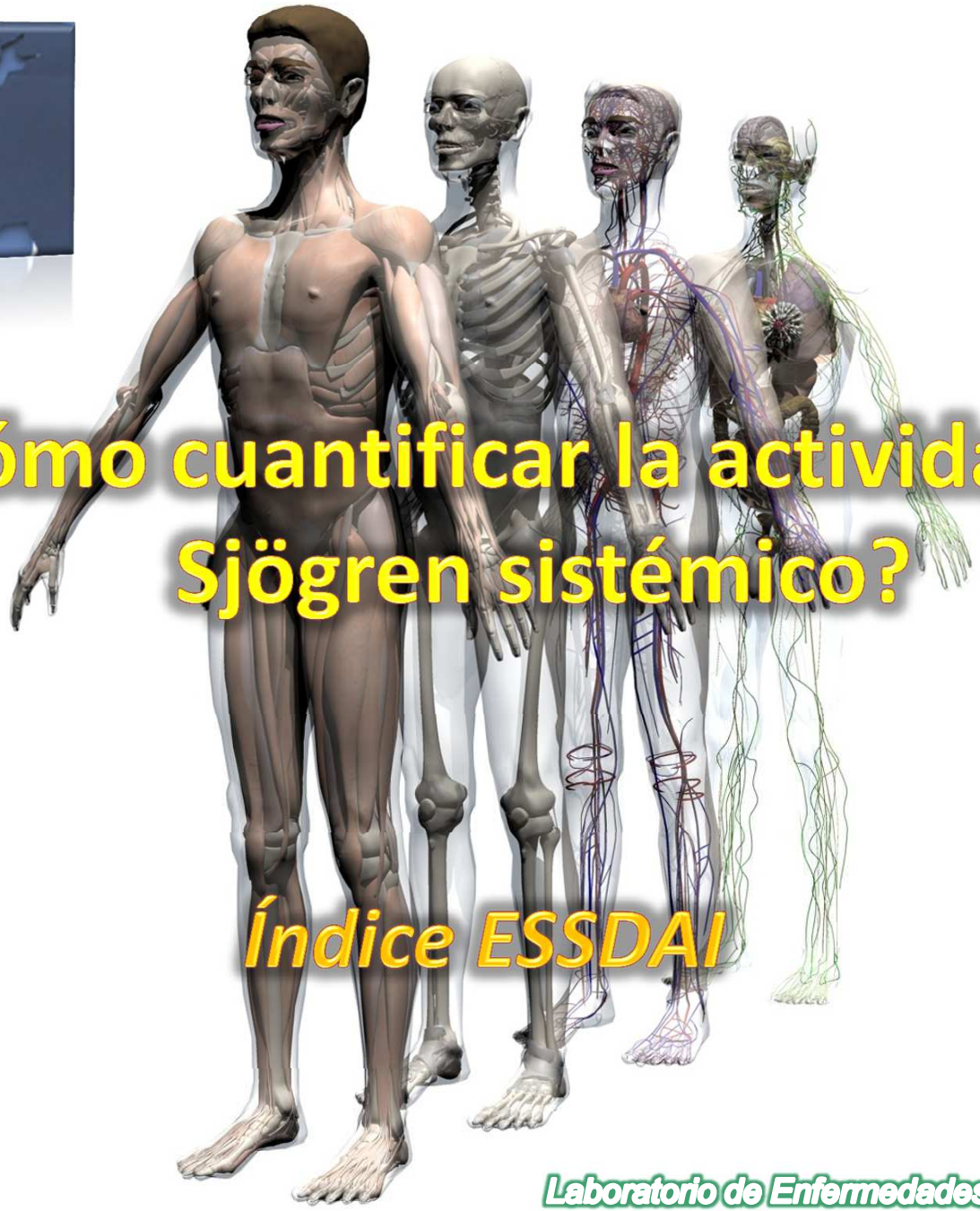
GEAS

Grupo de Enfermedades Autoinmunes Sistémicas



¿Cómo cuantificar la actividad del Sjögren sistémico?

Índice ESSDAI

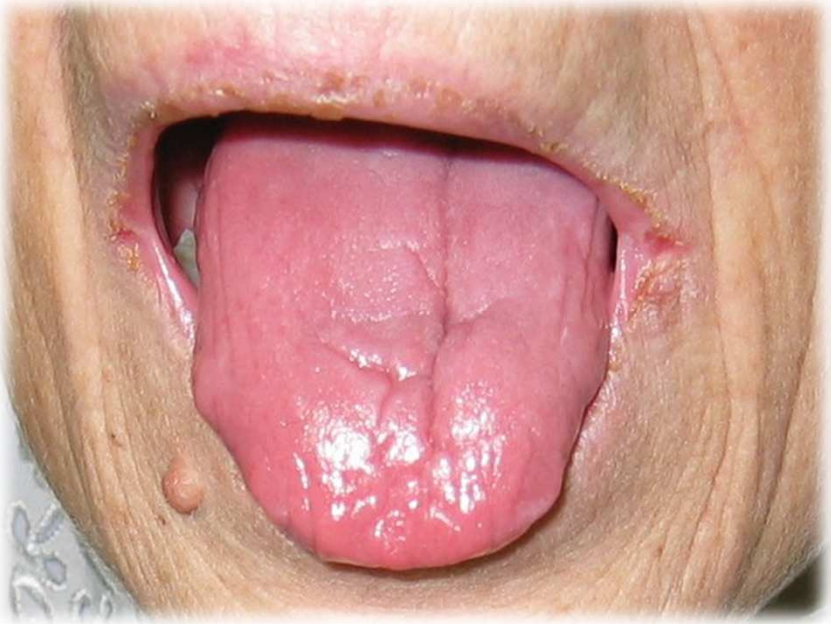
Pilar Brito-Zerón
Laboratorio de Enfermedades Autoimunes "Josep Font"
Hospital Clínic, Barcelona

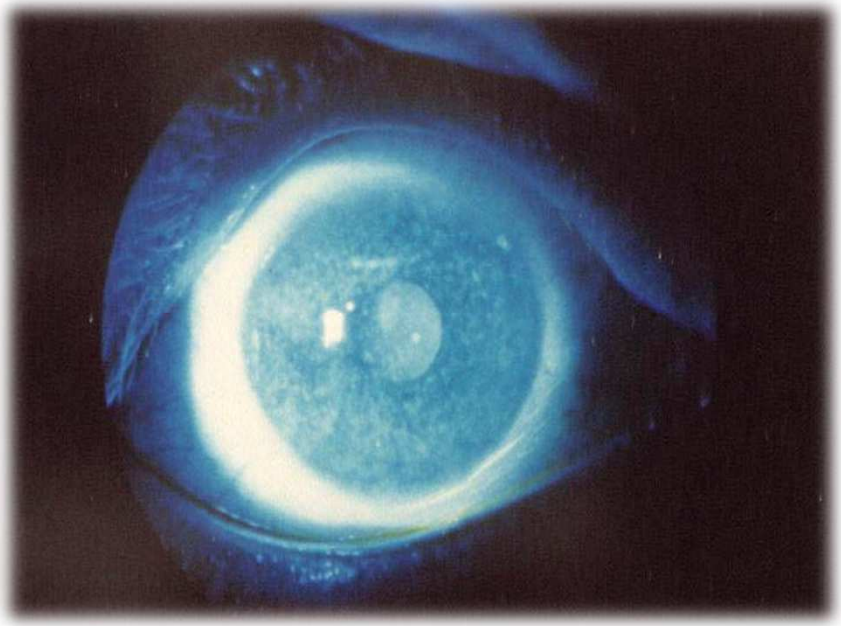
CONTENIDOS

1. **INTRODUCCIÓN** (3 min)
2. **ESSDAI**: ¿Cómo utilizarlo? (6 min)
3. **ESSDAI**: ¿Para qué sirve? (6 min)
4. **FUTURO** (1 min)

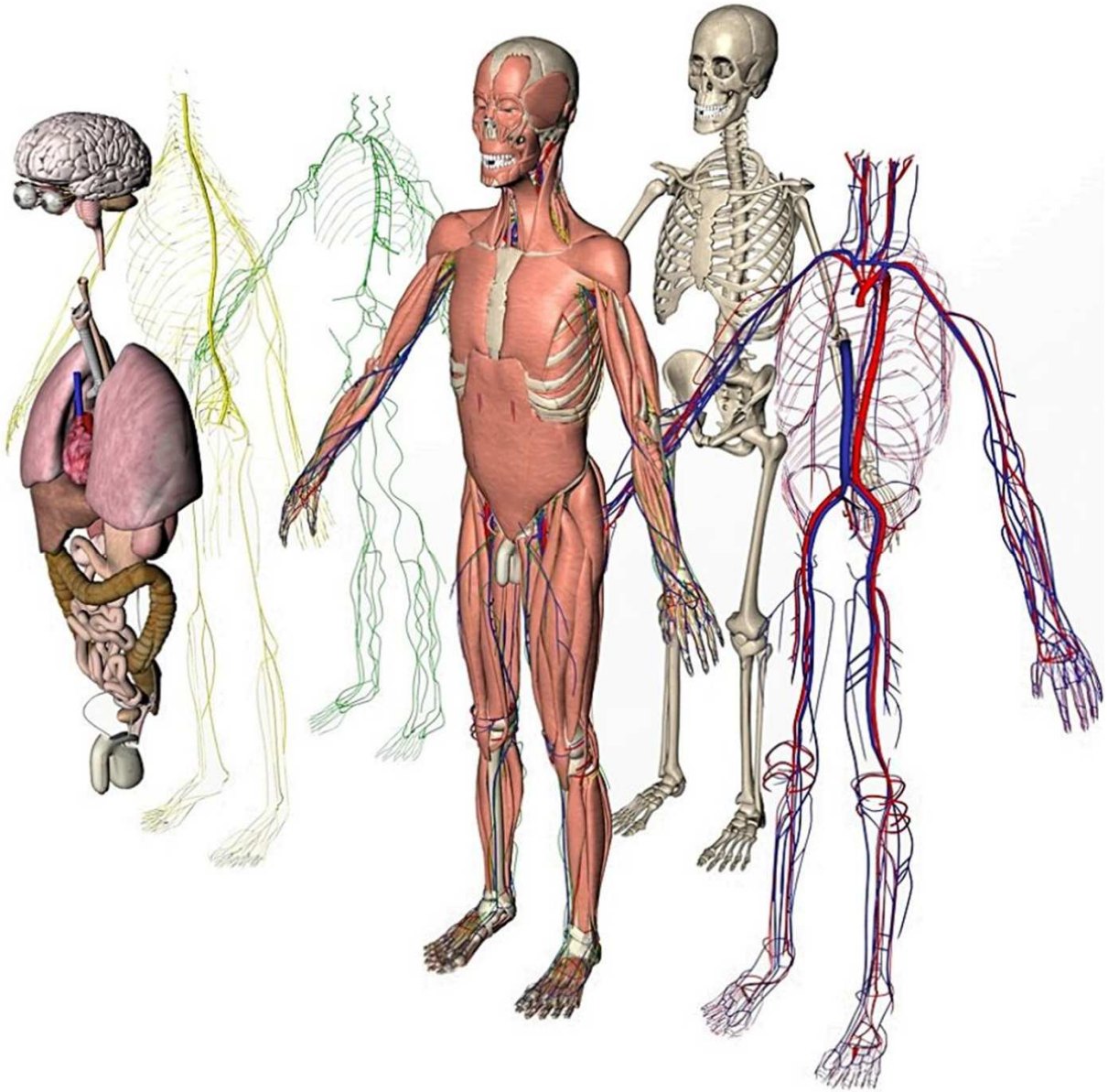
La sequedad de mucosas es la principal afectación

del síndrome de Sjögren





El síndrome de Sjögren es una enfermedad sistémica



REVIEW

CURRENT OPINION

Advances in the understanding and treatment of systemic complications in Sjögren syndrome

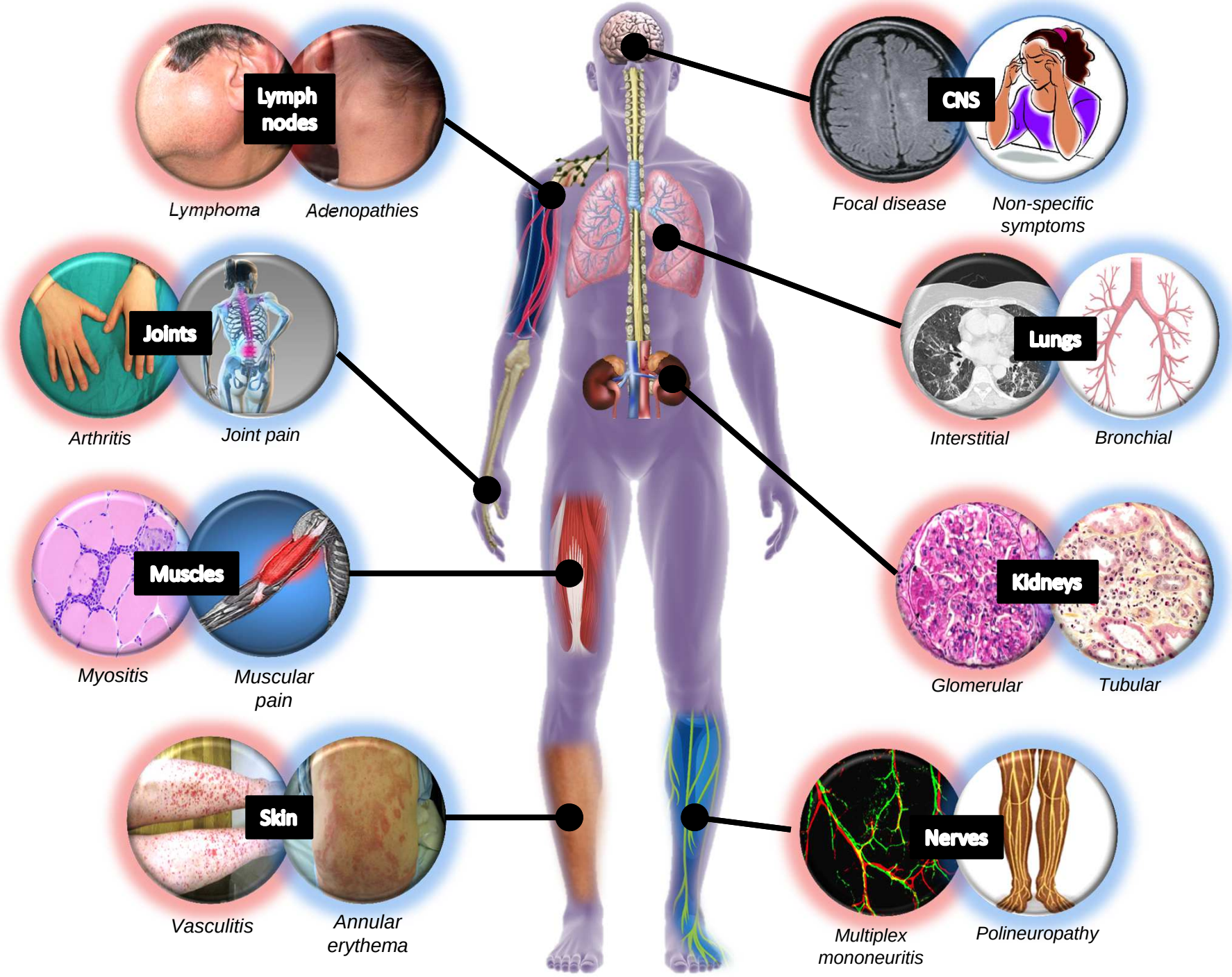
Pilar Brito-Zerón and Manuel Ramos-Casals, on behalf of the EULAR-SS task force group

Purpose of review
Primary Sjögren syndrome is a systemic autoimmune disease whose clinical spectrum extends from sicca syndrome to systemic involvement (extraglandular manifestations). Recent reports have focused on expanding the clinical characterization and improving the diagnostic and therapeutic management of systemic Sjögren.

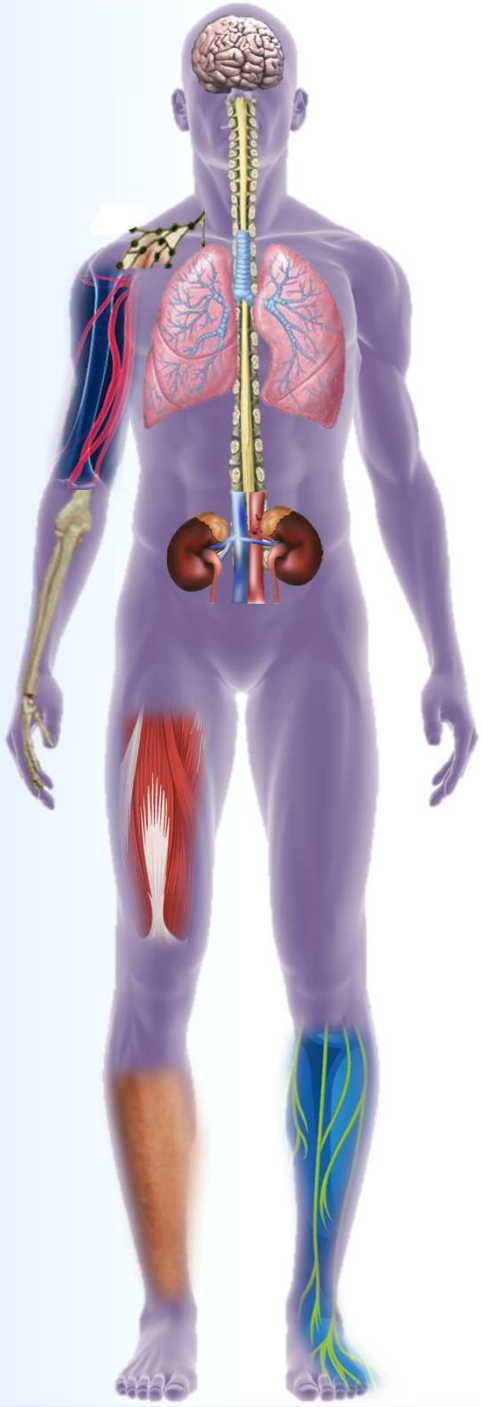
Recent findings
The development of the EULAR-SS disease activity index represents a step forward in the evaluation of systemic Sjögren, and three multicenter studies including more than 2500 European patients have confirmed that primary Sjögren syndrome is, undeniably, a systemic autoimmune disease. Systemic involvement plays a key role in the prognosis of primary Sjögren syndrome, and recent studies have focused on cutaneous, pulmonary, renal and neurological disease features. Other studies comparing the two sets of Sjögren syndrome criteria (American College of Rheumatology vs. American-European Consensus Group) in real-life patients found a moderate level of agreement. Autoantibodies are clues to an early diagnosis, as positivity confirms an autoimmune origin of the sicca syndrome and may appear several years before the disease diagnosis. In patients with a high clinical suspicion and negative results for the standard determination of anti-Ro/SS-A antibodies, specific detection of anti-Ro52/60 antibodies is recommended. Direct and indirect B-cell blocking seems to be the most promising biological pathway in primary Sjögren syndrome, especially for systemic involvement, although a large controlled trial has failed to demonstrate the efficacy of rituximab for nonsystemic symptomatology (dryness, fatigue and pain).

Summary
Dryness of the mucosal surfaces is the pivotal, but not only, clinical involvement that characterizes primary Sjögren syndrome. There is growing interest in and knowledge of the clinical characterization and therapeutic management of systemic Sjögren.

Keywords
belimumab, EULAR-SS disease activity index, lymphoma, rituximab, Sjögren syndrome, systemic autoimmune disease


Lymph nodes
Lymphoma
Adenopathies
CNS
Focal disease
Non-specific symptoms
Joints
Arthritis
Joint pain
Lungs
Interstitial
Bronchial
Muscles
Myositis
Muscular pain
Kidneys
Glomerular
Tubular
Skin
Vasculitis
Annular erythema
Nerves
Multiplex mononeuritis
Polineuropathy

¿CÓMO CUANTIFICAR LA AFECTACIÓN SISTÉMICA DEL SÍNDROME DE SJÖGREN?



The European League Against Rheumatism (EULAR) recientemente ha promovido y apoyado un estudio internacional (EULAR SS Task Force) con el objetivo de desarrollar unos índices de actividad consensuados para el síndrome de Sjögren.

Journal of Autoimmunity 39 (2012) 97–102

Contents lists available at SciVerse ScienceDirect

Journal of Autoimmunity

EULAR-Sjögren Task Force

Review

Outcome measures for primary Sjögren's syndrome

Raphaèle Seror [a,b,c], Hendrika Bootsma [d], Simon J. Bowman [e], Thomas Dörner [f], Jacques-Eric Gottenberg [g], Xavier Mariette [a], Manel Ramos-Casals [h], Philippe Ravaud [b,c], Elke Theander [i], Athanasios Tzioufas [j], Claudio Vitali [k,*]

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[b] *Center of Clinical Epidemiology, Hôpital Hôtel Dieu, Paris, France*
[c] *INSERM U738, Université Paris-René Descartes, Paris, France*
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[e] *Rheumatology Department, University Hospital Birmingham NHS Foundation Trust, Birmingham, UK*
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[g] *Department of Rheumatology, Strasbourg University Hospital, Université de Strasbourg, EA 4438, Strasbourg, France*
[h] *Laboratory of Autoimmune Diseases "Josep Font", Hospital Clinic, Barcelona, Spain*
[i] *Department of Rheumatology, Malmö University Hospital, Lund University, Sweden*
[j] *Department of Pathophysiology, School of Medicine, University of Athens, Greece*
[k] *Department of Internal Medicine and Section of Rheumatology, Villamarina Hospital, Piombino, Italy*

EULAR SS Task Force

Se han desarrollado 2 índices: ESSDAI (sistémico) y ESSPRI (calidad de vida)

Extended report

EULAR Sjögren's syndrome disease activity index: development of a consensus systemic disease activity index for primary Sjögren's syndrome

Raphaèle Seror,[1–5] Philippe Ravaud,[1–3] Simon J Bowman,[6] Gabriel Baron,[1–3] Athanasios Tzioufas,[7] Elke Theander,[8] Jacques-Eric Gottenberg,[9] Hendrika Bootsma,[10] Xavier Mariette,[4,5] Claudio Vitali[11]; on behalf of the EULAR Sjögren's Task Force

Extended report

EULAR Sjögren's Syndrome Patient Reported Index (ESSPRI): development of a consensus patient index for primary Sjögren's syndrome

Raphaèle Seror,[1–3] Philippe Ravaud,[1,2] Xavier Mariette,[3–5] Hendrika Bootsma,[6] Elke Theander,[7] Arne Hansen,[8] Manel Ramos-Casals,[9] Thomas Dörner,[8] Stefano Bombardieri,[10] Eric Hachulla,[11] Johan G Brun,[12] Aike A Kruize,[13,14] Sonja Praprotnik,[15] Matija Tomsic,[15] Jacques-Eric Gottenberg,[16] Valerie Devauchelle,[17] Salvatore Devita,[18] Cristina Vollenweider,[19] Thomas Mandl,[7] Athanasios Tzioufas,[20] Steven Carsons,[21] Alain Saraux,[17] Nurhan Sutcliffe,[22] Claudio Vitali,[23] Simon J Bowman[24]; on behalf of the EULAR Sjögren's Task Force

EULAR SS Task Force

Se acaba de publicar el estudio de validación de ambos índices

ARD Online First, published on January 17, 2014 as 10.1136/annrheumdis-2013-204615

EXTENDED REPORT

Validation of EULAR primary Sjögren's syndrome disease activity (ESSDAI) and patient indexes (ESSPRI)

Raphaèle Seror,[1,2] Elke Theander,[3] Johan G Brun,[4] Manel Ramos-Casals,[5] Valeria Valim,[6] Thomas Dörner,[7] Hendrika Bootsma,[8] Athanasios Tzioufas,[9] Roser Solans-Laqué,[10] Thomas Mandl,[3] Jacques-Eric Gottenberg,[11] Eric Hachulla,[12] Kathy L Sivils,[13] Wan-Fai Ng,[14] Anne-Laure Fauchais,[15] Stefano Bombardieri,[16] Guido Valesini,[17] Elena Bartoloni,[18] Alain Saraux,[19] Matija Tomsic,[20] Takayuki Sumida,[21] Sumusu Nishiyama,[22] Roberto Caporali,[23] Aike A Kruize,[24] Cristina Vollenweider,[25] Philippe Ravaud,[2] Claudio Vitali,[26] Xavier Mariette,[1] Simon J Bowman,[27] on behalf of the EULAR Sjögren's Task Force

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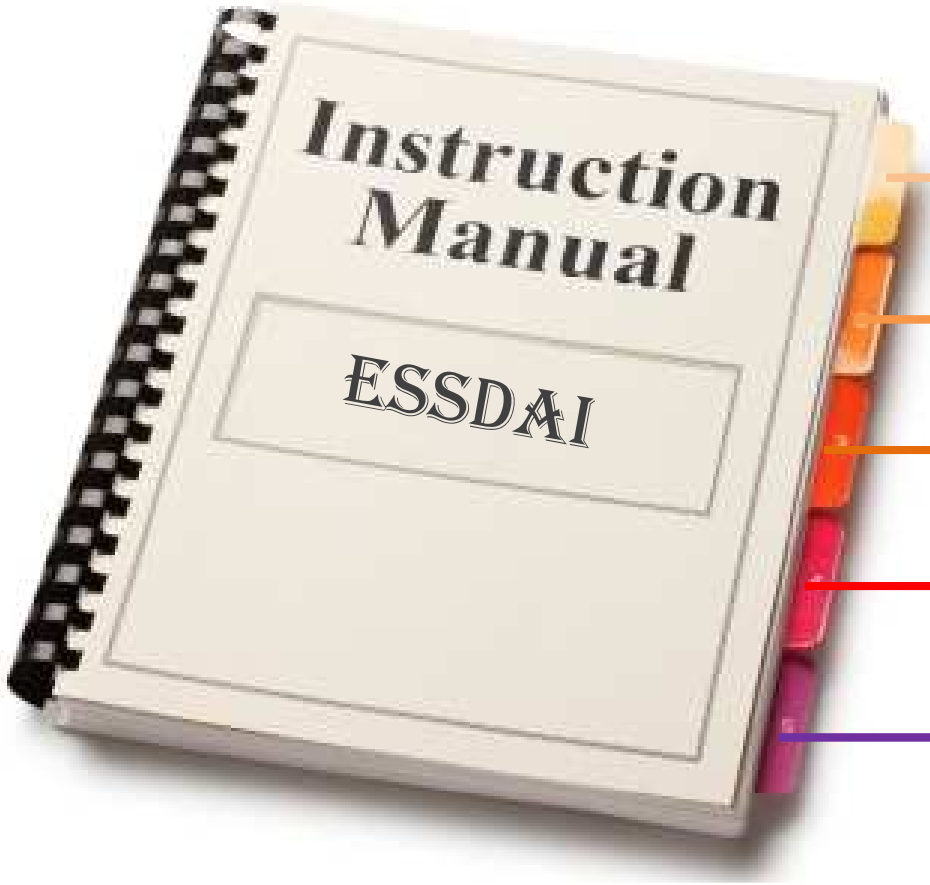
EXTENDED REPORT

Validation of EULAR primary Sjögren's syndrome disease activity (ESSDAI) and patient indexes (ESSPRI)



- ¿Qué es?
- Estructura
- ¿Cómo puntuar?
- ¿Cúando aplicarlo?
- Interés práctico

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EXTENDED REPORT

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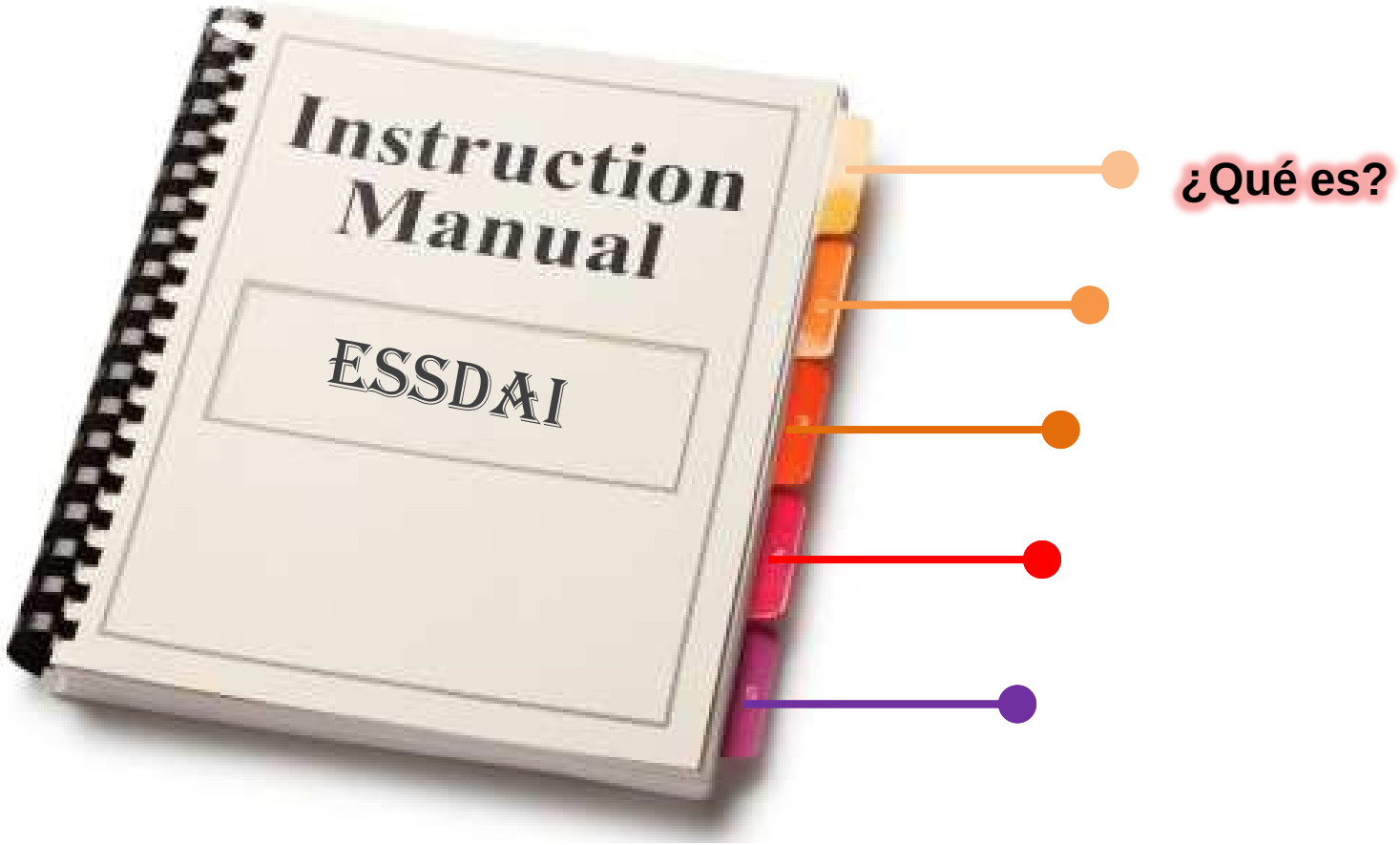

¿Qué es?

Journal of Autoimmunity xxx (2013) 1–6

Contents lists available at ScienceDirect

Journal of Autoimmunity

journal homepage: www.elsevier.com/locate/jautimm

Review

Outcome measures for primary Sjögren's syndrome: A comprehensive review

Raphaèle Seror [a,b,c,d,*,1], Elke Theander [e,1], Hendrika Bootsma [f,1], Simon J. Bowman [g,1], Athanasios Tzioufas [h,1], Jacques-Eric Gottenberg [i,1], Manel Ramos-Casals [j,1], Thomas Dörner [k,1], Philippe Ravaud [c,d,1], Xavier Mariette [a,b,1], Claudio Vitali [l,1]

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[b] Université Paris-Sud 11, INSERM U1012, Le Kremlin Bicêtre, France
[c] Center of Clinical Epidemiology, Hôpital Hôtel Dieu, Paris, France
[d] INSERM U738, Université Paris-René Descartes, Paris, France
[e] Department of Rheumatology, Skane University Hospital Malmö, Lund University, Sweden
[f] Department of Rheumatology and Clinical Immunology, University of Groningen, University Medical Center Groningen, P.O. Box 30.001, 9700 RB Groningen, The Netherlands
[g] Rheumatology Department, University Hospital Birmingham NHS Foundation Trust, Birmingham, UK
[h] Department of Pathophysiology, School of Medicine, University of Athens, Greece
[i] Rheumatology, Centre National de Référence des Maladies Auto-Immunes Rares, INSERM UMRS_1109, Fédération de Médecine Translationnelle de Strasbourg (FMTS), Strasbourg University Hospital, Université de Strasbourg, Strasbourg, France
[j] Laboratory of Autoimmune Diseases "Josep Font", IDIBAPS, ICMiD, Hospital Clinic, Barcelona, Spain
[k] Rheumatology Department, Charité, University Hospital, Berlin, Germany
[l] Sections of Rheumatology, Instituto San Giuseppe, Como and Casa di Cura di Lecco, Lecco, Italy

The ESSDAI is a disease activity index that was generated in 2009 in a cohort of 702 clinical vignettes based on 96 real patients. The score has been developed by consensus of a large group of worldwide experts from European and North American countries [22,27].

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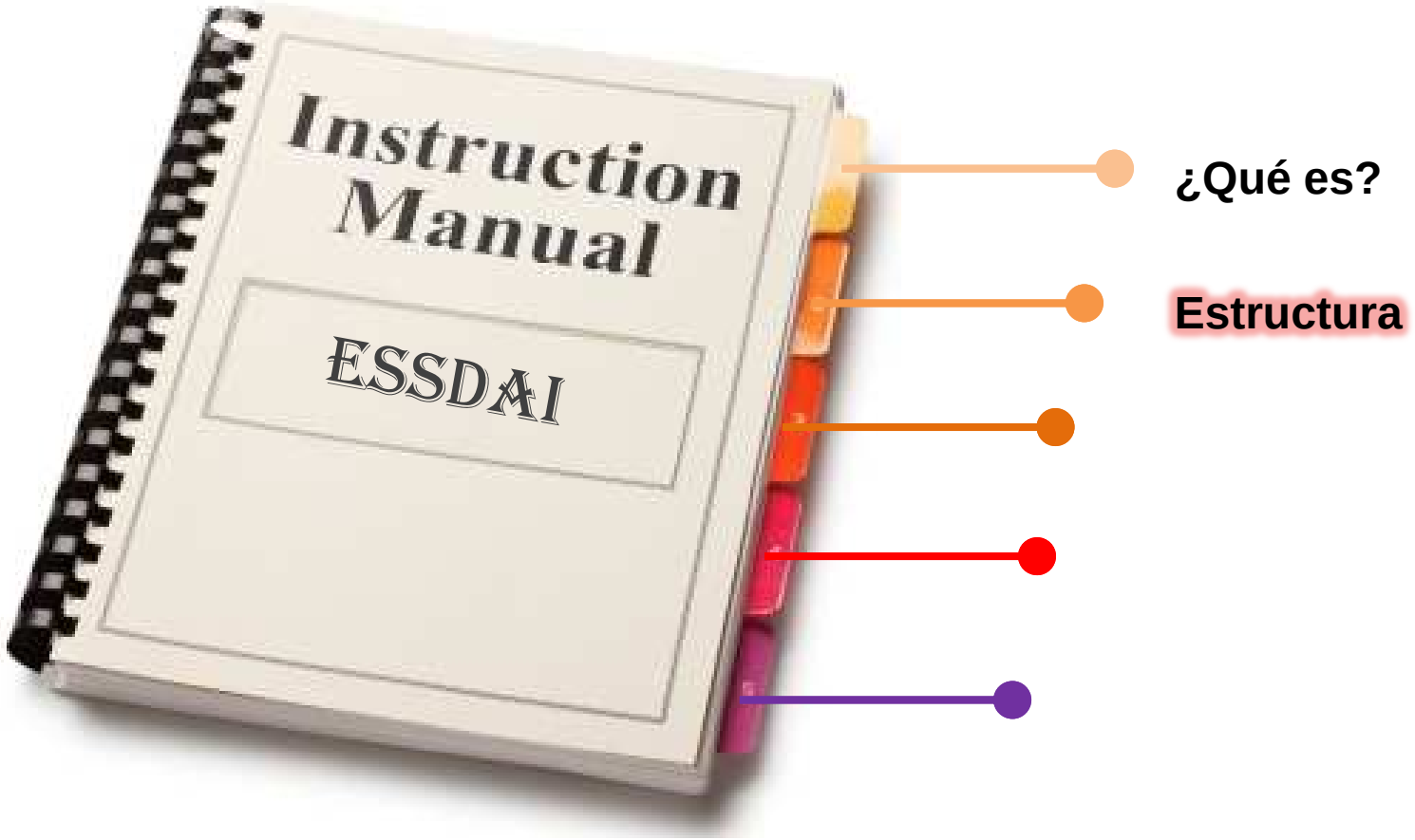
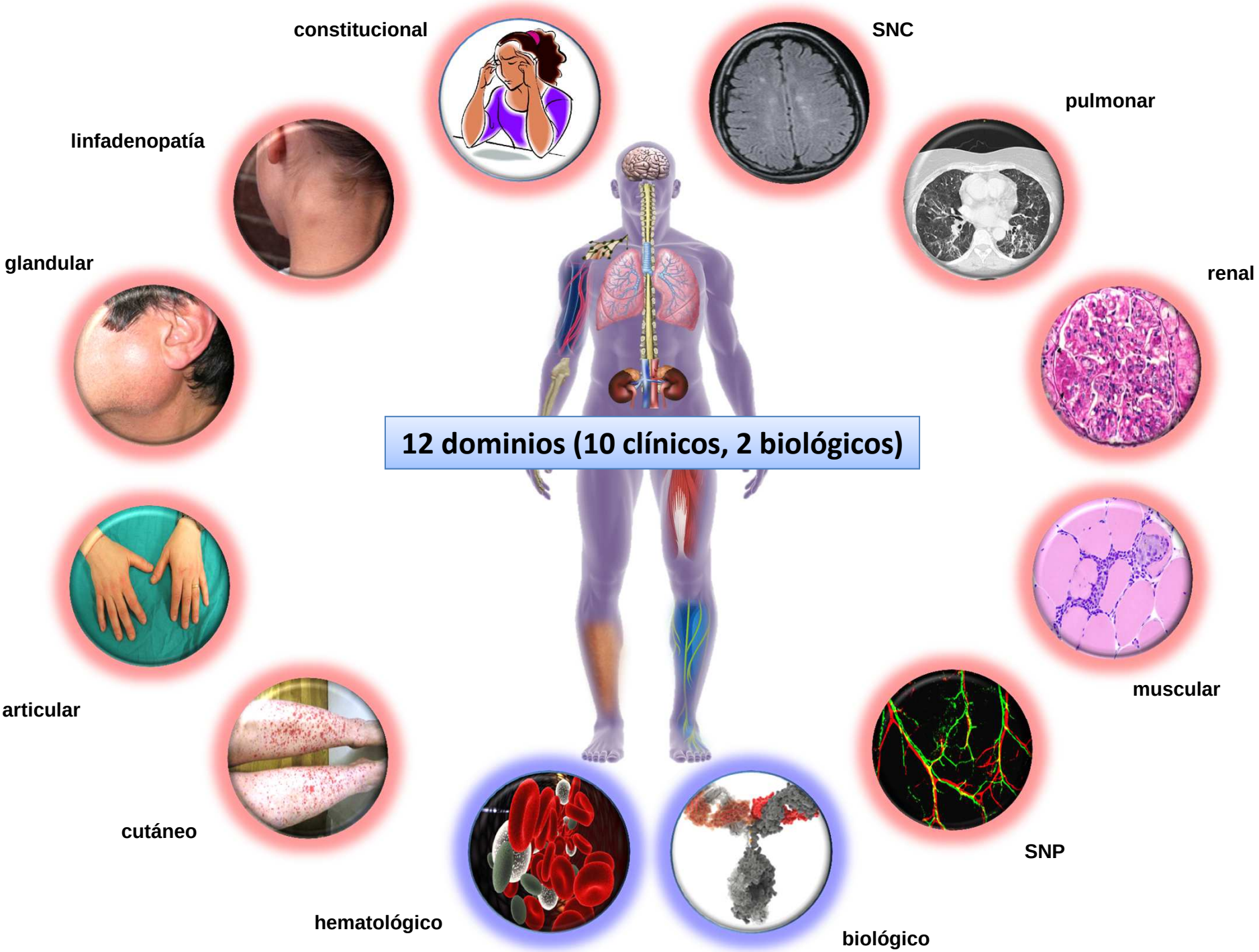


¿Qué es?

Estructura


constitucional
SNC
pulmonar
linfadenopatía
glandular
renal
12 dominios (10 clínicos, 2 biológicos)
articular
muscular
cutáneo
SNP
hematológico
biológico

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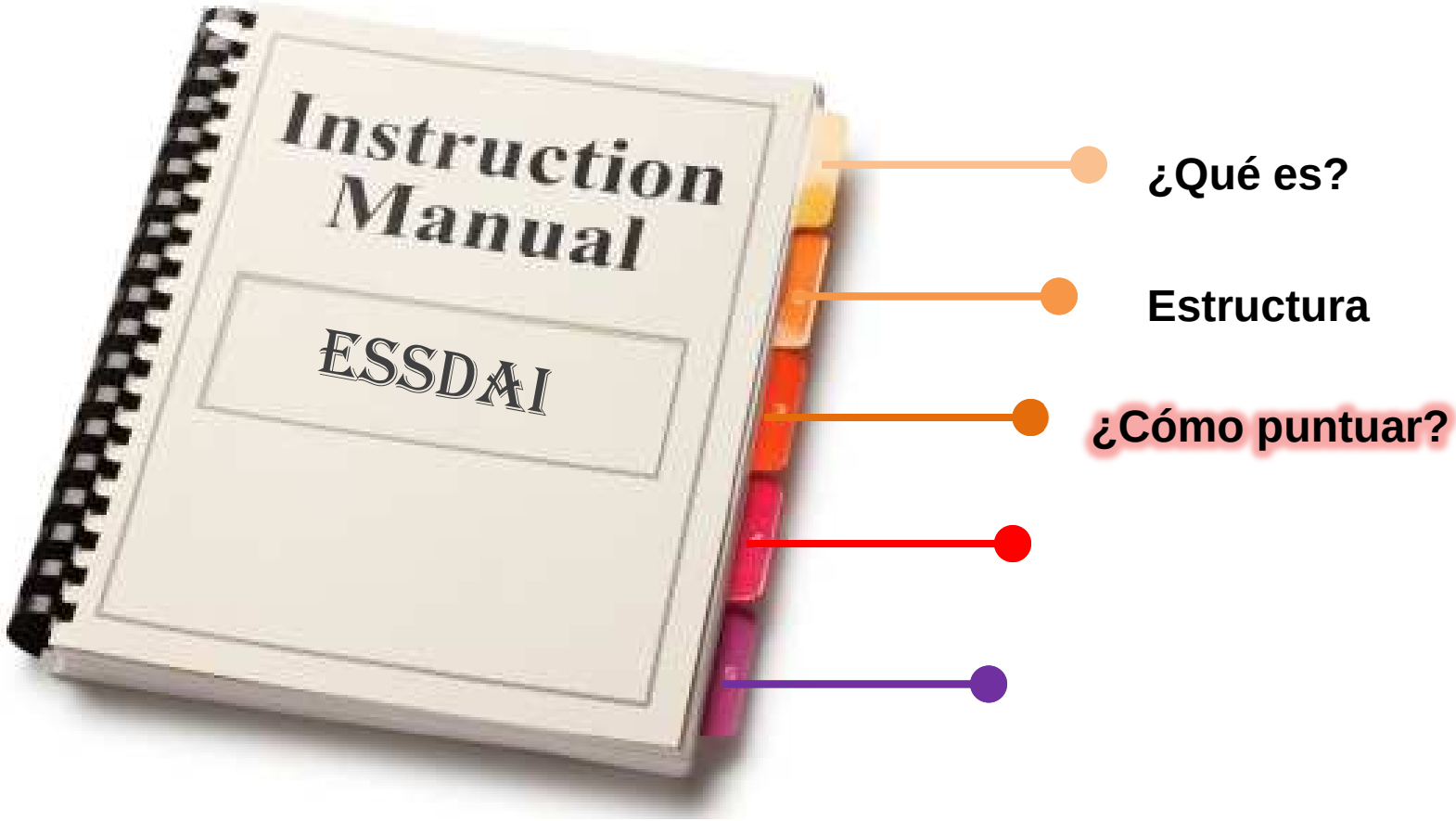


Table 1 The EULAR Sjögren's Syndrome Disease Activity Index (ESSDAI): domain and item definitions and scores

Domain	Activity level	Description
Constitutional *Exclusion of fever of infectious origin and voluntary weight loss*	No=0 Low=3 Moderate=6	Absence of the following symptoms Mild or intermittent fever (37.5°–38.5°C)/night sweats and/or involuntary weight loss of 5–10% of body weight Severe fever (>38.5°C) / night sweats and/or involuntary weight loss of >10% of body weight
Lymphadenopathy *Exclusion of infection*	No=0 Low=4 Moderate=8 High=12	Absence of the following features Lymphadenopathy ≥1 cm in any nodal region or ≥2 cm in inguinal region Lymphadenopathy ≥2 cm in any nodal region or ≥3 cm in inguinal region, and/or splenomegaly (clinically palpable or assessed by imaging) Current malignant B-cell proliferative disorder*
Glandular *Exclusion of stone or infection*	No=0 Low=2 Moderate=4	Absence of glandular swelling Small glandular swelling with enlarged parotid (≤3 cm), or limited submandibular or lachrymal swelling Major glandular swelling with enlarged parotid (>3 cm), or important submandibular or lachrymal swelling
Articular *Exclusion of osteoarthritis*	No=0 Low=2 Moderate=4 High=6	Absence of currently active articular involvement Arthralgias in hands, wrists, ankles and feet accompanied by morning stiffness (>30 min) 1–5 (of 28 total count) synovitis ≥6 (of 28 total count) synovitis
Cutaneous *Rate as 'No activity' stable long-lasting features related to damage*	No=0 Low=3 Moderate=6 High=9	Absence of currently active cutaneous involvement Erythema multiforma Limited cutaneous vasculitis, including urticarial vasculitis, or purpura limited to feet and ankle, or subacute cutaneous lupus Diffuse cutaneous vasculitis, including urticarial vasculitis, or diffuse purpura, or ulcers related to vasculitis
Pulmonary *Rate as 'No activity' stable long-lasting features related to damage, or respiratory involvement not related to the disease (tobacco use, etc.)*	No=0 Low=5 Moderate=10 High=15	Absence of currently active pulmonary involvement Persistent cough or bronchial involvement with no radiographic abnormalities on radiography Or radiological or HRCT evidence of interstitial lung disease with: No breathlessness and normal lung function test. Moderately active pulmonary involvement, such as interstitial lung disease shown by HRCT with shortness of breath on exercise (NHYA II) or abnormal lung function tests restricted to: 70%>DL_{CO}≥ 40% or 80%>FVC≥60% Highly active pulmonary involvement, such as interstitial lung disease shown by HRCT with shortness of breath at rest (NHYA III, IV) or with abnormal lung function tests: DL_{CO}< 40% or FVC<60%
Renal *Rate as 'No activity' stable long-lasting features related to damage, and renal involvement not related to the disease. If biopsy has been performed, please rate activity based on histological features first*	No=0 Low=5 Moderate=10 High=15	Absence of currently active renal involvement with proteinuria <0.5 g/day, no haematuria, no leucocyturia, no acidosis, or long-lasting stable proteinuria due to damage Evidence of mild active renal involvement, limited to tubular acidosis without renal failure or glomerular involvement with proteinuria (between 0.5 and 1 g/day) and without haematuria or renal failure (GFR ≥60 mL/min) Moderately active renal involvement, such as tubular acidosis with renal failure (GFR <60 mL/min) or glomerular involvement with proteinuria between 1 and 1.5 g/day and without haematuria or renal failure (GFR ≥60 mL/min) or histological evidence of extra-membranous glomerulonephritis or important interstitial lymphoid infiltrate Highly active renal involvement, such as glomerular involvement with proteinuria >1.5 g/day or haematuria or renal failure (GFR <60 mL/min), or histological evidence of proliferative glomerulonephritis or cryoglobulinemia related renal involvement
Muscular *Exclusion of weakness due to corticosteroids*	No=0 Low=6 Moderate=12 High=18	Absence of currently active muscular involvement Mild active myositis shown by abnormal EMG or biopsy with no weakness and creatine kinase (N<CK≤2N) Moderately active myositis proven by abnormal EMG or biopsy with weakness (maximal deficit of 4/5), or elevated creatine kinase (2N <CK ≤4N), Highly active myositis shown by abnormal EMG or biopsy with weakness (deficit ≤3/5) or elevated creatine kinase (>4N)

Continued

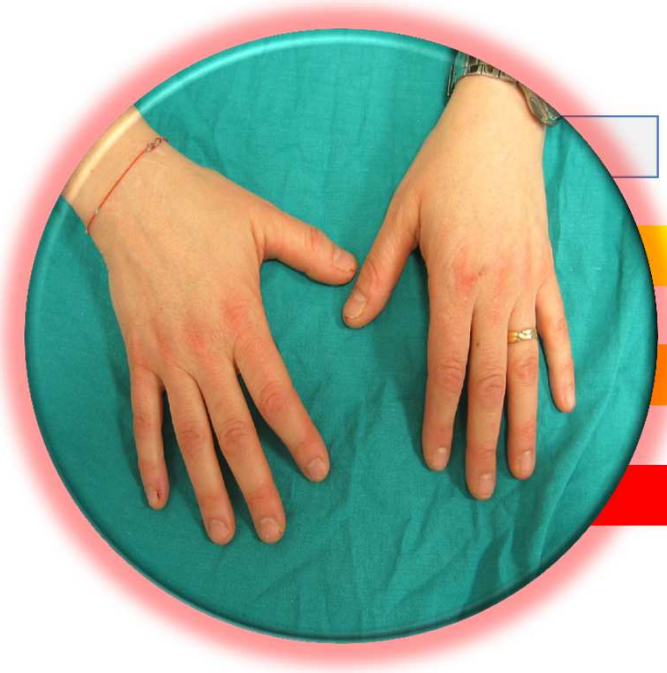
Table 1 Continued

Domain	Activity level	Description
PNS *Rate as 'No activity' stable long-lasting features related to damage or PNS involvement not related to the disease*	No=0 Low=5 Moderate=10 High=15	Absence of currently active PNS involvement Mild active peripheral nervous system involvement, such as pure sensory axonal polyneuropathy shown by NCS or trigeminal (V) neuralgia Moderately active peripheral nervous system involvement shown by NCS, such as axonal sensory-motor neuropathy with maximal motor deficit of 4/5, pure sensory neuropathy with presence of cryoglobulinemic vasculitis, ganglionopathy with symptoms restricted to mild/moderate ataxia, inflammatory demyelinating polyneuropathy (CIDP) with mild functional impairment (maximal motor deficit of 4/5or mild ataxia), Or cranial nerve involvement of peripheral origin (except trigeminal (V) neuralgia) Highly active PNS involvement shown by NCS, such as axonal sensory-motor neuropathy with motor deficit ≤3/5, peripheral nerve involvement due to vasculitis (mononeuritis multiplex, etc), severe ataxia due to ganglionopathy, inflammatory demyelinating polyneuropathy (CIDP) with severe functional impairment: motor deficit ≤3/5 or severe ataxia
CNS *Rate as 'No activity' stable long-lasting features related to damage or CNS involvement not related to the disease*	No=0 Moderate=10 High=15	Absence of currently active CNS involvement Moderately active CNS features, such as cranial nerve involvement of central origin, optic neuritis or multiple sclerosis-like syndrome with symptoms restricted to pure sensory impairment or proven cognitive impairment Highly active CNS features, such as cerebral vasculitis with cerebrovascular accident or transient ischaemic attack, seizures, transverse myelitis, lymphocytic meningitis, multiple sclerosis-like syndrome with motor deficit.
Haematological *For anaemia, neutropenia, and thrombopenia, only autoimmune cytopenia must be considered* *Exclusion of vitamin or iron deficiency, drug-induced cytopenia*	No=0 Low=2 Moderate=4 High=6	Absence of autoimmune cytopenia Cytopenia of autoimmune origin with neutropenia (1000<neutrophils<1500/mm^3), and/or anaemia (10<haemoglobin<12 g/dL), and/or thrombocytopenia (100 000<platelets<150 000/mm^3) Or lymphopenia (500<lymphocytes<1000/mm^3) Cytopenia of autoimmune origin with neutropenia (500≤neutrophils≤1000/mm^3), and/or anaemia (8≤haemoglobin≤10 g/dL), and/or thrombocytopenia (50 000≤platelets≤100 000/mm^3) Or lymphopenia (≤500/mm^3) Cytopenia of autoimmune origin with neutropenia (neutrophils <500/mm^3), and/or or anaemia (haemoglobin <8 g/dL) and/or thrombocytopenia (platelets <50 000/mm^3)
Biological	No=0 Low=1 Moderate=2	Absence of any of the following biological features Clonal component and/or hypocomplementemia (low C4 or C3 or CH50) and/or hypergammaglobulinemia or high IgG level between 16 and 20 g/L Presence of cryoglobulinemia and/or hypergammaglobulinemia or high IgG level >20 g/L, and/or recent onset hypogammaglobulinemia or recent decrease of IgG level (<5 g/L)

*Defined as indolent not treated lymphoma or currently treated lymphoma or myeloma (or treatment ended from less than 6 months). Do not rate past treated lymphoma or myeloma in complete remission.
CIDP, chronic inflammatory demyelinating polyneuropathy; CK, creatine kinase; CNS, central nervous system; DLCO, diffusing CO capacity; EMG, electromyogram; FVC, forced vital capacity; GFR, glomerular filtration rate; Hb, haemoglobin; HRCT, high-resolution CT; IgG, immunoglobulin G; NCS, nerve conduction studies; NHYA, New York Heart Association Classification; Plt, platelet; PNS, peripheral nervous system.

Rango de puntuación total de 0 a 123

4 niveles actividad (no, bajo, moderado, alto)



0 No actividad

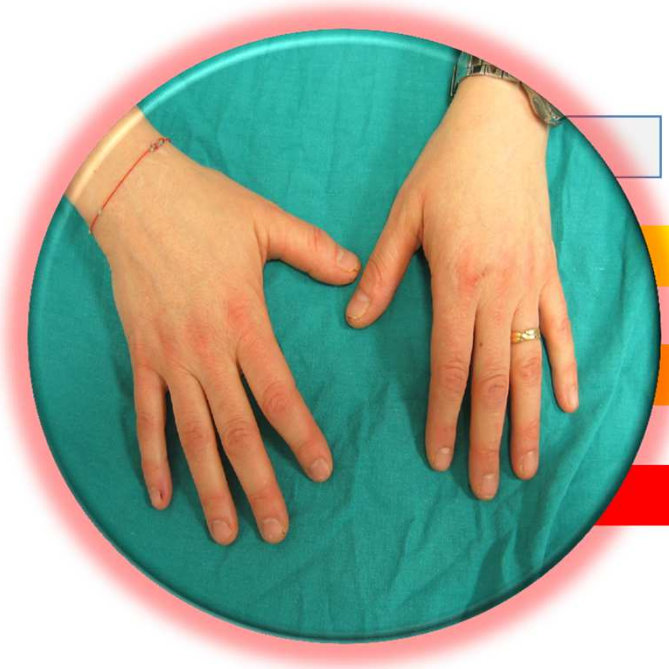
1 Baja/leve actividad

2 Moderada actividad

3 Elevada actividad

4 niveles actividad (no, bajo, moderado, alto)

EJEMPLO: Dominio articular



0 No afectación articular

1 Artralgias y rigidez matutina > 30 min

2 Artritis en 1-5 articulaciones

3 Artritis en >6 artic.

Criterios de exclusión: artrosis

Table 1 The EULAR Sjögren's Syndrome Disease Activity Index (ESSDAI): domain and item definitions and scores

Domain	Activity level	Description
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Glandular *Exclusion of stone or infection*	No=0 Low=2 Moderate=4	Absence of glandular swelling Small glandular swelling with enlarged parotid (≤3 cm), or limited submandibular or lachrymal swelling Major glandular swelling with enlarged parotid (>3 cm), or important submandibular or lachrymal swelling
Articular *Exclusion of osteoarthritis*	No=0 Low=2 Moderate=4 High=6	Absence of currently active articular involvement Arthralgias in hands, wrists, ankles and feet accompanied by morning stiffness (>30 min) 1–5 (of 28 total count) synovitis ≥6 (of 28 total count) synovitis
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Renal *Rate as 'No activity' stable long-lasting features related to damage, and renal involvement not related to the disease. If biopsy has been performed, please rate activity based on histological features first*	No=0 Low=5 Moderate=10 High=15	Absence of currently active renal involvement with proteinuria <0.5 g/day, no haematuria, no leucocyturia, no acidosis, or long-lasting stable proteinuria due to damage Evidence of mild active renal involvement, limited to tubular acidosis without renal failure or glomerular involvement with proteinuria (between 0.5 and 1 g/day) and without haematuria or renal failure (GFR ≥60 mL/min) Moderately active renal involvement, such as tubular acidosis with renal failure (GFR <60 mL/min) or glomerular involvement with proteinuria between 1 and 1.5 g/day and without haematuria or renal failure (GFR ≥60 mL/min) or histological evidence of extra-membranous glomerulonephritis or important interstitial lymphoid infiltrate Highly active renal involvement, such as glomerular involvement with proteinuria >1.5 g/day or haematuria or renal failure (GFR <60 mL/min), or histological evidence of proliferative glomerulonephritis or cryoglobulinemia related renal involvement
Muscular *Exclusion of weakness due to corticosteroids*	No=0 Low=6 Moderate=12 High=18	Absence of currently active muscular involvement Mild active myositis shown by abnormal EMG or biopsy with no weakness and creatine kinase (N<CK≤2N) Moderately active myositis proven by abnormal EMG or biopsy with weakness (maximal deficit of 4/5), or elevated creatine kinase (2N <CK ≤4N), Highly active myositis shown by abnormal EMG or biopsy with weakness (deficit ≤3/5) or elevated creatine kinase (>4N)

Continued

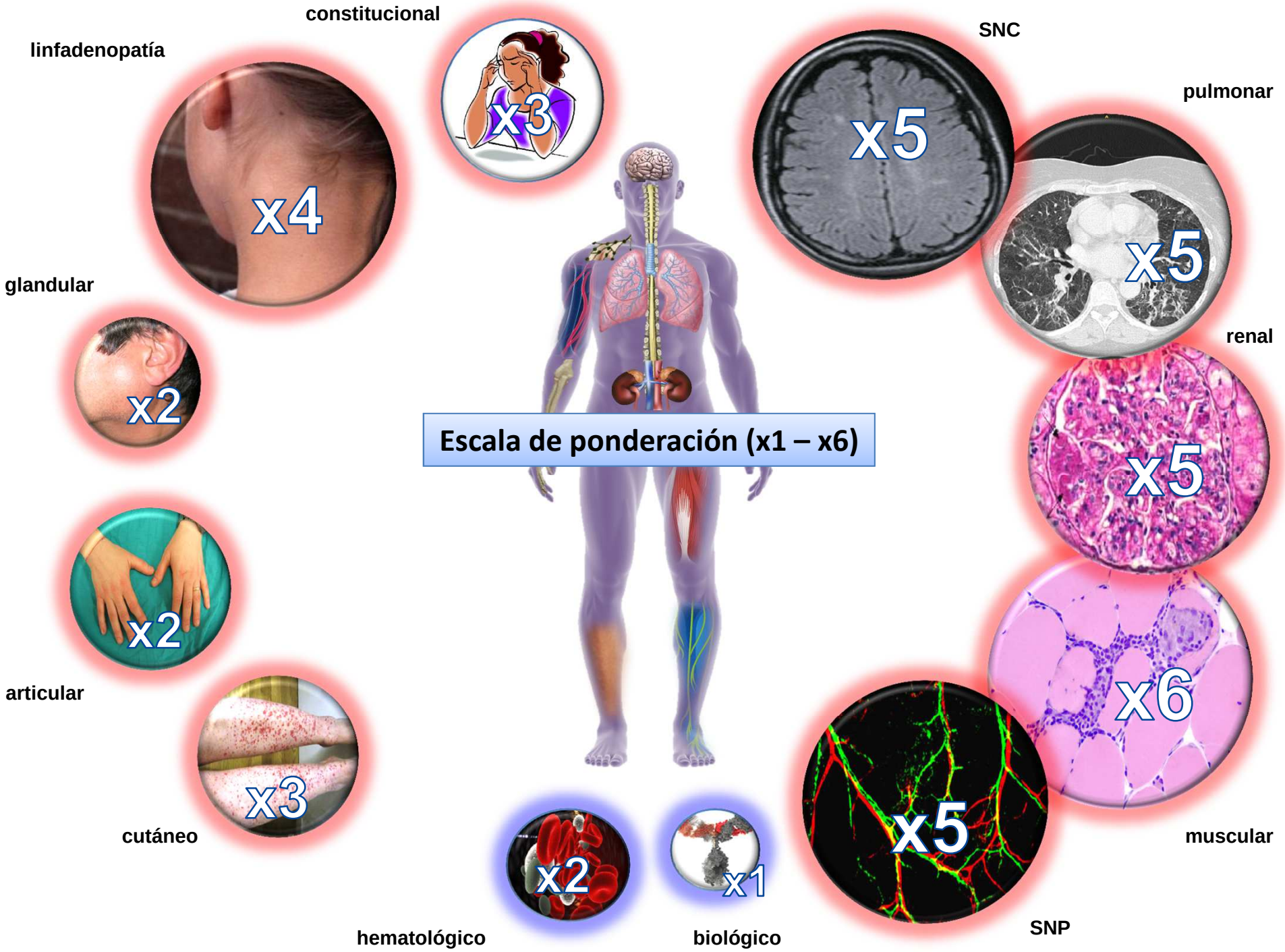
Table 1 Continued

Domain	Activity level	Description
PNS *Rate as 'No activity' stable long-lasting features related to damage or PNS involvement not related to the disease*	No=0 Low=5 Moderate=10 High=15	Absence of currently active PNS involvement Mild active peripheral nervous system involvement, such as pure sensory axonal polyneuropathy shown by NCS or trigeminal (V) neuralgia Moderately active peripheral nervous system involvement shown by NCS, such as axonal sensory-motor neuropathy with maximal motor deficit of 4/5, pure sensory neuropathy with presence of cryoglobulinemic vasculitis, ganglionopathy with symptoms restricted to mild/moderate ataxia, inflammatory demyelinating polyneuropathy (CIDP) with mild functional impairment (maximal motor deficit of 4/5or mild ataxia), Or cranial nerve involvement of peripheral origin (except trigeminal (V) neuralgia) Highly active PNS involvement shown by NCS, such as axonal sensory-motor neuropathy with motor deficit ≤3/5, peripheral nerve involvement due to vasculitis (mononeuritis multiplex, etc), severe ataxia due to ganglionopathy, inflammatory demyelinating polyneuropathy (CIDP) with severe functional impairment: motor deficit ≤3/5 or severe ataxia
CNS *Rate as 'No activity' stable long-lasting features related to damage or CNS involvement not related to the disease*	No=0 Moderate=10 High=15	Absence of currently active CNS involvement Moderately active CNS features, such as cranial nerve involvement of central origin, optic neuritis or multiple sclerosis-like syndrome with symptoms restricted to pure sensory impairment or proven cognitive impairment Highly active CNS features, such as cerebral vasculitis with cerebrovascular accident or transient ischaemic attack, seizures, transverse myelitis, lymphocytic meningitis, multiple sclerosis-like syndrome with motor deficit.
Haematological *For anaemia, neutropenia, and thrombopenia, only autoimmune cytopenia must be considered* *Exclusion of vitamin or iron deficiency, drug-induced cytopenia*	No=0 Low=2 Moderate=4 High=6	Absence of autoimmune cytopenia Cytopenia of autoimmune origin with neutropenia (1000<neutrophils<1500/mm^3), and/or anaemia (10<haemoglobin<12 g/dL), and/or thrombocytopenia (100 000<platelets<150 000/mm^3) Or lymphopenia (500<lymphocytes<1000/mm^3) Cytopenia of autoimmune origin with neutropenia (500≤neutrophils≤1000/mm^3), and/or anaemia (8≤haemoglobin≤10 g/dL), and/or thrombocytopenia (50 000≤platelets≤100 000/mm^3) Or lymphopenia (≤500/mm^3) Cytopenia of autoimmune origin with neutropenia (neutrophils <500/mm^3), and/or or anaemia (haemoglobin <8 g/dL) and/or thrombocytopenia (platelets <50 000/mm^3)
Biological	No=0 Low=1 Moderate=2	Absence of any of the following biological features Clonal component and/or hypocomplementemia (low C4 or C3 or CH50) and/or hypergammaglobulinemia or high IgG level between 16 and 20 g/L Presence of cryoglobulinemia and/or hypergammaglobulinemia or high IgG level >20 g/L, and/or recent onset hypogammaglobulinemia or recent decrease of IgG level (<5 g/L)

*Defined as indolent not treated lymphoma or currently treated lymphoma or myeloma (or treatment ended from less than 6 months). Do not rate past treated lymphoma or myeloma in complete remission.

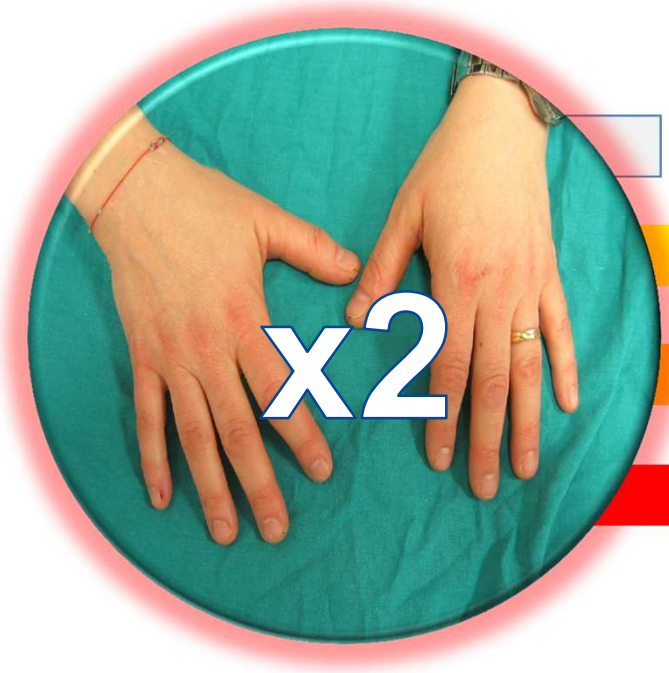
CIDP, chronic inflammatory demyelinating polyneuropathy; CK, creatine kinase; CNS, central nervous system; DLCO, diffusing CO capacity; EMG, electromyogram; FVC, forced vital capacity; GFR, glomerular filtration rate; Hb, haemoglobin; HRCT, high-resolution CT; IgG, immunoglobulin G; NCS, nerve conduction studies; NHYA, New York Heart Association Classification; Plt, platelet; PNS, peripheral nervous system.

Escala de ponderación de 1 a 6


constitucional
x3
linfadenopatía
x4
SNC
x5
pulmonar
x5
glandular
x2
renal
x5
Escala de ponderación (x1 – x6)
articular
x2
x6
cutáneo
x3
x5
muscular
hematológico
x2
biológico
x1
SNP

Puntuación final de cada dominio

EJEMPLO: Dominio articular



0 No afectación articular

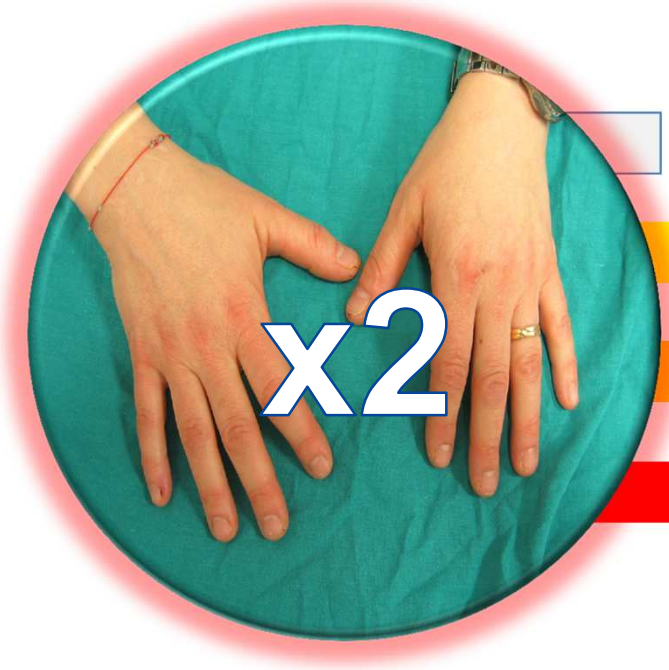
1 Artralgias y rigidez matutina > 30 min

2 Artritis en 1-5 articulaciones

3 Artritis en >6 artic.

Puntuación final de cada dominio

EJEMPLO: Dominio articular



0 No afectación articular

2 Artralgias y rigidez matutina > 30 min

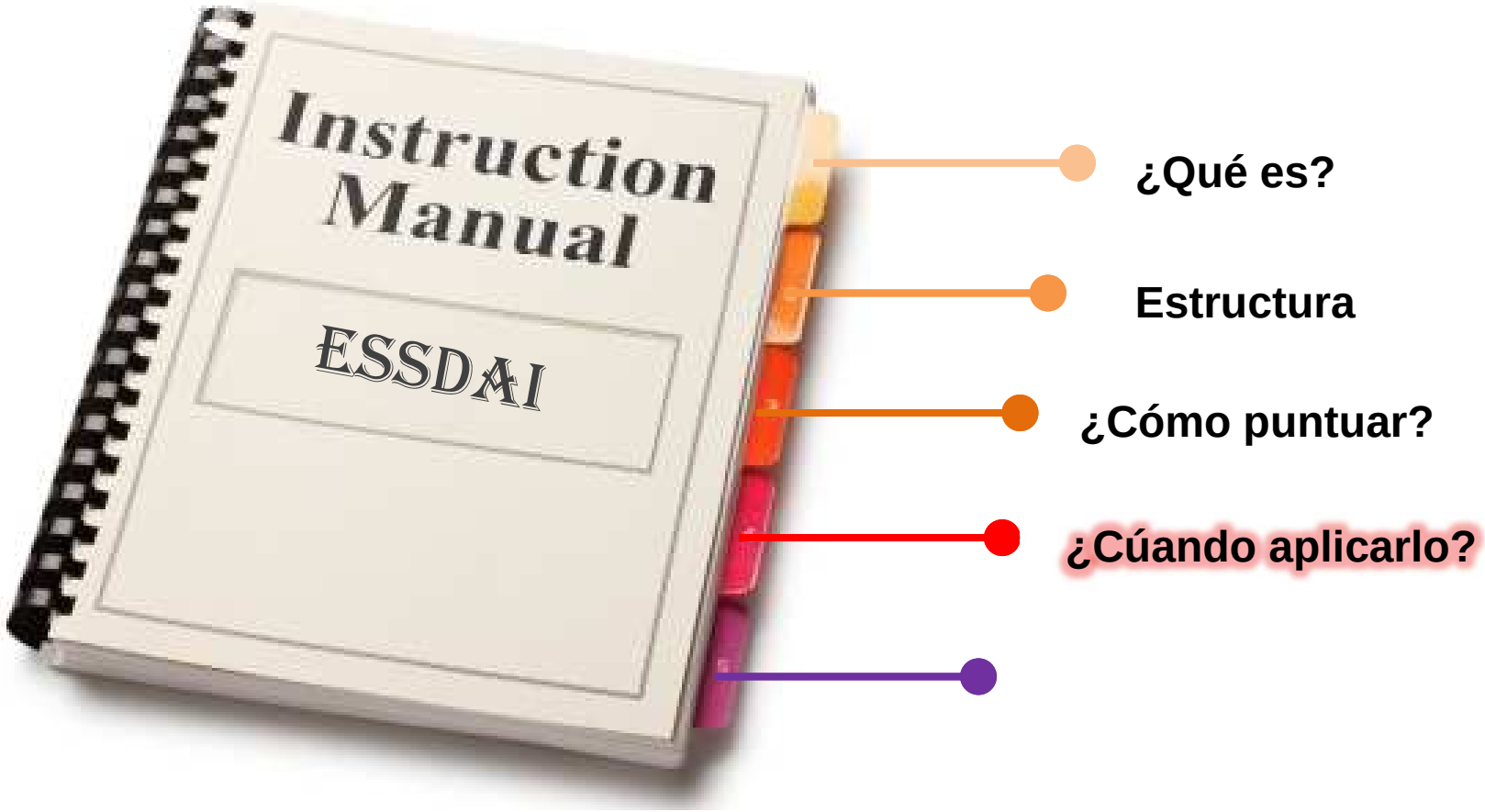
4 Artritis en 1-5 articulaciones

6 Artritis en >6 artic.

ARD Online First, published on January 17, 2014 as 10.1136/annrheumdis-2013-204615

EXTENDED REPORT

Validation of EULAR primary Sjögren's syndrome disease activity (ESSDAI) and patient indexes (ESSPRI)



¿Cuándo aplicarlo?

Cumplimiento criterios del 2002

Diagnóstico del síndrome de Sjögren

Última visita

ACTIVIDAD AL DIAGNÓSTICO DEL SJÖGREN

NUEVA ACTIVIDAD

ACTIVIDAD ACUMULADA

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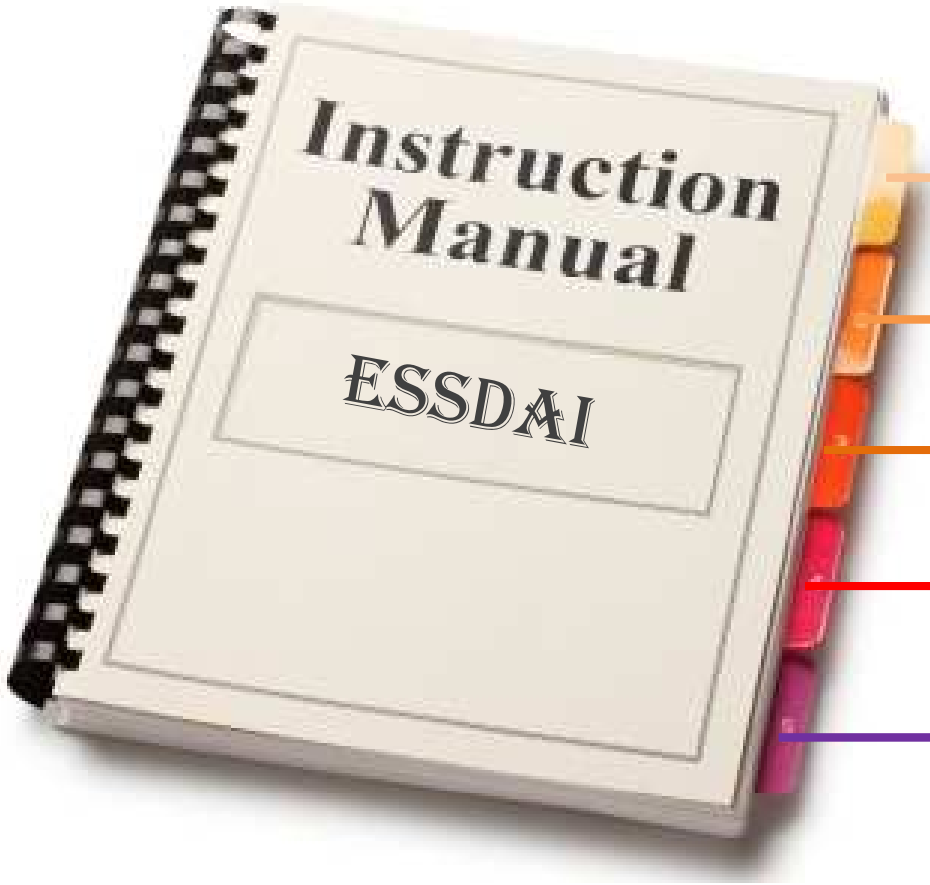

- ¿Qué es?
- Estructura
- ¿Cómo puntuar?
- ¿Cúando aplicarlo?
- Interés práctico

Interés práctico

A. Caracterización de la actividad sistémica
   1. ¿Cómo se distribuye en el tiempo?
   2. ¿Cómo se distribuye órgano por órgano?
   3. ¿Qué marcadores al dx se asocian con el desarrollo de actividad?
B. Instrumento de predicción pronóstica
   1. Papel de la puntuación global basal
   2. Actividad basal órgano por órgano
   3. Papel del nivel de actividad basal (alta actividad en al menos 1 órgano)
   4. Papel de la graduación de la actividad global basal (1-4, 5-13, >13)

RHEUMATOLOGY

Rheumatology 2014;53:321–331
doi:10.1093/rheumatology/ket349
Advance Access publication 25 October 2013

Original article

Systemic involvement in primary Sjögren's syndrome evaluated by the EULAR-SS disease activity index: analysis of 921 Spanish patients (GEAS-SS Registry)

Manuel Ramos-Casals[1], Pilar Brito-Zerón[1], Roser Solans[2], María-Teresa Camps[3], Arnau Casanovas[4], Bernardo Sopeña[5], Bernardino Díaz-López[6], Francisco-Javier Rascón[7], Rami Qanneta[8], Guadalupe Fraile[9], Roberto Pérez-Alvarez[10], José-Luis Callejas[11], Mar Ripoll[12], Blanca Pinilla[13], Miriam Akasbi[14], Eva Fonseca[15], Jesús Canora[16], María-Elvira Nadal[17], Gloria de la Red[18], Inés Fernández-Regal[19], Iratxe Jiménez-Heredia[20], Josep-Angel Bosch[2], María-del-Mar Ayala[3], Lluisa Morera-Morales[4], Brenda Maure[5], Arantxa Mera[6], Marc Ramentol[2], Soledad Retamozo[1], Belchin Kostov[21] and on behalf of the SS Study Group and Autoimmune Diseases Study Group (GEAS) of the Spanish Society of Internal Medicine (SEMI)*

Systemic activity and mortality in primary Sjögren syndrome

Predicting survival using the EULAR-SS disease activity index (ESSDAI) in 1045 patients

P. Brito-Zerón[1*], B. Kostov[1,12], R. Solans[2], G. Fraile[3], C. Suárez-Cuervo [4], A. Casanovas[5], F.J. Rascón[6], R. Qanneta[7], R. Pérez-Alvarez[8], M. Ripoll[9], M. Akasbi[10], B. Pinilla[11], J.A. Bosch[2], J. Nava-Mateos[3], B. Díaz-López [4], M.L. Morera-Morales[5], H. Gheitasi[1], S. Retamozo[1], M. Ramos-Casals[1], on behalf of the SS Study Group, Autoimmune Diseases Study Group (GEAS), Spanish Society of Internal Medicine (SEMI)**

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A1. ¿Cómo se distribuye en el tiempo?

Cumplimiento criterios del 2002

Diagnóstico del síndrome de Sjögren

Seguimiento (75 meses)

Última visita

ACTIVIDAD BASAL

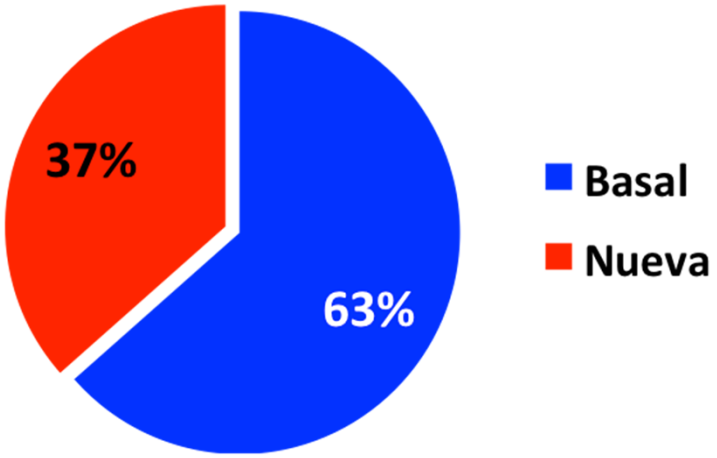
Media ESSDAI

5.81

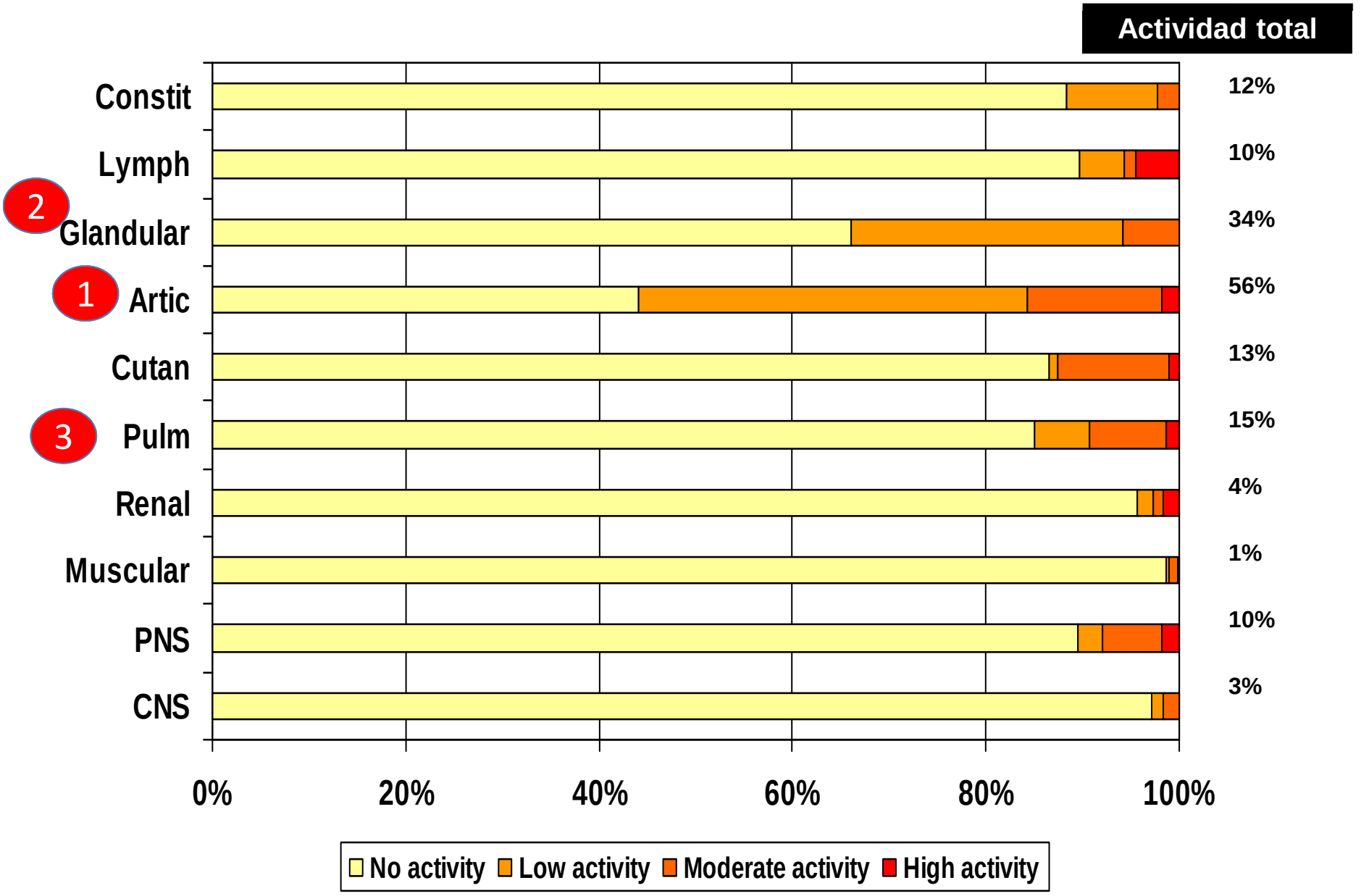
Nueva actividad

Media ESSDAI

3.34

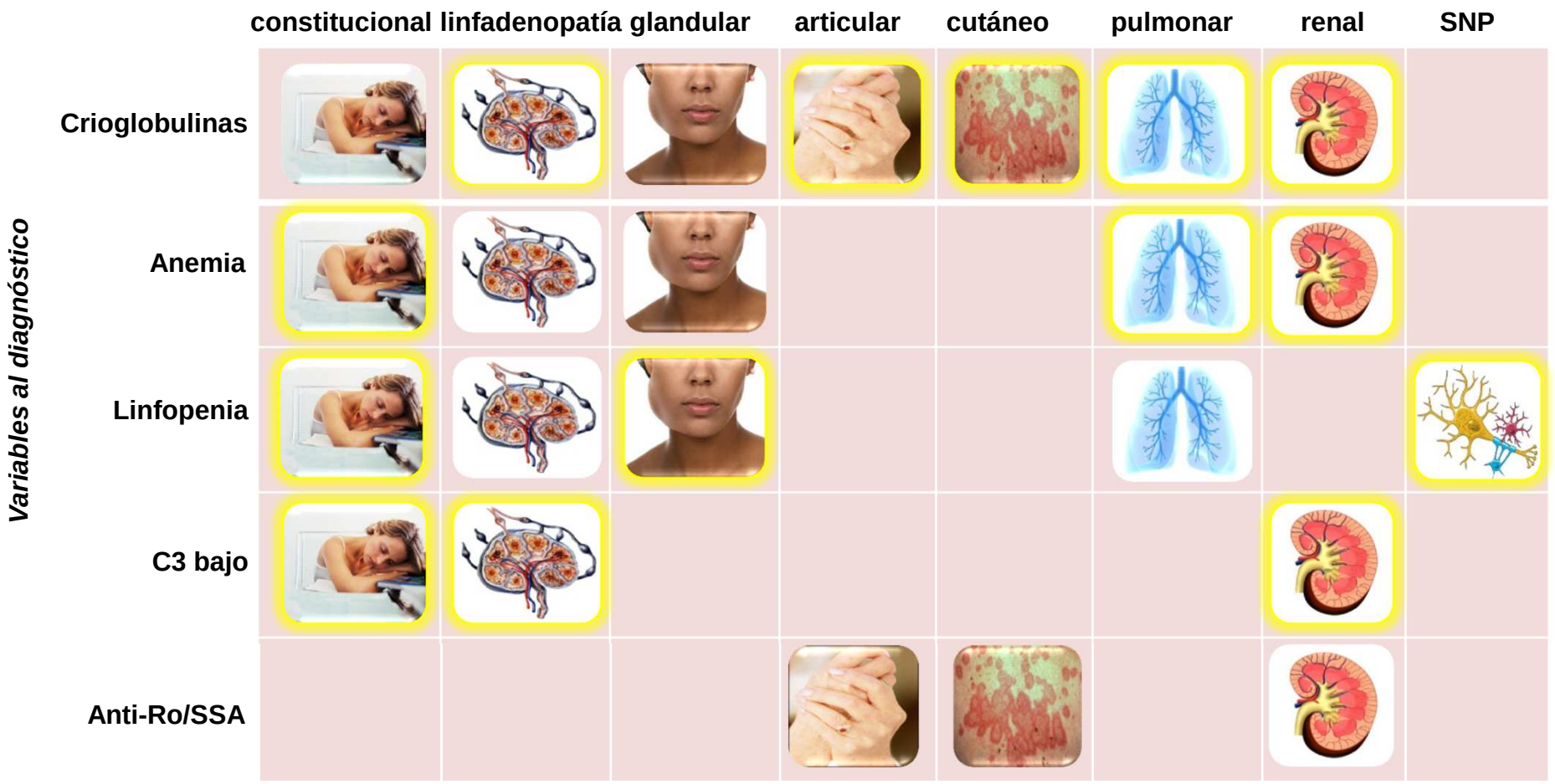


A2. ¿Cómo se distribuye órgano por órgano?



A3. ¿Qué marcadores al diagnóstico se asocian con el desarrollo de actividad?

Actividad acumulada por dominio



Interés práctico

A. Caracterización de la actividad sistémica
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B. Instrumento de predicción pronóstica
   1. **Papel de la puntuación global basal**
   2. **Actividad basal órgano por órgano**
   3. **Papel del nivel de actividad basal (alta actividad en al menos 1 órgano)**
   4. **Papel de la graduación de la actividad global basal (1-4, 5-13, >13)**

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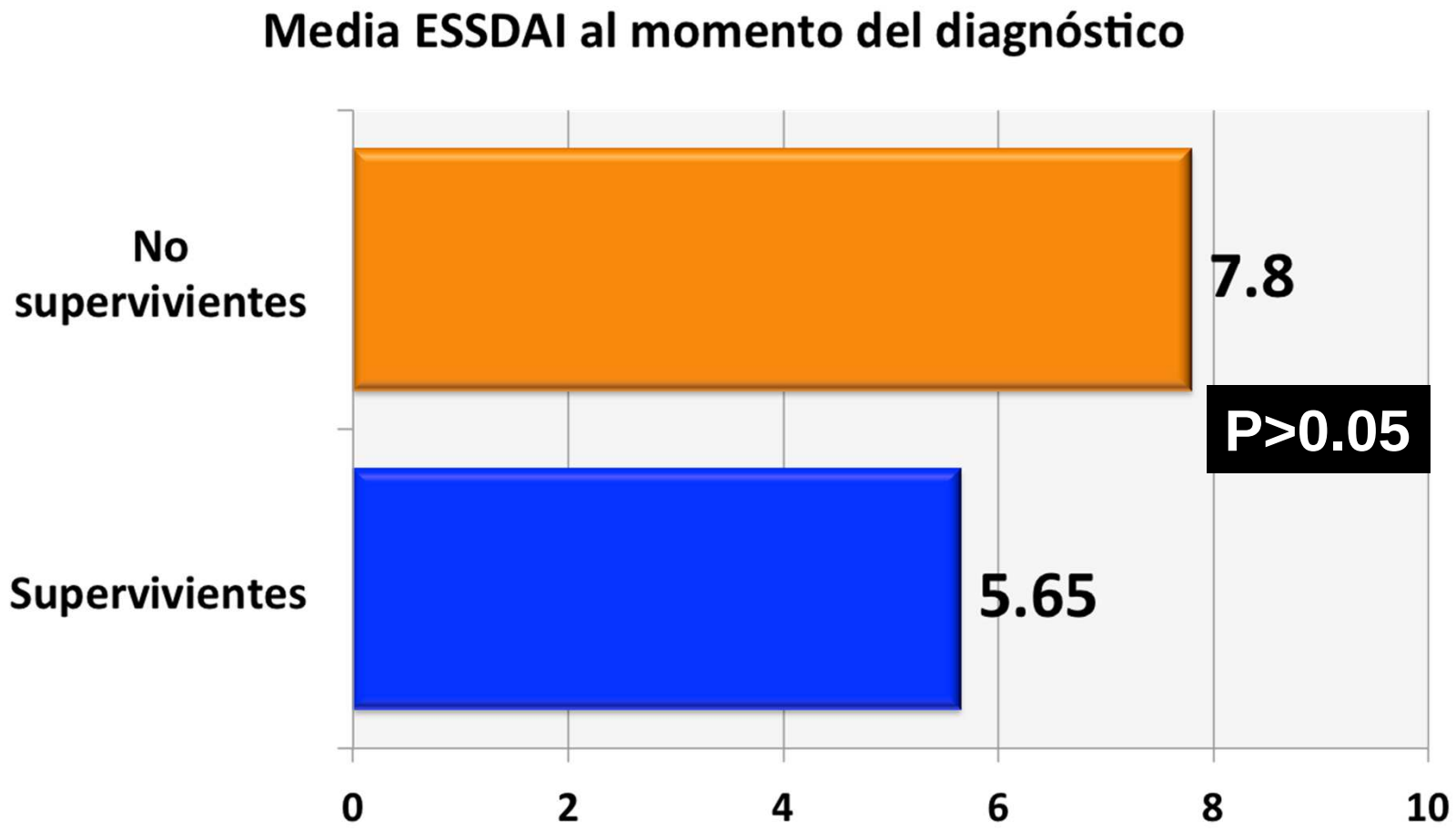
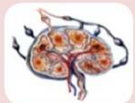
B1. Papel de la puntuación global basal

La media en la puntuación del ESSDAI al diagnóstico en toda la cohorte fue de 5.89



B2. Actividad basal órgano por órgano y mortalidad

Dominios ESSDAI asociados con mayor mortalidad

DOMINIOS ESSDAI	Constitucional	Hematológico	Biológico	Glandular	Linfadenopatía	Articular	Cutáneo	Pulmonar	Renal	SNP	SNC
											
Grado de actividad	score > 1		score > 1					score > 1			

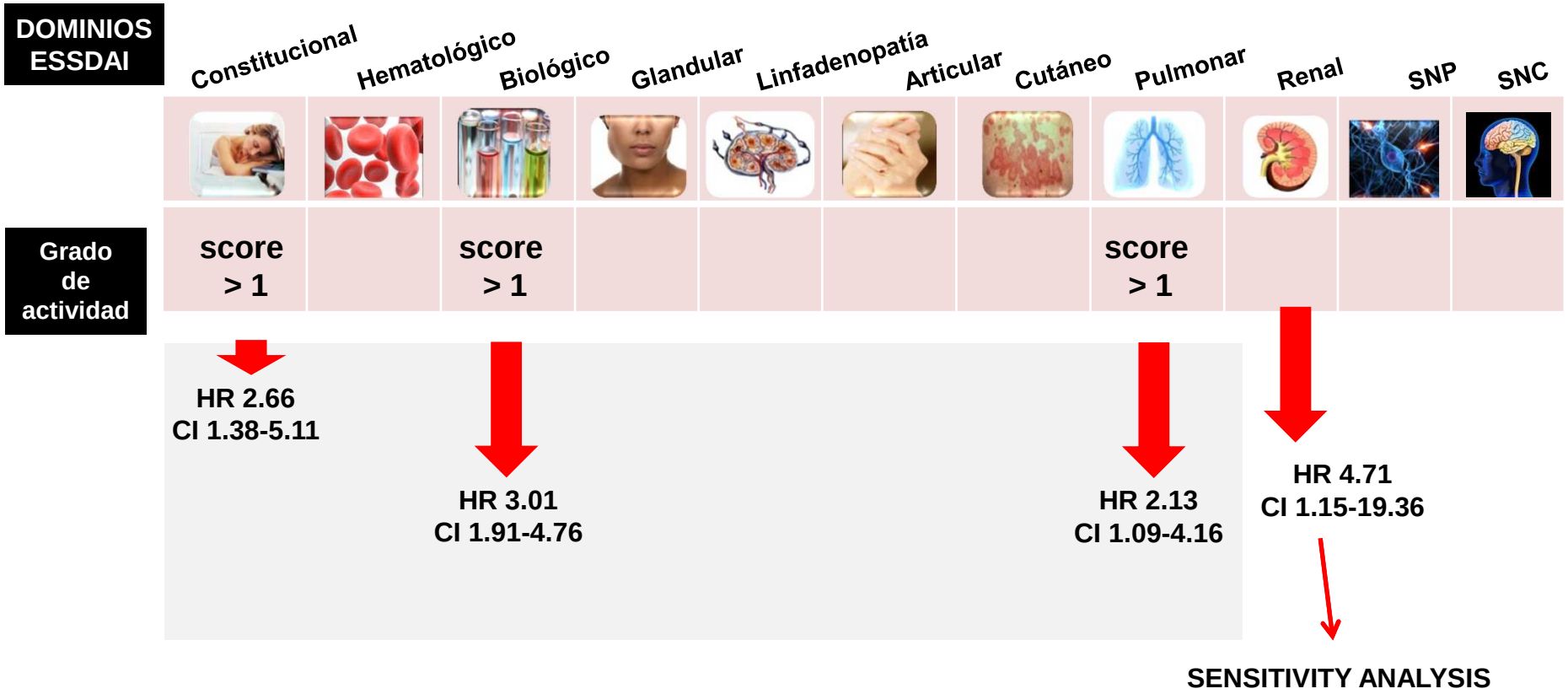
HR 2.66
CI 1.38-5.11

HR 3.01
CI 1.91-4.76

HR 2.13
CI 1.09-4.16

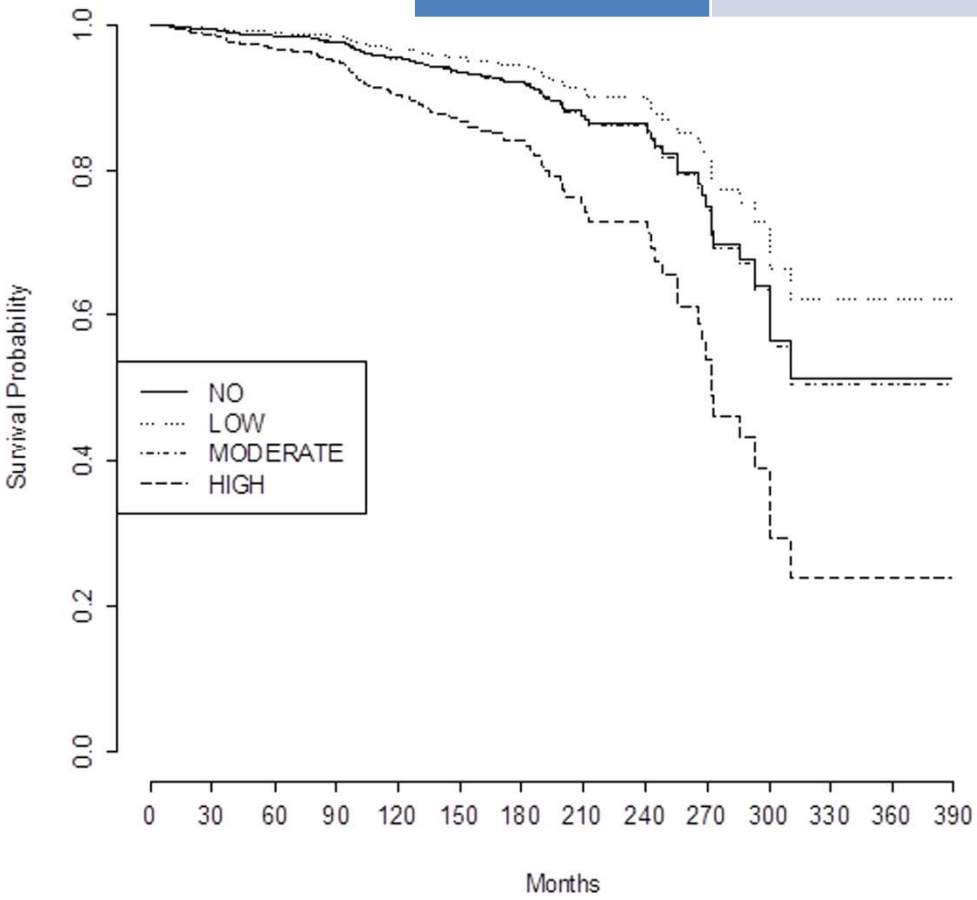
B2. Actividad basal órgano por órgano y mortalidad

Dominios ESSDAI asociados con mayor mortalidad



B3. Papel del nivel de actividad basal (alta actividad en al menos 1 órgano)

Follow-up (years)	5 years	10 years	20 years	30 years
Survival rate (%)				
- No activity	98.4	95.3	86.3	51.4
- Low activity	98.8	96.6	90.0	62.1
- Moderate	98.3	95.2	86.0	50.7
- High activity	96.5	90.3	72.9	24.0



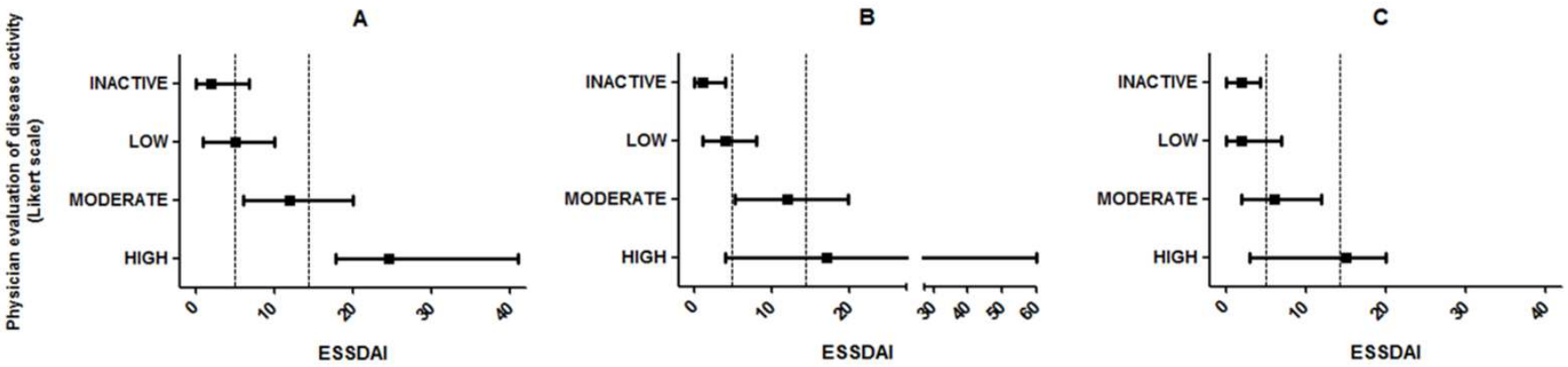
Mayor mortalidad si al dx hay al menos un dominio ESSDAI de alta actividad

P=0.011

Defining disease activity sates and meaningful differences in primary Sjögren's Syndrome with EULAR primary Sjögren's Syndrome disease activity (ESSDAI) and patient reported indexes (ESSPRI)

Raphaèle Seror (1,2), Hendrika Bootsma (3), Alain Saraux (4), Simon J Bowman (5), Elke Theander (6), Johan G Brun (7), Gabriel Baron (2), Véronique Leguern (8), Valérie Devauchelle-Pensec (4), Manel Ramos-Casals (9), Valeria Valim (10), Thomas Dörner (11, Athanasios Tzioufas (12, Jacques-Eric Gottenberg (13), Roser Solans Laqué (14), Thomas Mandl (6), Eric Hachulla (15), Kathy L. Sivils (16), Wan-Fai Ng (17), Anne-Laure Fauchais (18) Stefano Bombardieri (19), Guido Valesini (20), Elena Bartoloni (21), Vincent Goeb (22), Sonja Praprotnik (23), Takayuki Sumida (24), Sumusu Nishiyama (25), Roberto Caporali (26), Aike A Kruize (27), Cristina Vollenweider (28), Philippe Ravaud (2), Petra Meiners (3), Pilar Brito-Zerón (9), Claudio Vitali (29), Xavier Mariette (1) on behalf of the EULAR Sjögren's Task Force*

Figure 1. Distribution of ESSDAI score according to disease activity levels



In patients form EULAR cohort at inclusion (A), at 6 months (B) and in the ASSESS cohort (C).
Vertical dot lines represents disease activity thresholds of moderate activity (ESSDAI=5), and high activity (ESSDAI=14).
ESSDAI score distribution is represented with mean value (square) and interquartile range, in each subgroups of activity as assesse

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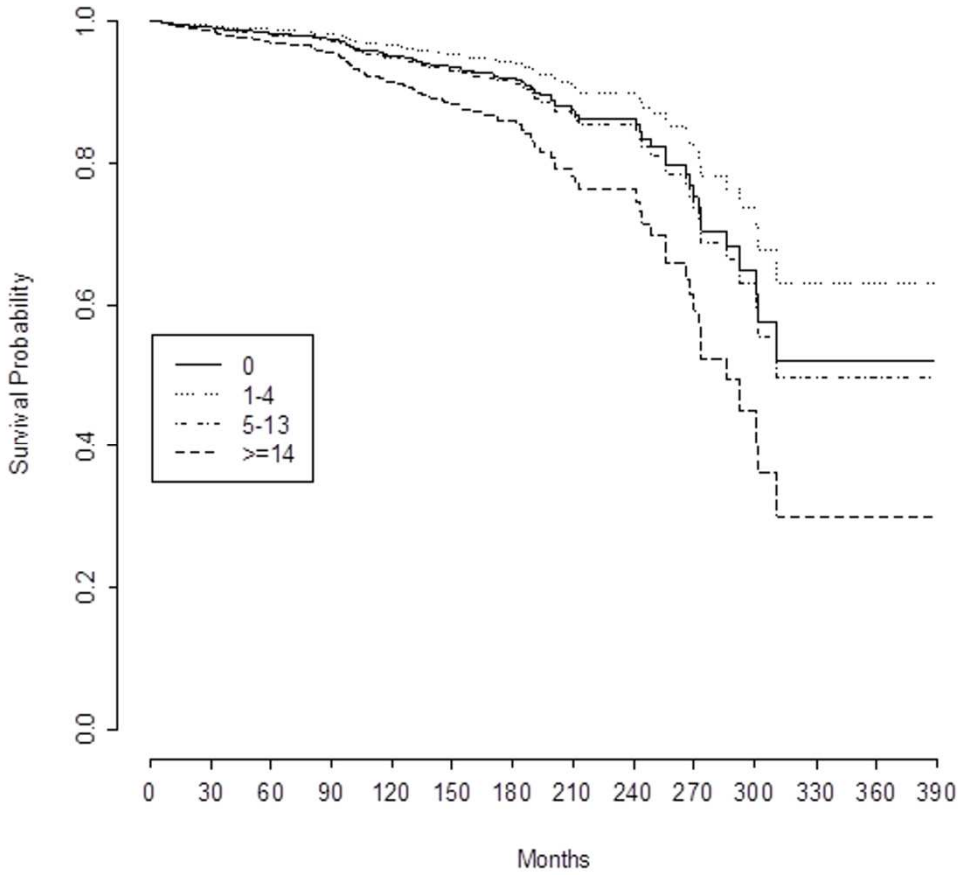
Actividad baja: puntuación global ESSDAI 1-4

Actividad moderada: puntuación global ESSDAI 5-13

Actividad alta: puntuación global ESSDAI >13

B4. Papel de la graduación de la actividad global basal (1-4, 5-13, >13)

Follow-up (years)	5 years	10 years	20 years	30 years
Survival rate (%)				
- ESSDAI = 0	98.4	95.4	86.4	52.2
- ESSDAI 1-4	98.9	96.7	90.2	63.2
- ESSDAI 5-13	98.3	95.0	85.5	49.8
- ESSDAI ≥14	97.1	91.6	76.3	30.1



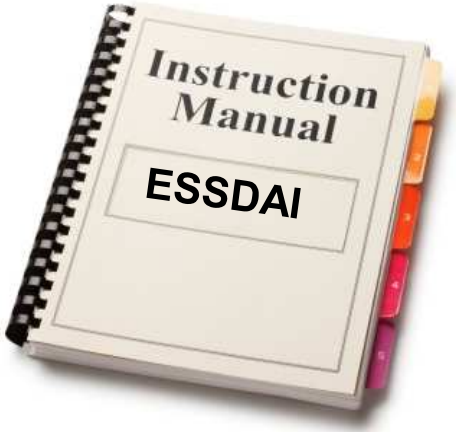
Mayor mortalidad si el paciente presenta al dx un indice ESSDAI superior a 13.

P=0.037


The Future
NEXT EXIT

- **EULAR ESSDAI Glossary**
- **EULAR evidence-based therapeutic guidelines for systemic Sjögren**

EULAR Glossary



EULAR Sjögren's syndrome disease activity index : A User Guide

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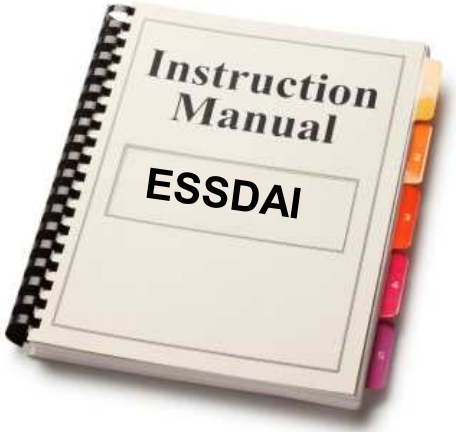
EULAR Glossary



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Inclusión de pacientes con otras enfermedades autoinmunes asociadas

Definir de forma clara la afectación de cada órgano

Definir los criterios de exclusión (afectación no autoinmune)

EULAR evidence-based therapeutic guidelines for systemic Sjögren

EULAR evidence-based therapeutic guidelines for systemic Sjögren

Participantes de 25 países

EULAR evidence-based therapeutic guidelines for systemic Sjögren

Coordinador del proyecto: Manel

Participantes de 25 países

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Systemic activity and mortality in primary Sjögren syndrome

Predicting survival using the EULAR-SS disease activity index (ESSDAI) in 1045 patients

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