Con la colaboración de

NOVARTIS

Sociedad Española de Reumatología

EULAR REVIEW

MADRID JUNIO 2017

Annual European Congress of Rheumatology

Enfermedades Autoinmunes Sistémicas. Clínica

Dra. Paloma García de la Peña

MADRID JUNIO 2017
EULAR

HOJA DE RUTA-RESUMEN VIERNES

RUTA DE VIAJE

- Sjögren
- Lupus eritematoso
- Esclerosis sistémica
- Vasculitis

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EULAR

Junio 2017

Se	Lun	Mar	Míe	Jue	Vie	Sáb	Dom
22				1	2	3	4
23	5	6	7	8	9	10	11
24	12	13	14	15	16	17	18
25	19	20	21	22	23	24	25
26	26	27	28	29	30		

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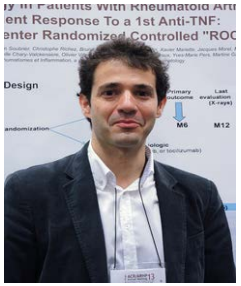
WIN & HOT SESSION 10:15-11:45



Jacques-Eric Gottenberg

SJÖGREN

Simon Bowman

- Nuevos Criterios diagnósticos
- Estandarización de las biopsias de glándula saliva
- Caracterización de los centros germinales con tinciones especiales CD20/CD21/CD23
- Inclusión de toda el área glandular
- Mejor definición de calidad de vida de los pacientes HRLQ
- Nuevos score de actividad ESSDAI/ESSPRI
- Etiopatogenia
  - Inmunidad Innata ¨la firma del ITF Tipo I”
  - Influencia viral-VIROMA (VEB/VHδ/reovirus)
  - Genética: polimorfismo/ 50% Firma ITF
  - Epigenética: hipometilación
- Inmunidad Adaptativa: CD8+ multiomic approach/MICAL / LB
- Cáncer y autoinmunidad
- Novedades en manifestaciones clínicas: neurológicas/renales/pulmón (AFECTACIÓN QUÍSTICA).
- Factores de riesgo para linfoma: ESSDAI/FR+/Histología de la GS (high focus Score/centros germinales)

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EULAR

POSTER TOUR 11:45-13:30
"NOVELTY IN THE CLINICAL APPROACH TO SLE, SJÖGREN´S AND APS II""

SJÖGREN

RI0256
ULTRASOUND CONSENSUS DEFINITIONS ON NORMAL AND ABNORMAL FINDINGS IN SALIVARY GLANDS IN SJÖGREN'S SYNDROME: RESULTS OF AN OMERACT DELPHI PROCESS.
S. Jousse-Joulin1,*, F. gandjbakhch2, S. ohrndorf3, M. backhaus3, G. tamborrini4, I. chary-valckenaere5, L. terslev6, A. iagnocco7, P. collado8, C. hernandez-diaz9, E. naredo8, W. schmidt3, G. filippou10, C. dejaco11, M. mortada12, A. hocevar13, S. chrysidis14, G. mardenly15, J. J. de agustin16, R. thiele17, D. Mac Carter18, S. finzel19, P. hanova20, C. glaser19, D. hammerfors21, M. A. d'agostino22, G. bruyn23

FRI0263
SYSTEMIC DISEASE ACTIVITY PROGRESSION IN A LARGE COHORT OF PRIMARY SJÖGREN'S SYNDROME: A LONG-TERM FOLLOW-UP DATA BASED ON THE ESSDAI SCORE
L. Quartuccio1,*, S. Zandonella Callegher1, S. Gandolfo1, C. Fabro1, S. De Vita1

Estudio retrospectivo- N=254-tiempo seguimiento medio de 9,1 años,

Table1 (% of agreement)	Definition of normal US findings	Procedure of scanning	Definition of abnormal US findings	SG to evaluate in pSS	Definition of scoring
Parotid glands	Uniformly echoic texture with a clear delineation from the superficial tissue. Tissue comparable to thyroid parenchyma (81%)	Longitudinal and transverse plane (90%)	Focal or diffuse an/hypoechoic areas (95%)	yes	4-grade semiquantitative scoring system (i.e. grade 0, normal parenchyma; grade 1, minimal change: grade 2, moderate: grade 3, severe: diffuse inhomogeneity occupying all the surface of the gland) (79%)
Submandibular glands	SMG are usually of finer granular echo texture compared to PG or to the normal thyroid parenchyma (89.1%)	Longitudinal and transverse plane (90%)	Idem PG(95%)	yes	Idem PG (79%)
Sublingual glands	SLG has no clear delineation from the superficial tissue because of no true fascial capsule(77%)	longitudinal and transverse plane (77%)	Idem PG(95%)	no	Not useful

Conclusions: We developed a consensual US definition of normal and abnormal USGS findings through a Delphi exercise process. This finding will be used as a basis to further proceed with the validation of USGS for clinical application.

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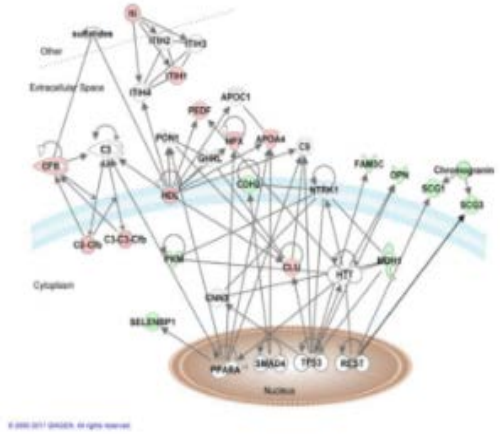
EULAR

CLINICAL SESSIONS 13:30-15:00
"BASIC AND TRASLATIONAL S SCIENCE SESSION"

SJÖGREN

OP0312
A PROTEOMIC SIGNATURE OF FATIGUE IN PRIMARY SJÖGREN'S SYNDROME
R. Omdal1,2,*, E. Larssen3,4, C. Brede5, A. Hjelle3, A. B. Tjensvoll6, K. B. Norheim2, K. Bårdsen3, K. Jonsdottir7, P. Ruoff8, M. M. Nilsen3,4

IPA= Ingenuty Pathway Analysis



LCR

- Apolipoproteina A4
- Hemopexina Factor derivado del epitelio pigmentario
- Secretogranina-3
- Proteína ligando del Selenio 1
- Proteína B del complemento

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CLINICAL SESSIONS 13:30-15:00
"EMBARAZO ENFERMEDADES REUMÁTICAS"

EMBARAZO-ENFERMEDADES REUMATOLÓGICAS

CONSEJO PRECONCEPCIONAL

Rebeca Fisher-Betz

No necesario suspender Metotrexato en los varones y controversia en cuanto al micofenolato

Laura Andreoli

- 24 Centros/24 centros
- 390 pacientes/320 niños
- 1/3 no han recibido consejo preconcepcional
- 50% han tenido menos hijos de lo deseado por miedo
- 3,7% de la descendencia desarrollan Enfermedades autoinmunes y 3,4% alteraciones en el desarrollo neurológico
- No alteración de la inteligencia/Sí alteraciones del aprendizaje

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POSTER TOUR 11:45-13:30
"NOVELTY IN THE CLINICAL APPROACH TO SLE, SJÖGREN´S AND APS II""

LUPUS ERITEMATOSO SISTÉMICO

FRI0257
URINARY ANGIOSTATIN, CXCL4 AND VCAM-1 AS BIOMARKERS FOR LUPUS NEPHRITIS
C. C. Mok1,*, S. Soliman2, L. Y. Ho1, F. Mohamed3, F. I. Mohammed3, C. Mohan2

Inhibidor endógeno de la angiogénesis

N=227 (80 LES no activos/67 LES activos sin NL/80 LES y NL)

EULAR PROYECTS IN CLINICAL AFFAIRS 13:30-15:00

"Biomarkers in cardiovascular rheumatology-state of the art"

OP0319
HIGH SENSITIVITY CARDIAC TROPONIN T IS A BIOMARKER FOR ATHEROSCLEROSIS IN SYSTEMIC LUPUS ERYTHEMATOUS PATIENTS: A CROSS-SECTIONAL CONTROLLED STUDY
G. Divard1, R. Abbas1, C. Chenevier-Gobeaux2, B. Escoubet1, M.-P. Chauveheid1, A. Dossier1, M. Dehoux1, T. Papo1, K. Sacre

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FRI0258

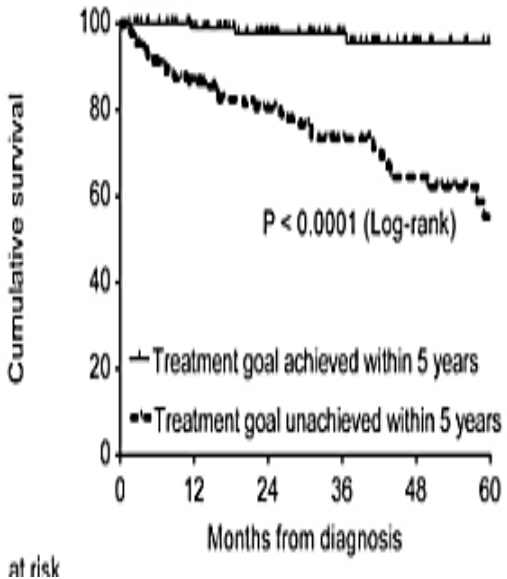
LONG-TERM PROGNOSIS OF PATIENTS WITH SYSTEMIC LUPUS ERYTHEMATOSUS-ASSOCIATED PULMONARY ARTERIAL HYPERTENSION: CSTAR-PAH COHORT STUDY

J. Qian1, M. Li2, X. Zhang3, Q. Wang2,*, J. Zhao2, Z. Tian4, X. Zeng2 on behalf of CSTAR on behalf of CSTAR group

N=310- Tiempo medio seguimiento=24 meses

Table 1 Baseline characteristics of patients with SLE-associated PAH

Characteristics	SLE-associated PAH (n = 310)
Age at recruitment, yr	35.0 ± 10.1
Female sex, %	99.4
WHO Fc I-II, %	51.7
6MWD, m	408.6 ± 98.0
NT-proBNP, pg/ml	1660.5 ± 2275.1
SLEDAI	6.1 ± 5.5
mPAP, mmHg	46.5 ± 12.1
CI, L/min × m^2	2.8 ± 0.9
RAP, mmHg	5.8 ± 5.6



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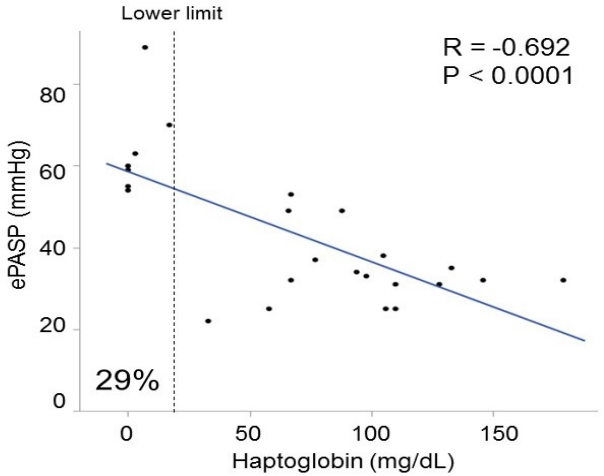
ESCLEROSIS SISTÉMICA/OTRAS
POSTER TOUR 11:45-13:30
"INDEXES AND PREDICTORS IN SYSTEMIC SCLEROSIS AND MYOSITIS""

FRI0364

USEFULNESS OF SERUM HAPTOGLOBIN LEVELS AS A NOVEL MARKER FOR PULMONARY ARTERIAL HYPERTENSION COMPLICATED WITH CONNECTIVE TISSUE DISEASE

H. Nakamura1,*, M. Kato1, M. Kono1, S. Tanimura1, R. Hisada1, E. Sugawara1, K. Ohmura1, S. Shimamura1, Y. Fujieda1, K. Oku1, T. Bohgaki1, O. Amengual1, S. Yasuda1, T. Atsumi1



Microangiopatía trombótica arterial pulmonar

N=24 pacientes/32 controles

8 ES/8EMTC/LES

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FRI0363

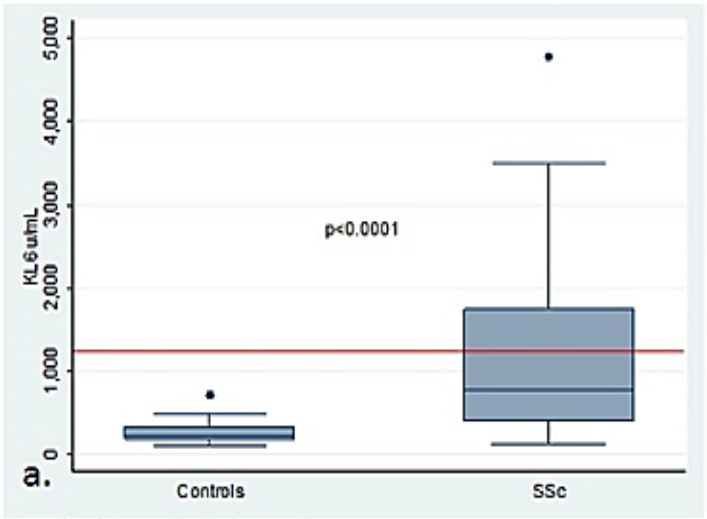
KL6 AND NOT CCL-18 IS A PREDICTOR OF EARLY PROGRESSION IN SYSTEMIC SCLEROSIS RELATED INTERSTITIAL LUNG DISEASE

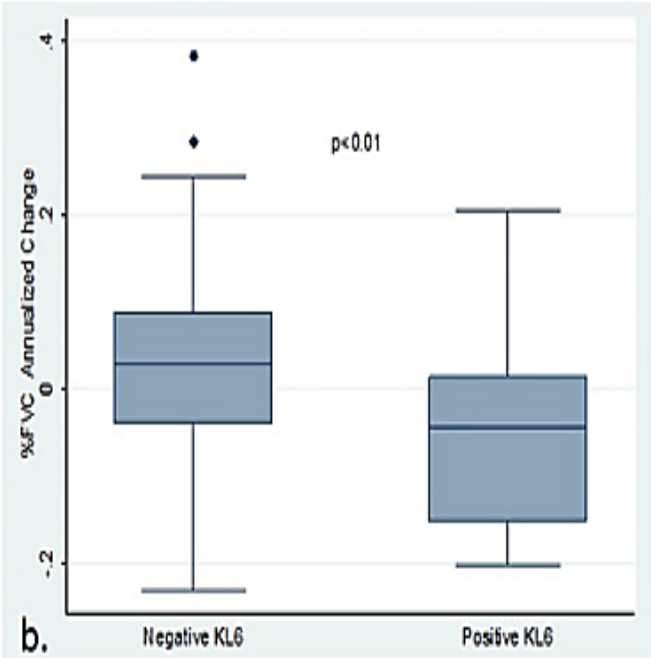
G. Salazar1,*, M. Kuwana2, M. Wu1, R. M. Estrada-Y-Martin3, J. Ying1, J. Charles1, C. Bellocchi4, M. Mayes4, S. Assassi4

N=82 pacientes/45 EScd

Validación: punto de corte KL6 1273U/mL

Figure 1. KL-6 levels between patients and controls and %FVC change





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POSTER TOUR 11:45-13:30
"INDEXES AND PREDICTORS IN SYSTEMIC SCLEROSIS AND MYOSITIS""

ESCLEROSIS SISTÉMICA

FRI0365
NEW COLLAGEN BIOMARKERS PREDICT PROGRESSION OF FIBROSIS IN SYSTEMIC SCLEROSIS
R. Dobrota1,*, S. Jordan1, P. Juhl2, B. Maurer1, L. Wildi1, A.-C. Bay-Jensen2, M. A. Karsdal2, A. S. Siebuhr2, O. Distler1

N=149 ES (23 Progresión)
Controles sanos 29

↓ 10% CFV

≥25% mRSS ≥25%

Degradación MEC
- C3M
- VICM
- C4M2
- BGM

Formación MEC
- P1NP
- P4NP7S
- Pro-C3
- Pro-C5
- Pro-C6

Ratio ProC3/c3M ¿Nuevo índice de progresión?

MADRID JUNIO 2017
EULAR

VASCULITIS-OTRASPOSTER TOUR 11:45-13:30
"RISK FACTORS FOR RMDS OR COMORBID CONDITIONS

VASCULITIS-OTRAS

FRI0304
THE UTILITY OF 18F FDG-PET/CT IN DISTINGUISHING BENIGN FROM MALIGNANT RETROPERITONEAL FIBROSIS
Y. Wang1,*, Z. Guan2, D. Gao1, J. Zhu1, J. Zhang1, F. Huang1

Table 1 Morphologic features, FDG-uptake of retroperitoneal lesions and lymph nodes (LNs)

Parameters	Benign RPF (71)	Malignant RPF (21)	p value
Morphologic features			
Craniocaudal length, median (IQR), mm	107.0(80.5,136.4)	164.9(104.7,229.2)	0.001
Axial width, median (IQR), mm	41.0(32.3,52.0)	85.0(62.3,98.5)	<0.001
Aorta lumen to anterior limit, median (IQR), mm	8.9(6.0,12.0)	20.7(13.9,45.3)	<0.001
Aorta lumen to posterior limit, median (IQR), mm	2.8(2.2,4.4)	7.4(4.5,14.5)	<0.001
FDG-uptake of retroperitoneal lesions			
High FDG-uptake, n (%)	55(77.5)	21(100.0)	0.017
SUV max, mean(S.D.)	4.8(1.7)	12.2(7.1)	<0.001
LNs with high FDG-uptake, n (%)			
Hilar/Mediastinal	27(38.0)	13(61.9)	0.054
Cervical	6(8.5)	5(23.8)	0.058
Axillary	4(5.6)	6(28.6)	0.003
Retroperitoneal	3(4.2)	16(76.2)	<0.001
Supraclavicular	2(2.8)	8(38.1)	<0.001
Inguinal	1(1.4)	5(23.8)	<0.001
Peritoneal	0(0.0)	10(47.6)	<0.001
Number of specific LNs, n (%)	0(0.0)	2(1.3)	<0.001

FRI0305
RELATIVE FDG ACCUMULATION OF THE AORTIC WALL LESIONS TO AORTIC BLOOD POOL IN 18F-FDG-PET AND PET/CT COULD BE A USEFUL PARAMETER FOR THE PREDICTION OF DISEASE RELAPSE AFTER SUCCESSFUL TREATMEN IN TAKAYASU ARTERITIS
A. Ihata1,*, Y. Kunishita2, K. Minegishi2, R. Yoshimi2, Y. Kirino2, H. Nakajima2

Characteristics of two groups		
Relapse	(-)	(+)
#	17	20
Age(yr)	47 [30-72]	28.5 [14-68]
CRP(mg/dl)	0.13±0.03	0.13±0.03
Steroid dose (mg/d equivalent to prednisolone)	15.6±2.4	17.7±3.6
Immunosuppressant	2/17	7/35
Duration until relapse (Days)		702.5 [4-1769]

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An Rheum Dis. 2017 **Comparison of performance of the 2016 ACR-EULAR classification criteria for primary Sjögren's syndrome with other sets of criteria in Japanese patients.**
Tsuboi H[1,2], Hagiwara S[1], Asashima H[1], Takahashi H[1], Hirota T[1], Noma H[3], Umehara H[2,4,5], Kawakami A[2,6], Nakamura H[2,6], Sano H[2,7], Tsubota K[2,8], Ogawa Y[2,8], Takamura E[2,9], Saito I[2,10], Inoue H[2,11], Nakamura S[2,12], Moriyama M[2,12], Takeuchi T[2,13], Tanaka Y[2,14], Hirata S[2,14], Mimori T[2,5], Matsumoto I[1], Sumida T[1,2].

Ann Rheum Dis. 2017 Jul;76(7):1161-1168. doi: 10.1136/annrheumdis-2016-210448. Epub 2016 Dec 13.
Standardisation of labial salivary gland histopathology in clinical trials in primary Sjögren's syndrome.
Fisher BA1,2, Jonsson R3,4, Daniels T5, Bombardieri M6, Brown RM7, Morgan P8, Bombardieri S9, Ng WF10, Tzioufas AG11, Vitali C12, Shirlaw P13, Haacke E14, Costa S15, Bootsma H16, Devauchelle-Pensec V17, Radstake TR18, Mariette X19, Richards A20, Stack R1, Bowman SJ1,2, Barone F1; Sjögren's histopathology workshop group (appendix) from ESSENTIAL (EULAR Sjögren's syndrome study group).

EULAR Sjögren's syndrome disease activity index (ESSDAI): a user guide.
Seror R, Bowman SJ, Brito-Zeron P, Theander E, Bootsma H, Tzioufas A, Gottenberg JE, Ramos-Casals M, Dörner T, Ravaud P, Vitali C, Mariette X, Asmussen K, Jacobsen S, Bartoloni E, Gerli R, Bijlsma JW, Kruize AA, Bombardieri S, Bookman A, Kallenberg C, Meiners P, Brun JG, Jonsson R, Caporali R, Carsons S, De Vita S, Del Papa N, Devauchelle V, Saraux A, Fauchais AL, Sibilia J, Hachulla E, Illei G, Isenberg D, Jones A, Manoussakis M, Mandl T, Jacobsson L, Demoulins F, Montecucco C, Ng WF, Nishiyama S, Omdal R, Parke A, Praprotnik S, Tomsic M, Price E, Scofield H, L Sivils K, Smolen J, Laqué RS, Steinfeld S, Sutcliffe N, Sumida T, Valesini G, Valim V, Vivino FB, Vollenweider C.

Ann Rheum Dis. 2016 Feb;75(2):382-9. doi: 10.1136/annrheumdis-2014-206008. Epub 2014 Dec 5.
Defining disease activity states and clinically meaningful improvement in primary Sjögren's syndrome with EULAR primary Sjögren's syndrome disease activity (ESSDAI) and patient-reported indexes (ESSPRI).
Seror R1, Bootsma H2, Saraux A3, Bowman SJ4, Theander E5, Brun JG6, Baron G7, Le Guern V8, Devauchelle-Pensec V3, Ramos-Casals M9, Valim V10, Dörner T11, Tzioufas A12, Gottenberg JE13, Solans Laqué R14, Mandl T5, Hachulla E15, Sivils KL16, Ng WF17, Fauchais AL18, Bombardieri S19, Priori R20, Bartoloni E21, Goeb V22, Praprotnik S23, Sumida T24, Nishiyama S25, Caporali R26, Kruize AA27, Vollenweider C28, Ravaud P7, Meiners P29, Brito-Zerón P9, Vitali C30, Mariette X31; EULAR Sjögren's Task Force.

EBioMedicine. 2016 Aug;10:216-26. doi: 10.1016/j.ebiom.2016.06.041. Epub 2016 Jun 29.
Targeting the Ca(2+) Sensor STIM1 by Exosomal Transfer of Ebv-miR-BART13-3p is Associated with Sjögren's Syndrome.
Gallo A1, Jang SI1, Ong HL2, Perez P1, Tandon M1, Ambudkar I2, Illei G1, Alevizos I3.

Ann Rheum Dis. 2016 May;75(5):933-40. doi: 10.1136/annrheumdis-2014-206998. Epub 2015 Jul 16.
Overlap between differentially methylated DNA regions in blood B lymphocytes and genetic at-risk loci in primary Sjögren's syndrome.
Miceli-Richard C1, Wang-Renault SF2, Boudaoud S1, Busato F3, Lallemand C3, Bethune K3, Belkhir R1, Nocturne G1, Mariette X1, Tost J3.

Ann Rheum Dis. 2016 Apr;75(4):780-3. doi: 10.1136/annrheumdis-2015-207731. Epub 2015 Sep 2.
Germline variation of TNFAIP3 in primary Sjögren's syndrome-associated lymphoma.
Nocturne G1, Tarn J2, Boudaoud S1, Locke J2, Miceli-Richard C3, Hachulla E4, Dubost JJ5, Bowman S6, Gottenberg JE7, Criswell LA8, Lessard CJ9, Sivils KL9, Carapito R10, Bahram S11, Seror R1, Ng WF2, Mariette X12.

Characterization of systemic disease in primary Sjögren's syndrome: EULAR-SS Task Force recommendations for articular, cutaneous, pulmonary and renal involvements.
Ramos-Casals M, Brito-Zerón P, Seror R, Bootsma H, Bowman SJ, Dörner T, Gottenberg JE, Mariette X, Theander E, Bombardieri S, De Vita S, Mandl T, Ng WF, Kruize A, Tzioufas A, Vitali C; EULAR Sjögren Syndrome Task Force.

Joint Bone Spine. 2017 May;84(3):317-321. doi: 10.1016/j.jbspin.2016.05.019. Epub 2016 Sep 19.
Cystic lung disease in Sjögren's syndrome: An observational study.
Lechtman S1, Debray MP2, Crestani B3, Bancal C4, Hourseau M5, Dossier A1, Alexandra JF1, Chauveheid MP1, Papo T6, Sacre K7.

Predicting the risk for lymphoma development in Sjogren syndrome: An easy tool for clinical use.
Fragkioudaki S, Mavragani CP, Moutsopoulos HM.

Br J Haematol. 2015 Feb;168(3):317-27. doi: 10.1111/bjh.13192. Epub 2014 Oct 15.
Sjögren Syndrome-associated lymphomas: an update on pathogenesis and management.
Nocturne G1, Mariette X.