry Marhadour[1] and Valérie Devauchelle-Pensec[1,2,6*]

Table 1 Pragmatic AECG [1] and ACR [2] classification criteria for Sjögren's syndrome

	Pragmatic 2002 AECG criteria	2012 ACR criteria
Items	1. Ocular dryness symptoms	1. Positive anti-SSA or anti-SSB antibodies or positive rheumatoid factor plus ANA ≥1:320
	2. Oral dryness symptoms	2. Focus score ≥1 focus/4 mm^2 on minor salivary gland biopsy
	3. Ocular signs: Schirmer's test ≤5 mm/5 minutes	3. Keratoconjunctivitis sicca with ocular staining score ≥3
	4. Focus score ≥1 focus/4 mm^2 on minor salivary gland biopsy	
	5. Salivary gland involvement: unstimulated whole salivary flow ≤0.1 ml/minute	
	6. Positive anti-SSA or anti-SSB antibodies	
Rules for classification	Presence of any four of the six items with at least item 4 or 6, or presence of any three of the four objective items (items 3, 4, 5 and 6)	In a patient with suspected Sjögren's syndrome, any two of the three items

There was two distincts criteria published by ACR/EULAR in 2002 and ACR ten years later

Level of agreement between 2002 American–European Consensus Group and 2012 American College of Rheumatology classification criteria for Sjögren's syndrome and reasons for discrepancies

Divi Cornec[1,2], Alain Saraux[1,2], Béatrice Cochener[3], Jacques-Olivier Pers[2,4], Sandrine Jousse-Joulin[1,2], Yves Renaudineau[2,5], Thierry Marhadour[1] and Valérie Devauchelle-Pensec[1,2,6*]

Both sets	ACR set only	AECG set only	Neither set
(*n* = 27)	(*n* = 8)	(*n* = 15)	(*n* = 55)

With discordance between them

Justiying new criteria

Cornec *et al. Arthritis Research & Therapy* 2014, **16**:R74

ACR/EULAR

The principle is based on five objective items, and a total score ≥ 4, derived from the sum of the weights assigned to each positive test/item

Item	Weight / Score
LSG with FLS and FS ≥ 1[3]	3
Anti-SSA (Ro) +	3
OSS ≥ 5 (or VB ≥ 4) on at least one eye[4]	1
Schirmer ≤ 5 mm/5min on at least one eye	1
UWS[5] flow rate ≤ 0.1 ml/min	1

Shiboski C, submitted

but ultrasonography of the major salivary glands have been developed in recent years, and should be soonly included into classification criteria.

- Parotid: transversal plane
- Parotid: longitudinal plane
- Submandibular

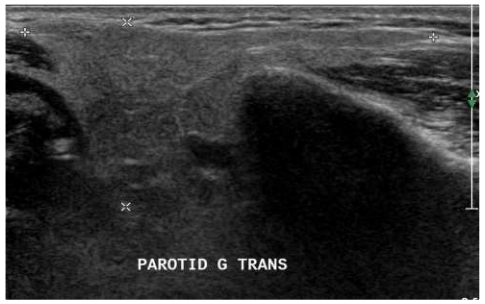
ARTHRITIS & RHEUMATISM
Vol. 65, No. 1, January 2013, pp 216–225
DOI 10.1002/art.37698

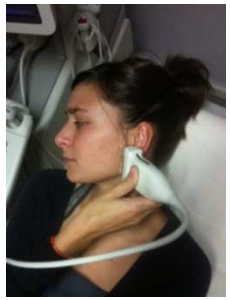
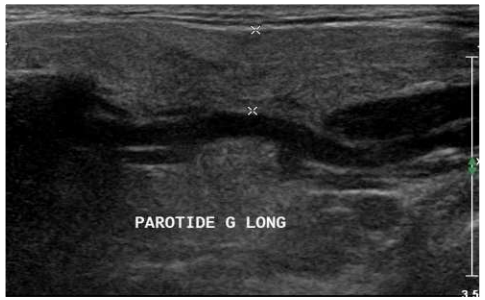
Contribution of Salivary Gland Ultrasonography to the Diagnosis of Sjögren's Syndrome

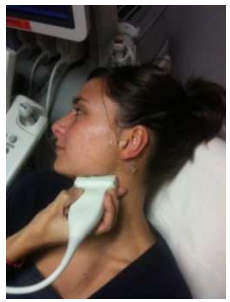
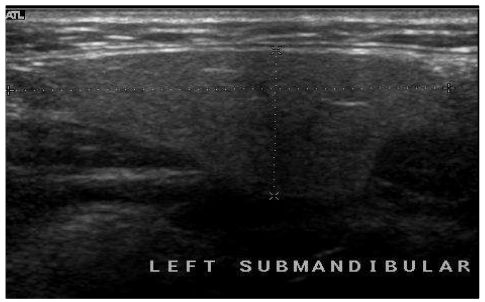
Toward New Diagnostic Criteria?

Divi Cornec,[1] Sandrine Jousse-Joulin,[1] Jacques-Olivier Pers,[1] Thierry Marhadour,[1] Béatrice Cochener,[2] Sylvie Boisramé-Gastrin,[3] Emmanuel Nowak,[4] Pierre Youinou,[1] Alain Saraux,[1] and Valérie Devauchelle-Pensec[1]





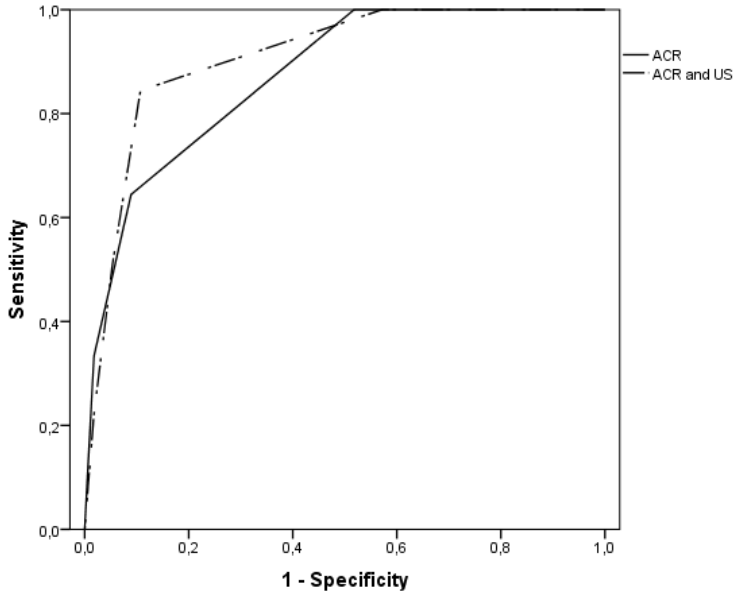




RHEUMATOLOGY 273, 294

Concise report doi:10.1093/rheumatology/keu037

Salivary gland ultrasonography improves the diagnostic performance of the 2012 American College of Rheumatology classification criteria for Sjögren's syndrome

Divi Cornec[1,2], Sandrine Jousse-Joulin[1,2], Thierry Marhadour[1], Jacques-Olivier Pers[2,3], Sylvie Boisramé-Gastrin[3], Yves Renaudineau[2,4], Alain Saraux[1,2] and Valérie Devauchelle-Pensec[1,2]



	Se	Sp	PPV	NPV
ACR criteria	64.4	91.1	85.3	76.1
ACR criteria + SGUS	84.4	89.3	86.7	87.7

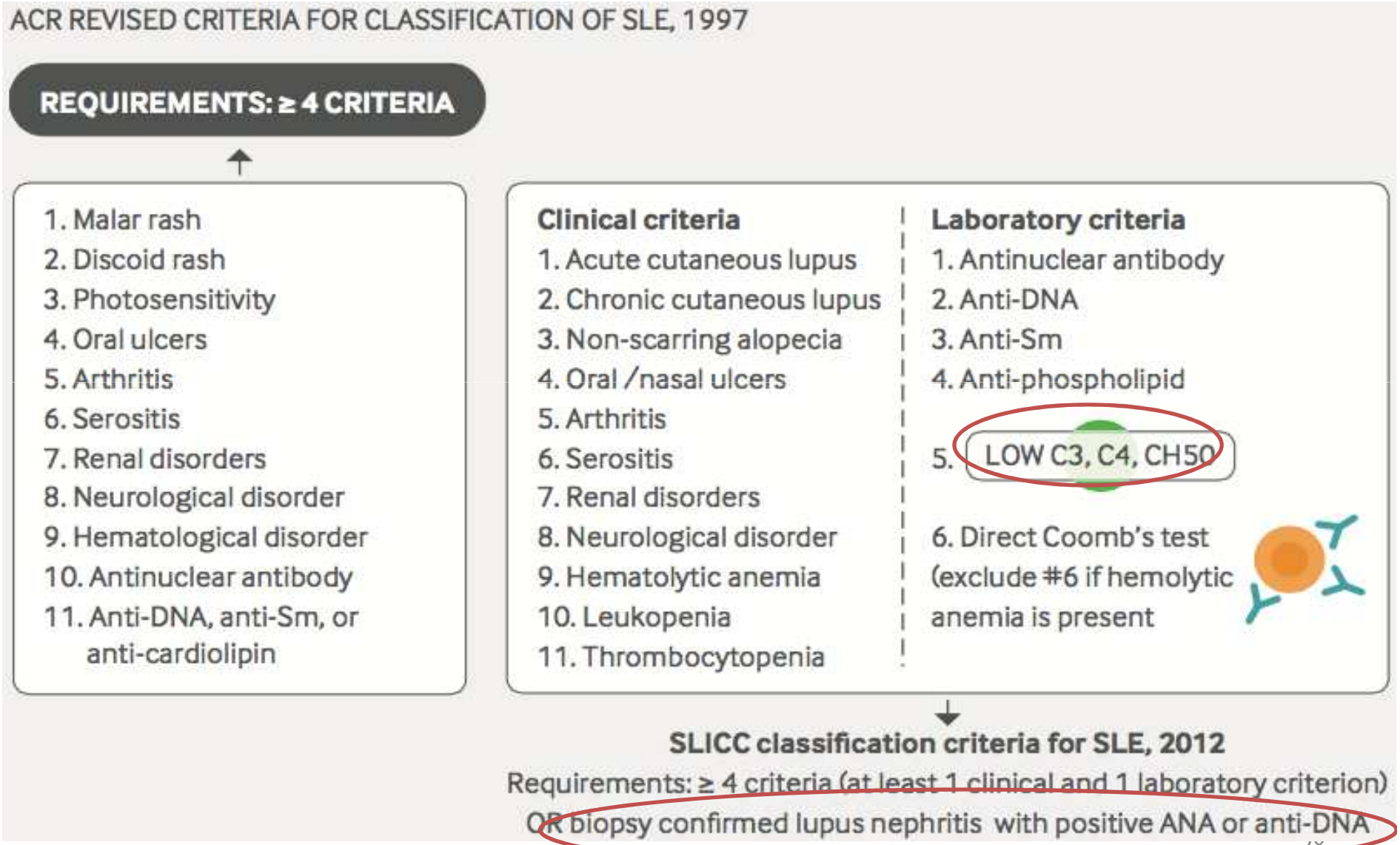
US clearly improve previous criteria.

Do criteria change and why?

Another reason to change criteria may be the development of new treatment

	Infection	*Crystal*	*Systemic*	*Primitif*
new mechanism				*Autoinflammatory*
new tool	*Acute Rheumatic fever*	*Gout*		
discordance			*Sjogren*	
new treatment			***SLE***	
All these reasons				*PMR* *RA* *SpA* *Vasculitis*

The SLICC criteria increase the sensitivity and allow the inclusion of a larger population



Do criteria change and why?

But finally, all these reasons explain the evolution of criteria in PMR, RA, SpA and vasculitis

	Infection	*Crystal*	*Systemic*	*Primitif*
new mechanism				*Autoinflammatory*
new tool	Acute Rheumatic fever	*Gout*		
discordance			*Sjogren*	*PMR*
new treatment			*SLE*	
All these reasons				***PMR*** ***RA*** ***SpA*** ***Vasculitis***

Polymyalgia rheumatica

There was discordance between previous criteria

Criteria for diagnosis of polymyalgia rheumatica suggested by various authors.

	Healey 1984	Chuang et al. 1982	Jones and Hazleman 1981	Bird et al. 1979	Hamrin 1972
Age	>50 years	>50		>65 years	>50 years
Onset				<2 weeks	
Duration		>1 month	>2 months		>2 months
Area of pain	neck, shoulder, or pelvic girdle	1. neck or torso, 2. shoulders or arms, 3. hips or thighs (at least 2 of 3)	shoulder and pelvic girdle	bilateral shoulder pain and stiffness	neck, shoulder or pelvic girdle (at least 2 of 3)
Morning stiffness	>1 h	>30 min.	present	>1 h	
Tenderness				upper arms	
Systemic symptoms				depression, weight loss	present
Erythrocyte sedimentation rate (ESR)	elevated	>40 mm/h[a]	>30 mm/h, or C-reactive protein >6 mg/l	>40 mm/h	>50 mm/h
Response to glucocorticoids	rapid, to 20 mg or less		prompt and dramatic		
Requirements for diagnosis	age must be > 50, plus 3 of the other criteria	all criteria	all criteria	if any 3 criteria present – sensitivity 92% specificity 80%	criteria of age, pain, and ESR are obligatory

Journal of Autoimmunity 48-49 (2014) 76–78

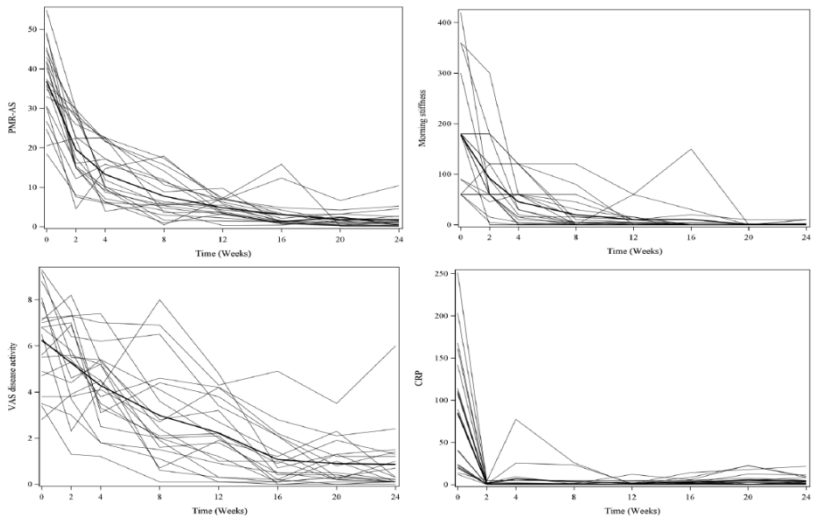
Polymyalgia rheumatica

And new tools (US, PET) and new treatments (anti-IL6)

Efficacy of first-line tocilizumab therapy in early polymyalgia rheumatica: a prospective longitudinal study

Valérie Devauchelle-Pensec[1,3], Jean Marie Berthelot[2], Divi Cornec[1,3], Yves Renaudineau[3] Thierry Marhadour[1], Sandrine Jousse-Joulin[1,3], Solène Querellou[4], Florent Garrigues[5], Michel De Bandt[6], Maelenn Gouillou[7], Alain Saraux[1,3]

doi:10.1136/annrheumdis-2015-208742



Eur J Nucl Med Mol Imaging
DOI 10.1007/s00259-015-3287-z

ORIGINAL ARTICLE

Value of ^{18}F-FDG PET/CT for therapeutic assessment of patients with polymyalgia rheumatica receiving tocilizumab as first-line treatment

X. Palard-Novello[1] · S. Querellou[1,5] · M. Gouillou[2] · A. Saraux[3,6] · T. Marhadour[3] · F. Garrigues[4] · R. Abgral[1,5] · P. Y. Salaün[1,5] · V. Devauchelle-Pensec[3,6]

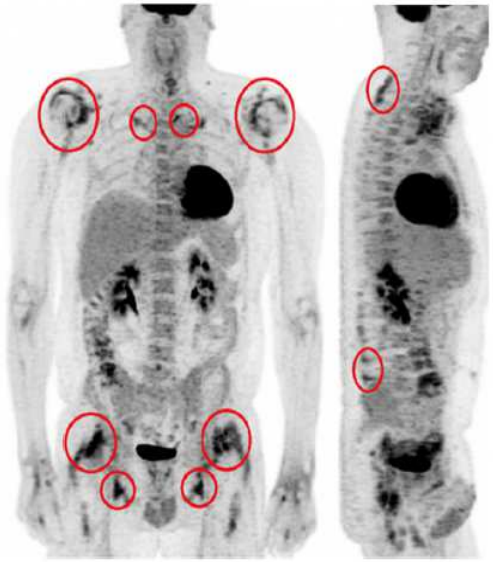


Fig. 1 Maximum intensity projection ^{18}F-FDG PET/CT images. SUVmax measurements were obtained in the ten regions of interest (*red circles*)

These new criteria require

- Age older than 50 years,
- Shoulder pain
- Inflammation

≥4 points

- Morning stiffness
- Hip pain
- Absence of RF or ACCP
- Absence of other joint

or ≥ 5 points

- Using ultrasound

	Points
Clinical criteria for scoring algorithm*	
Morning stiffness lasting more than 45 min	2
Hip pain or restricted range of motion	1
Absence of rheumatoid factor and antibody to cyclic citrullinated peptide	2
Absence of other joint involvement	1
Ultrasound criteria for scoring algorithm*	
At least one shoulder with subdeltoid bursitis, biceps tenosynovitis, or glenohumeral synovitis; and at least one hip with synovitis or trochanteric bursitis	1
Both shoulders with subdeltoid bursitis, biceps tenosynovitis, or glenohumeral synovitis	1

Required criteria: age 50 years or older, bilateral shoulder pain, and abnormal ESR, C-reactive protein, or both. *With only clinical criteria, a score of ≥4 had a sensitivity of 68% and specificity of 78% for discriminating polymyalgia rheumatica from comparison patients. With a combination of clinical criteria and ultrasound criteria, a score of ≥5 had a sensitivity of 66% and specificity of 81% for discriminating patients with the disorder from comparison patients.

Table: **European League Against Rheumatism and American College of Rheumatology provisional criteria for classification of polymyalgia rheumatica[50,51]**

Dejaco C, Singh YP, Perel P, et al. Ann Rheum Dis. 2015;74:1799-807

Tanaz A Kermani, Kenneth J Warrington **Lancet 2013; 381: 63–72**

THE AMERICAN RHEUMATISM ASSOCIATION 1987 REVISED CRITERIA FOR THE CLASSIFICATION OF RHEUMATOID ARTHRITIS

FRANK C. ARNETT, STEVEN M. EDWORTHY, DANIEL A. BLOCH, DENNIS J. McSHANE, JAMES F. FRIES, NORMAN S. COOPER, LOUIS A. HEALEY, STEPHEN R. KAPLAN, MATTHEW H. LIANG, HARVINDER S. LUTHRA, THOMAS A. MEDSGER, Jr., DONALD M. MITCHELL, DAVID H. NEUSTADT, ROBERT S. PINALS, JANE G. SCHALLER, JOHN T. SHARP, RONALD L. WILDER, and GENE G. HUNDER

Table 5. The 1987 revised criteria for the classification of rheumatoid arthritis (traditional format)*

Criterion	Definition
1. Morning stiffness	Morning stiffness in and around the joints, lasting at least 1 hour before maximal improvement
2. Arthritis of 3 or more joint areas	At least 3 joint areas simultaneously have had soft tissue swelling or fluid (not bony overgrowth alone) observed by a physician. The 14 possible areas are right or left PIP, MCP, wrist, elbow, knee, ankle, and MTP joints
3. Arthritis of hand joints	At least 1 area swollen (as defined above) in a wrist, MCP, or PIP joint
4. Symmetric arthritis	Simultaneous involvement of the same joint areas (as defined in 2) on both sides of the body (bilateral involvement of PIPs, MCPs, or MTPs is acceptable without absolute symmetry)
5. Rheumatoid nodules	Subcutaneous nodules, over bony prominences, or extensor surfaces, or in juxtaarticular regions, observed by a physician
6. Serum rheumatoid factor	Demonstration of abnormal amounts of serum rheumatoid factor by any method for which the result has been positive in <5% of normal control subjects
7. Radiographic changes	Radiographic changes typical of rheumatoid arthritis on posteroanterior hand and wrist radiographs, which must include erosions or unequivocal bony decalcification localized in or most marked adjacent to the involved joints (osteoarthritis changes alone do not qualify)

In early RA, the earlier use of methotrexate and the need of criteria to introduce it, the identification of anti CCP, the development of biologics explained that 1987 ACR criteria became old

Ability of the American College of Rheumatology 1987 Criteria to Predict Rheumatoid Arthritis in Patients With Early Arthritis and Classification of These Patients Two Years Later

Alain Saraux,[1] Jean M. Berthelot,[2] Gérard Chalès,[3] Catherine Le Henaff,[4] Jean B. Thorel,[4] Sylvie Hoang,[4] Isabelle Valls,[1] Valérie Devauchelle,[1] Antoine Martin,[4] Dominique Baron,[1] Yvon Pennec,[1] Estelle Botton,[1] Jean Y. Mary,[5] Paul Le Goff,[1] and Pierre Youinou[1]

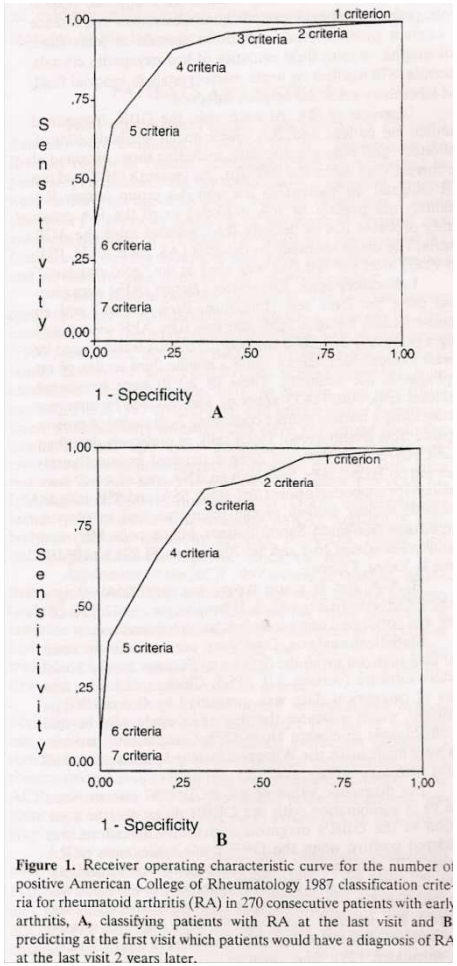
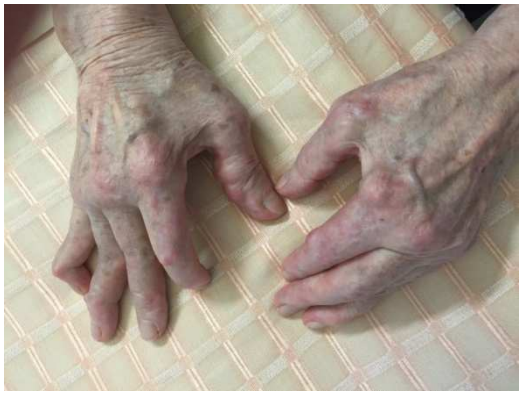
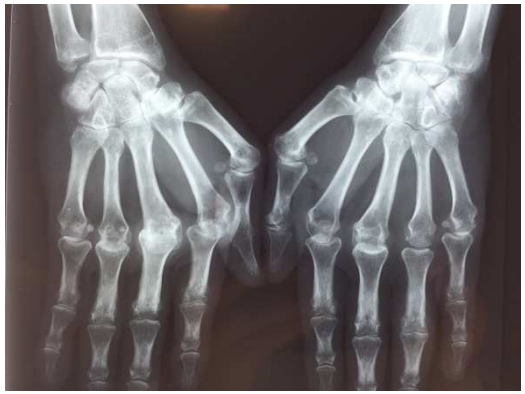


Figure 1. Receiver operating characteristic curve for the number of positive American College of Rheumatology 1987 classification criteria for rheumatoid arthritis (RA) in 270 consecutive patients with early arthritis, A, classifying patients with RA at the last visit and B predicting at the first visit which patients would have a diagnosis of RA at the last visit 2 years later.

Nodules and radiographics changes are too rare in early arthritis





Low classification value at inclusion

Saraux A, Arthritis Rheum, 2001 Saraux A, arthritis Rheum 2002

Anti-CCP Revised Criteria for the Classification of Rheumatoid Arthritis

Katherine P. Liao, **Kerri L. Batra**, **Lori Chibnik**, **Peter H. Schur**, and **Karen H. Costenbader**
Division of Rheumatology, Allergy and Immunology, Department of Medicine, Brigham and Women's Hospital, Boston, MA

anti CCP are useful

Criteria sets investigated for classification of RA

	1987 ACR Criteria	1987 ACR Criteria + anti-CCP	CCP 7 Criteria	CCP 6 Criteria
1. morning stiffness > 1 hr*	✓	✓	✓	✓
2. arthritis ≥ 3 joints*	✓	✓	✓	✓
3. hand arthritis*	✓	✓	✓	✓
4. symmetric arthritis*	✓	✓	✓	✓
5. rheumatoid nodules	✓	✓		
6. RF +	✓	✓	✓	✓
7. radiographic changes	✓	✓	✓	
8. anti-CCP +		✓	✓	✓
# criteria required	≥4 out of 7	≥4 out of 8	≥3 out of 7	≥3 out of 6

*Arthritis symptoms ≥ 6 weeks

Criteria	Sensitivity (%)	Specificity (%)
All subjects, n = 248		
1987 ACR Criteria	51	91
1987 ACR Criteria + Anti-CCP	55	91
CCP 7 Criteria	77	79
CCP 6 Criteria	74	81
Subjects with arthritis symptoms ≤ 6 months, n = 66		
1987 ACR Criteria	25	86
1987 ACR Criteria + Anti-CCP	44	86
CCP 7 Criteria	63	72
CCP 6 Criteria	63	72
Subjects with arthritis symptoms > 6 months, n = 182		
1987 ACR Criteria	58	93
1987 ACR Criteria + Anti-CCP	58	93
CCP 7 Criteria	81	82
CCP 6 Criteria	77	85

Liao KP, Ann Rheum Dis 2008, 67:1557-61

So, the 2010 ACR/EULAR criteria were built and their originality is that both synovitis and exclusion criteria on the basis of the opinion of clinician are mandatory

Table 2. The 2010 ACR/EULAR classification criteria and the scoring system for rheumatoid arthritis.

Target population to be tested 1. Patients who have at least one joint with definitive clinical synovitis (swelling) 2. Patients with synovitis not better explained by other disease	
A. Joint involvement	**Score**
1 large joint	0
2–10 large joints	1
1–3 small joints (with or without involvement of large joints)	2
4–10 small joints (with or without involvement of large joints)	3
> 10 joints (at least 1 small joint)	5
B. Serology (at least one test is needed for classification)	
Negative RF and negative ACPA	0
Low-positive RF or low-positive ACPA	2
High-positive RF or high-positive ACPA	3
C. Acute phase reactants	
(at least one test result is needed for classification)	
Normal CRP and ESR	0
Abnormal CRP or abnormal ESR	1
D. Duration of symptoms	
< 6 weeks	0
≥ 6 weeks	1

we observed that the difference between all previous set of classification criteria are very low in the cohort of early arthritis in Brittany if we apply these exclusion criteria.

Diagnostic Accuracy of ACR/EULAR 2010 Criteria for Rheumatoid Arthritis in a 2-Year Cohort

SOPHIE VARACHE, DIVI CORNEC, JOHANNE MORVAN, VALÉRIE DEVAUCHELLE-PENSEC, JEAN-MARIE BERTHELOT, CATHERINE LE HENAFF-BOURHIS, SYLVIE HOANG, JEAN-BAPTISTE THOREL, ANTOINE MARTIN, GÉRARD CHALÈS, EMMANUEL NOWAK, SANDRINE JOUSSE-JOULIN, PIERRE YOUINOU, and ALAIN SARAUX

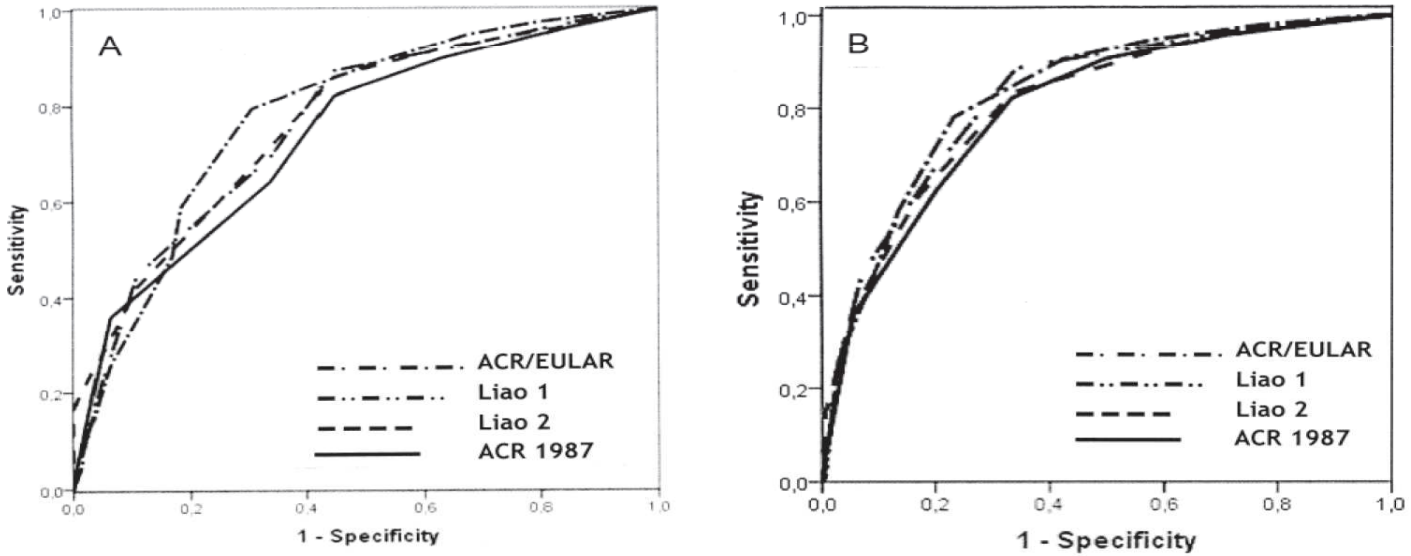


Figure 2. Receiver operating characteristic curve of the criteria sets in the subgroup meeting the 3 conditions for ACR/EULAR scoring (synovitis, no better alternative diagnosis, and no typical erosions; panel A; n = 143) and in the overall population (panel B; n = 270).

The Journal of Rheumatology 2011; 38:7; doi:10.3899/jrheum.101227

and it was confirmed on an independent cohort

Potential Classification Criteria for Rheumatoid Arthritis After Two Years: Results From a French Multicenter Cohort

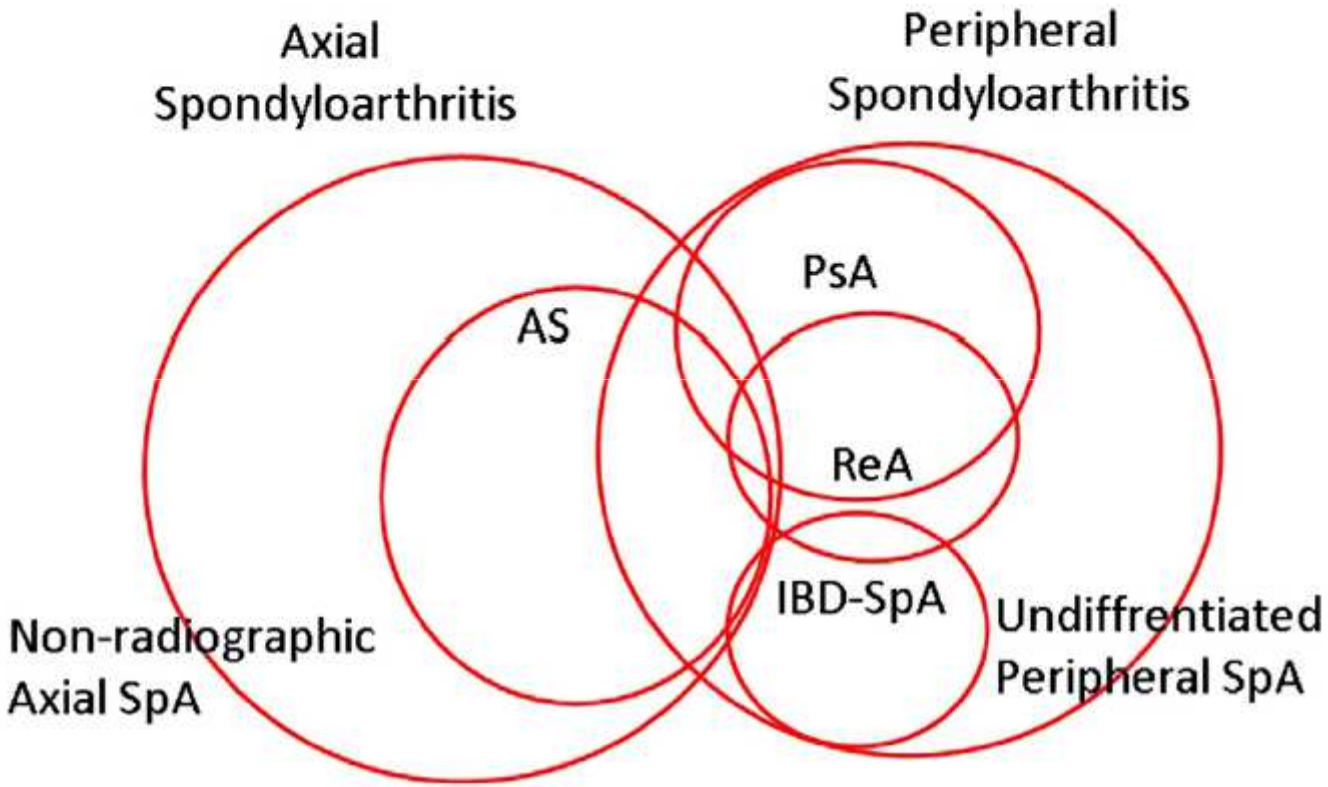
ALAIN SARAUX,[1] GABRIEL J. TOBÓN,[2] MATHILDE BENHAMOU,[3] VALÉRIE DEVAUCHELLE-PENSEC,[1] MAXIME DOUGADOS,[4] XAVIER MARIETTE,[5] FRANCIS BERENBAUM,[6] GILLES CHIOCCHIA,[4] ANNE-CHRISTINE RAT,[7] THIERRY SCHAEVERBEKE,[8] NATHALIE RINCHEVAL,[9] OLIVIER MEYER,[10] BRUNO FAUTREL,[3] AND BERNARD COMBE[10]

Table 2. Agreement among all definitions of RA*

	1987 ACR criteria	Rheumatologist diagnosis of RA with >50.0% certainty	No better alternative diagnosis with >50.0% certainty	Persistent disease†	Joint erosion
ACR/EULAR 2010					
Present at any time‡	0.43 (0.39, 0.47)	0.38 (0.34, 0.42)	0.32 (0.27, 0.37)	0.32 (0.27, 0.37)	0.09 (0.02, 0.11)
Present at last visit§	0.39 (0.35, 0.43)	0.71 (0.68, 0.74)	0.81 (0.78, 0.84)	0.38 (0.34, 0.42)	0.16 (0.13, 0.19)
1987 ACR criteria		0.40 (0.32, 0.48)	0.32 (0.23, 0.41)	0.28 (0.20, 0.37)	0.09 (0.06, 0.12)
Rheumatologist diagnosis of RA with >50.0% certainty			0.69 (0.63, 0.76)	0.46 (0.38, 0.54)	0.16 (0.13, 0.20)
No better alternative diagnosis with >50.0% certainty				0.35 (0.26, 0.43)	0.02 (−0.02, 0.07)
Persistent disease†					0.13 (0.10, 0.16)

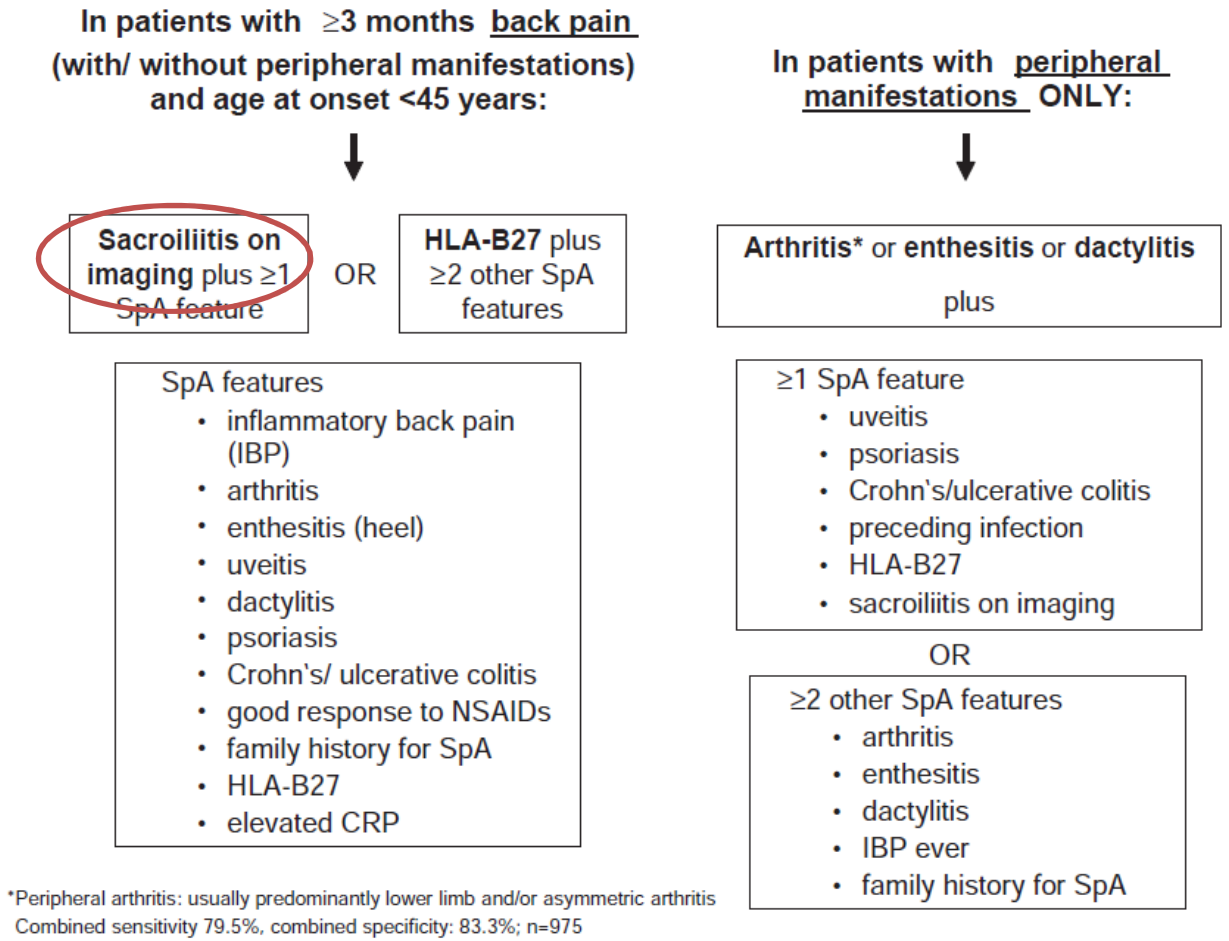
* Values are the kappa coefficient (95% confidence interval). The definitions of rheumatoid arthritis (RA) were the 1987 American College of Rheumatology (ACR) criteria, 2010 ACR/European League Against Rheumatism (EULAR) criteria, rheumatologist diagnosis of RA or no better alternative diagnosis with >50.0% certainty, persistent synovitis, and ≥1 joint erosions.
† Defined as having synovitis, receiving disease-modifying antirheumatic drug therapy, or both.
‡ 2010 ACR/EULAR criteria were present if noted at any time during followup.
§ 2010 ACR/EULAR criteria were present if noted at any time during followup, except the exclusion criterion (no better alternative diagnosis), which was considered present only if it was met at the last visit.

The problem was quite similar for spondyloarthritis



S.P. Raychaudhuri, A. Deodhar / Journal of Autoimmunity 48-49 (2014) 128–133

For which new ASAS criteria were published
to improve sensitivity
and allow to clinician the abibility to use biologics before sacroiliitis on X-Rays.



M Rudwaleit

Ann Rheum Dis 2011;**70**:25–31. doi:10.1136/ard.2010.133645

Clinical presentation of patients suffering from recent onset chronic inflammatory back pain suggestive of spondyloarthritis: The DESIR cohort

Maxime Dougados[a,b], Adrien Etcheto[b], Anna Molto[a,b], Sandrine Alonso[c], Sophie Bouvet[c], Jean-Pierre Daurès[c], Paul Landais[c], Maria-Antonietta d'Agostino[d], Francis Berenbaum[e], Maxime Breban[d], Pascal Claudepierre[f], Bernard Combe[g], Bruno Fautrel[h], Antoine Feydy[i], Philippe Goupille[j], Pascal Richette[k], Thao Pham[l], Christian Roux[a], Jean-Marc Treluyer[a], Alain Saraux[m], Désirée van der Heijde[n], Daniel Wendling[o,*]

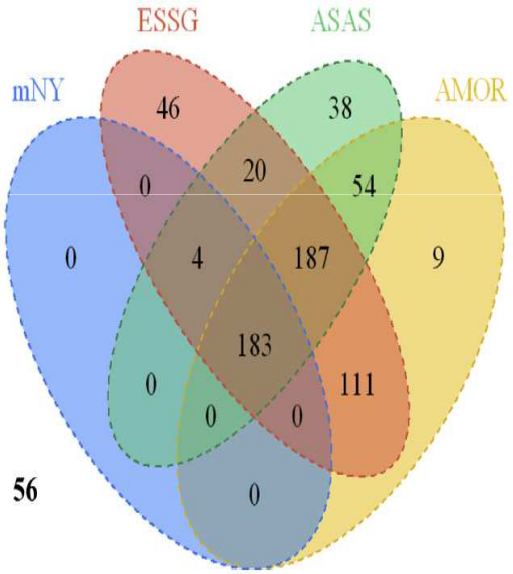


Fig. 1. Venn diagram representing the overlap between the various classification criteria for axial spondylo arthritis.

Criteria set	Sensitivity[a]	Specificity[b]
mNY	40.2 [34.9; 45.8]	85.5 [81.6; 88.9]
Amor	88.5 [84.5; 91.8]	29.2 [24.6; 34.2]
mAmor[e]	90.4 [86.6; 93.4]	25.7 [21.3; 30.5]
ESSG	87.2 [83.1; 90.6]	30.3 [25.7; 35.1]
mESSG[e]	91.2 [87.5; 94.0]	23.9 [19.7; 28.6]
ASAS	81.1 [76.3; 85.2]	39.0 [34.0; 44.2]

The difficulty is now the low concordance between all criteria and the validity of the ASAS criteria in patients older than forty years according to the low specificity of MRI in this context.

M. Dougados et al. / Joint Bone Spine 82 (2015) 345–351

2012 Revised International Chapel Hill Consensus Conference Nomenclature of Vasculitides

J. C. Jennette,[1] R. J. Falk,[1] P. A. Bacon,[2] N. Basu,[3] M. C. Cid,[4] F. Ferrario,[5] L. F. Flores-Suarez,[6] W. L. Gross,[7] L. Guillevin,[8] E. C. Hagen,[9] G. S. Hoffman,[10] D. R. Jayne,[11] C. G. M. Kallenberg,[12] P. Lamprecht,[13] C. A. Langford,[10] R. A. Luqmani,[14] A. D. Mahr,[15] E. L. Matteson,[16] P. A. Merkel,[17] S. Ozen,[18] C. D. Pusey,[19] N. Rasmussen,[20] A. J. Rees,[21] D. G. I. Scott,[22] U. Specks,[16] J. H. Stone,[23] K. Takahashi,[24] and R. A. Watts[25]

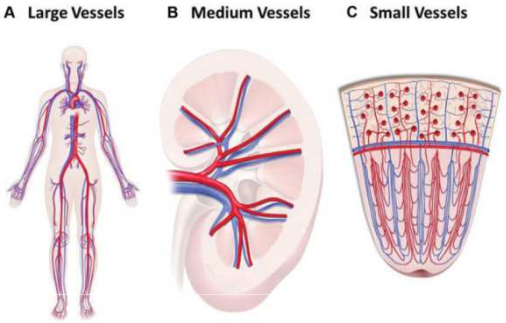
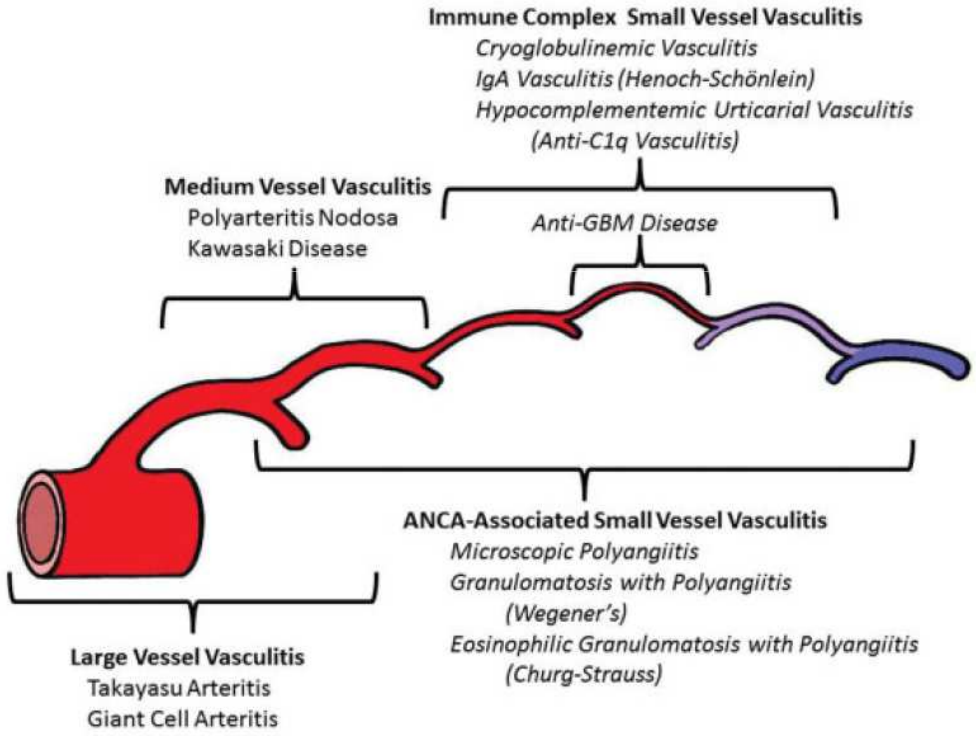


Table 1 | Are ANCA present in all patients with AAV?

Diagnosis	PR3-ANCA	MPO-ANCA	Elastase-ANCA	No ANCA detected	% of patients with ANCA
GPA (n=364)	323	25	4	12*	96
EGPA (n=36)	0	23	0	13	64
MPA (n=85)	16	67	1	1	98
NCGN (n=54)	4	47	1	2	94

*10 of 12 were ENT-limited GPA. Unpublished data. Abbreviations: AAV, ANCA-associated vasculitis; ANCA, anti-neutrophil cytoplasmic antibodies; ENT, ear nose and throat; EGPA, eosinophilic granulomatosis with polyangiitis; GPA, granulomatosis with polyangiitis; MPA, microscopic polyangiitis; NCGN, necrotizing and crescentic glomerulonephritis.



And it was the same in vasculitis for which we have new antibodies (ANCA), new names, and new treatment (Rituximab)

ARTHRITIS & RHEUMATISM
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Evolution over the time in France?

We used the ACR 1987 criteria for RA and the ESSG criteria for SpA in 2001 to evaluate the prevalence of RA in France and they are not used today…

Prevalence of spondyloarthropathies in France: 2001

A Saraux, F Guillemin, P Guggenbuhl, C H Roux, P Fardellone, E Le Bihan, A Cantagrel, I Chary-Valckenaere, L Euller-Ziegler, R-M Flipo, R Juvin, J-M Behier, B Fautrel, C Masson, J Coste

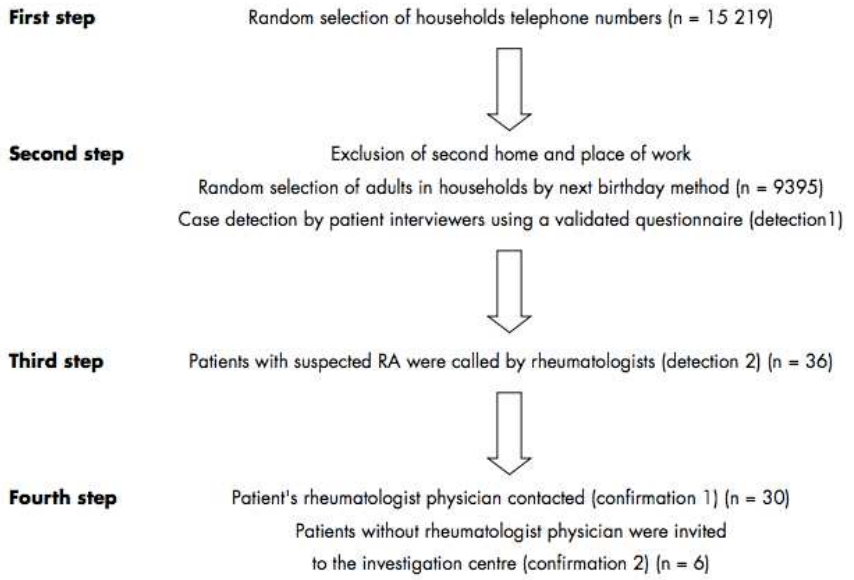
Table 1 Diagnosis and ESSG criteria in 29 patients with spondyloarthropathy

Patient	Age (years)	Sex	Diagnosis	Date of diagnosis	ESSG synovitis *	ESSG spinal pain †	ESSG criterion‡	Other ESSG criteria§
1	51	F	AS	1994	+	+	+	Enthesitis, buttock pain, sacroiliitis
2	50	F	Undifferentiated	1984	+	+	+	Psoriasis, enthesitis, buttock pain
3	31	F	PsA	1997	−	+	+	Psoriasis, buttock pain, sacroiliitis
4	26	F	Undifferentiated	?	−	+	+	Family history
5	67	M	AS	1992	−	+	+	Enthesitis, buttock pain, sacroiliitis
6	45	F	AS	1990	−	+	+	Family history, enthesitis, buttock pain, sacroiliitis
7	33	M	AS	1990	−	+	+	Family history, enthesitis, buttock pain, sacroiliitis
8	43	M	PsA	1991	−	+	+	Psoriasis, enthesitis, buttock pain,
9	21	M	Undifferentiated	?	+	+	+	Family history, psoriasis, infection
10	50	M	AS	1985	+	+	+	Family history, sacroiliitis
11	70	F	AS and PsA	1992	+	−	+	Family history, psoriasis, infection, enthesitis, sacroiliitis
12	78	F	PsA	1970	+	+	+	Family history, psoriasis, buttock pain, sacroiliitis
13	23	F	PsA	1992	−	+	+	Family history, psoriasis
14	43	F	PsA	1986	−	+	+	Psoriasis, sacroiliitis
15	60	M	PsA	1967	+	+	+	Buttock pain, sacroiliitis
16	52	F	Undifferentiated	1990	+	+	−	
17	66	F	AS	1979	−	+	+	Family history, buttock pain, sacroiliitis
18	36	M	AS	1988	+	+	+	Buttock pain, sacroiliitis
19	66	F	AS	1972	−	+	+	Enthesitis, buttock pain, sacroiliitis
20	31	F	AS	1997	−	+	+	Family history, buttock pain, sacroiliitis
21	40	F	PsA	1991	−	+	+	Psoriasis, buttock pain
22	27	M	AS	1990	+	+	+	Enthesitis, buttock pain, sacroiliitis
23	39	M	AS	1992	−	+	+	Enthesitis, buttock pain, sacroiliitis
24	68	F	AS	1988	−	+	+	Buttock pain
25	32	F	AS	1995	−	+	+	Family history, buttock pain
26	57	F	PsA	1989	+	+	+	Psoriasis, buttock pain
27	69	M	PsA	1960	+	−	+	Psoriasis, enthesitis, IBD
28	53	M	PsA	1975	+	−	+	Family history, psoriasis
29	63	F	PsA	1995	+	−	+	Family history, psoriasis, infection

Ann Rheum Dis 2005;**64**:1431–1435.

Prevalence of rheumatoid arthritis in France: 2001

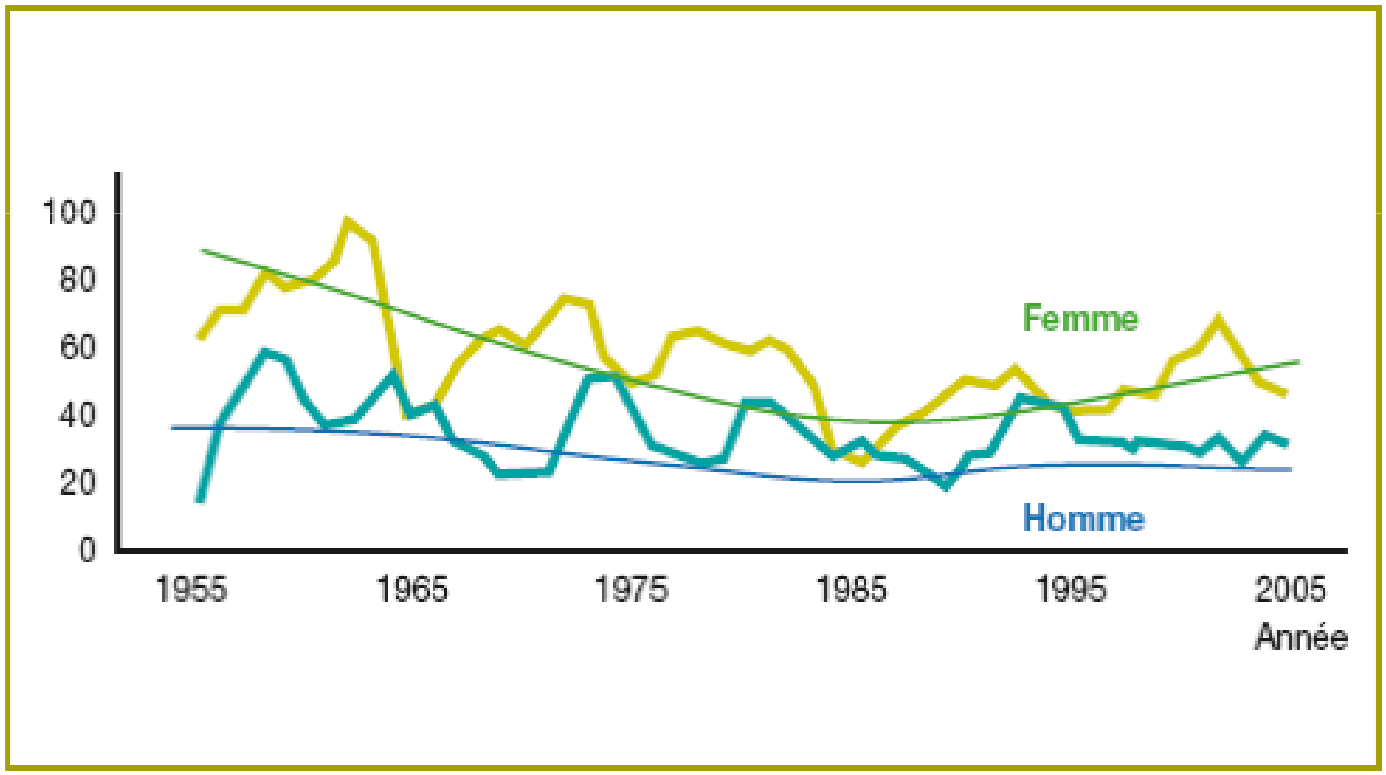
F Guillemin, A Saraux, P Guggenbuhl, C H Roux, P Fardellone, E Le Bihan, A Cantagrel, I Chary-Valckenaere, L Euller-Ziegler, R-M Flipo, R Juvin, J-M Behier, B Fautrel, C Masson, J Coste



Ann Rheum Dis 2005;**64**:1427–1430.

Evolution over the time in US

The longer evaluation that we have is about RA according to criteria obtained retrospectively in the Olmsted county (USA) using the 1987 ACR criteria



Myasedova E. Arthritis Rheum. 2010;62:1576-82.

How to do criteria?

The ACR decided to not build diagnosis criteria but to limit their efforts on classification

- Classification criteria
  - choice of a gold standard
  - comparison to another population
  - identification of a combination of signs separating disease from control
- Diagnosis criteria
  - inclusion of a cohort of patients suspected as having the disease
  - gold standard is the diagnostic after the follow up

How to do criteria?

Rheumatoid Arthritis

•Phase 1, used cohort data to identify the key factors to be considered in the new criteria, and their associated weights

•Phase 2

1) assembly of an expert panel: results of phase 1 and published data
2) development and rank ordering of patient case scenarios using multiple possible combinations of clinical features, decision analytic software (1000Minds)
3) 2-day in-person consensus meeting,
4) assessment of face and construct validity: Cases were rank ordered from highest probability of developing RA (score closest to 100) to lowest probability of developing RA (score closest to 0) and the panelists indicated if they would treat with MTX or another disease-modifying anti-rheumatic drug (DMARD) or enroll the patient into a clinical trial of an investigational biologic therapy

Neogi T, Arthritis Rheum. 2010 Sep; 62(9): 2582–2591.

The use and abuse of diagnostic/classification criteria

Rayford R. June [a, 1], Rohit Aggarwal [b, *]

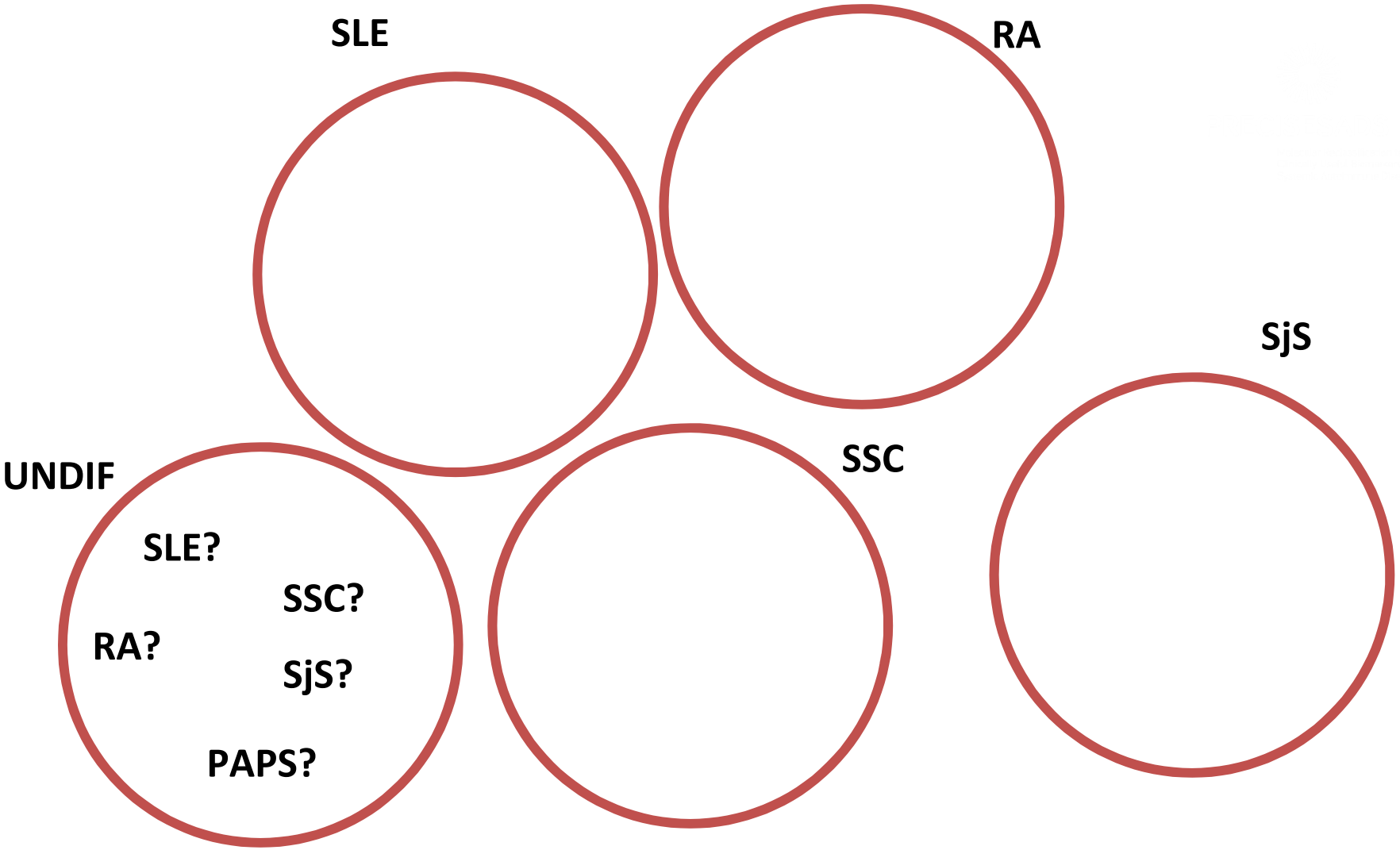
Practice points

- Meeting disease classification criteria is not necessary for clinical diagnosis or treatment
- Classification criteria are designed for clinical research to create well-defined relatively homogeneous groups for disease investigation
- Classification criteria should be used as a guide to clinicians in the clinical diagnosis and evaluation of common diseases, but clinicians should use their clinical knowledge for making the final diagnosis
- Physicians should understand the sensitivity, specificity, positive predictive value, negative predictive values, and influence of epidemiology on any criteria before applying them
- Clinicians and researchers should be aware of the trend of decreasing specificity with increased sensitivity of some of the newer classification criteria—understanding that more early disease may be classified with the risk of misclassification

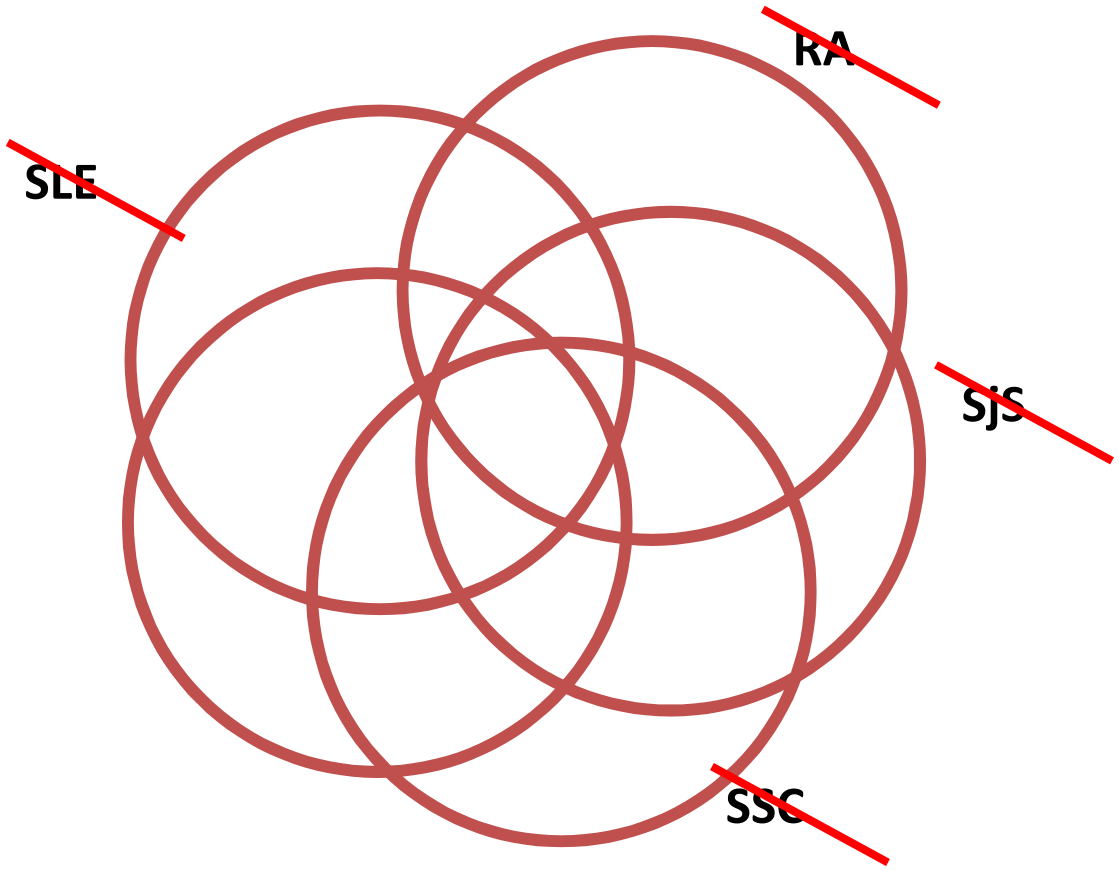
Best Practice & Research Clinical Rheumatology 28 (2014) 921–934

Future direction: Molecular taxonomy

PRECISEADS: molecular classification of systemic autoimmune diseases

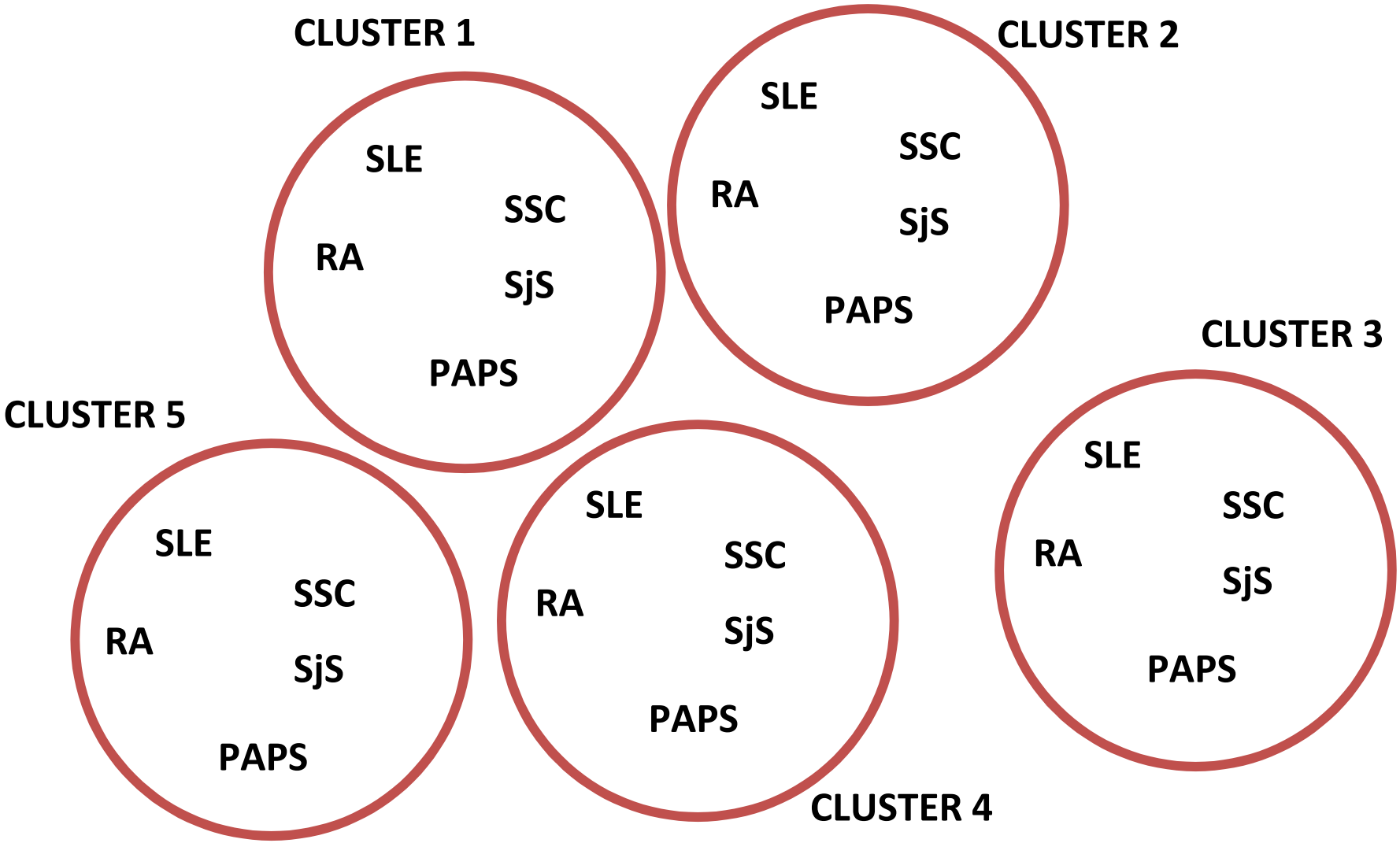


We have seen that for autoinflammatory disease that a new gold standard based on mutation gave new classification. The same approach should be obtained



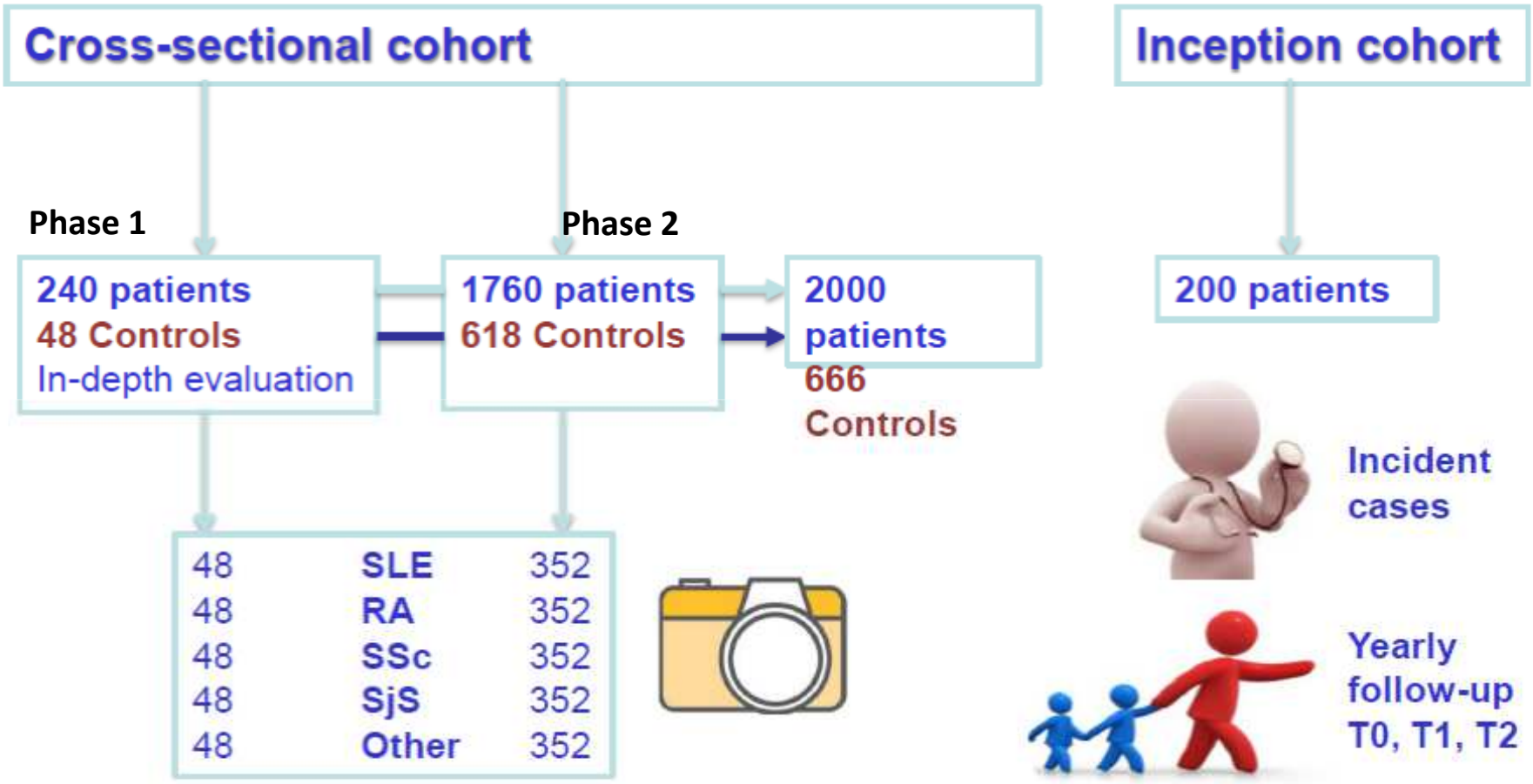
The principle is to mix all autoimmune diseases

Identification de clusters



And then to identify clusters on the basis of omics

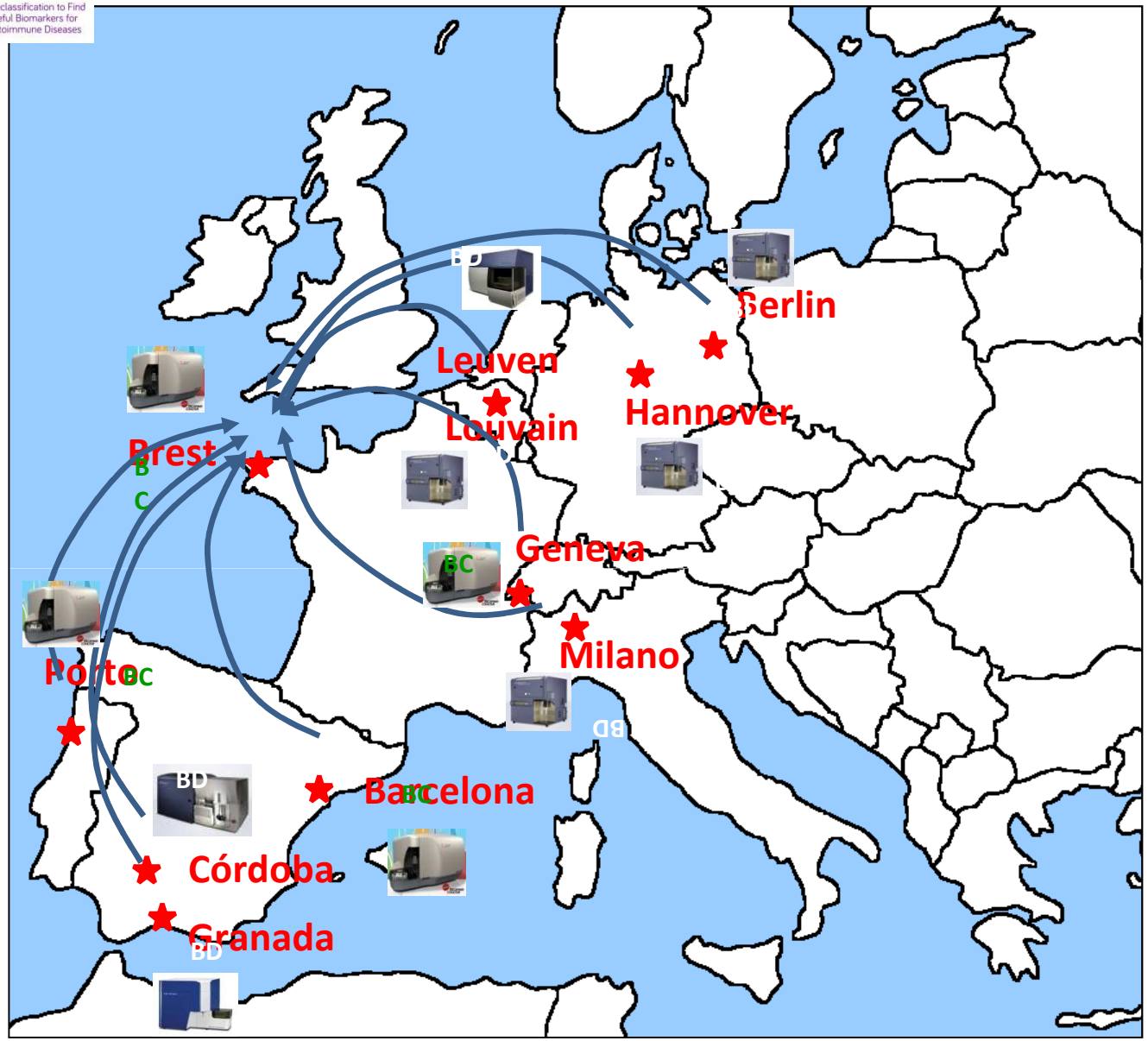
Project Structure – Patients' cohorts



We are including patients in both cross sectional and inception cohorts

PRECISESADS
Molecular Reclassification to Find Clinically Useful Biomarkers for Systemic Autoimmune Diseases

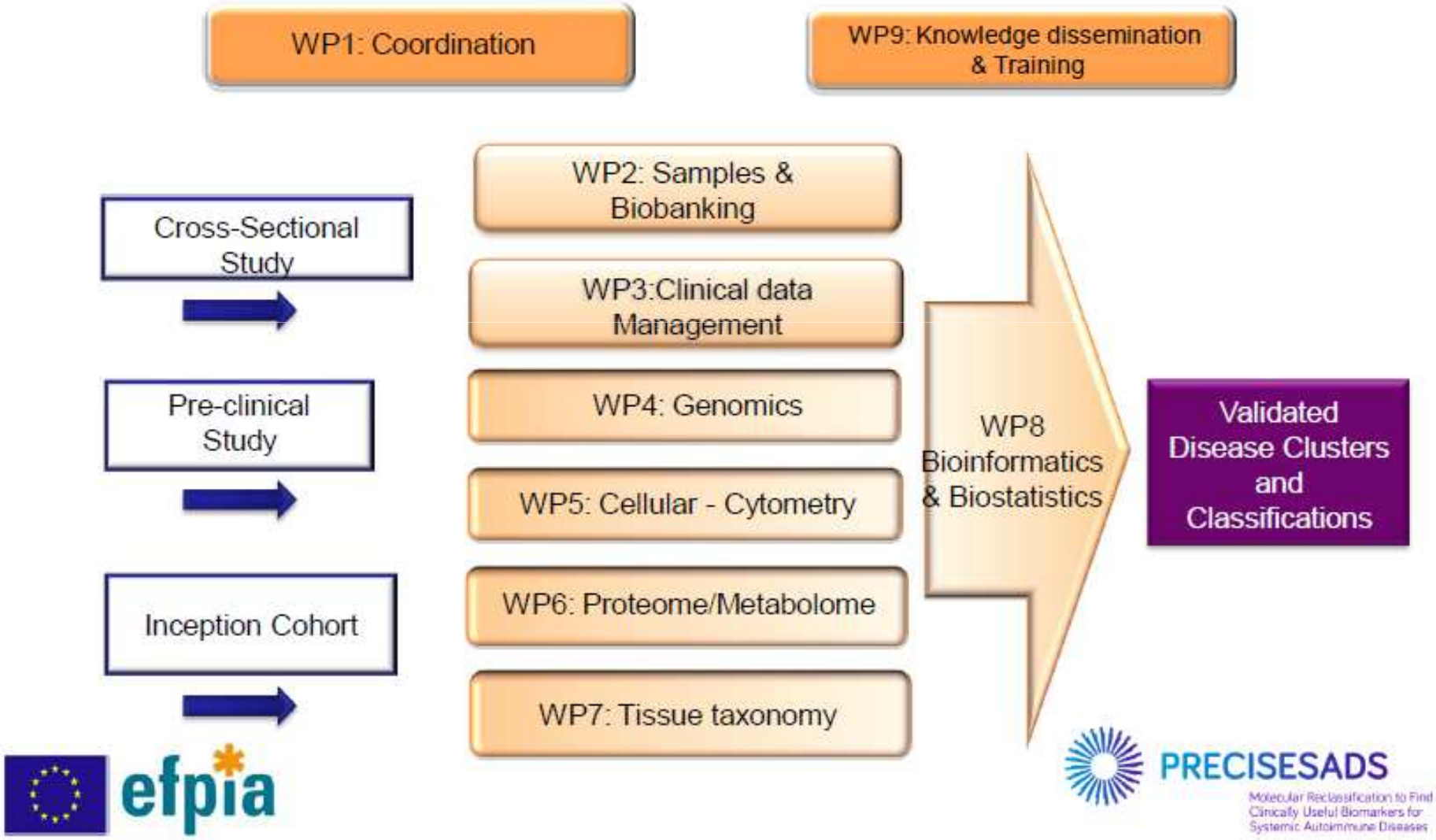
innovative medicines initiative



In Europe

Complementary Work Packages Deliver Complex Data

Bioinformatic analysis delivers insight to underlying taxonomy



Conclusion

- Diagnosis of a disease remains doctor dependant, not based on diagnosis criteria
- But classification criteria help clinicians and homogenize the meaning.
- To do a diagnosis, exclusion of differential diagnosis is as important as the presence of positive signs
- Criteria are good indicators for epidemiological studies but they are changing over the time, on the basis of knowledge about etiology, diagnoses tools, discordance between criteria, or new treatment

and that explain why we are not able to follow the epidemiology of IRD over the time…..