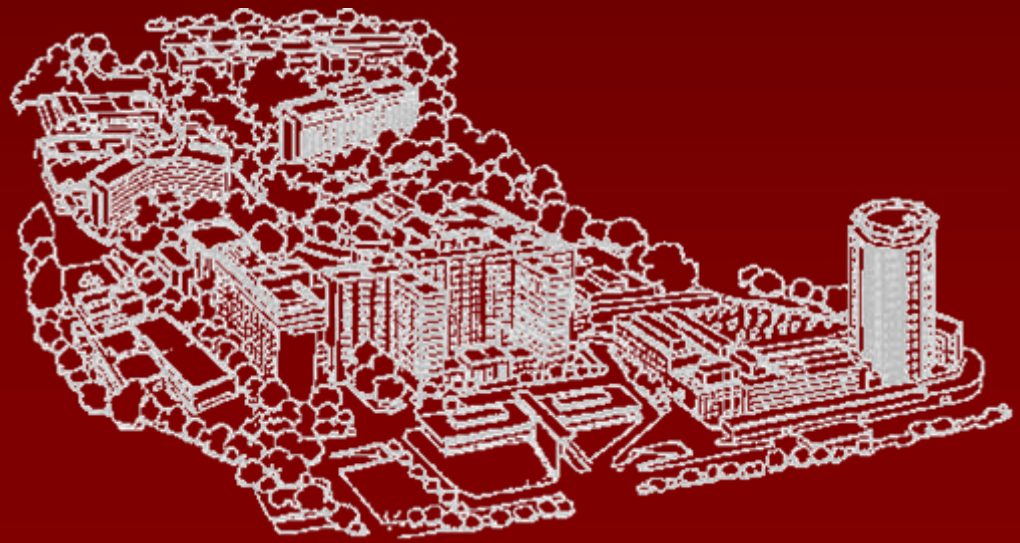
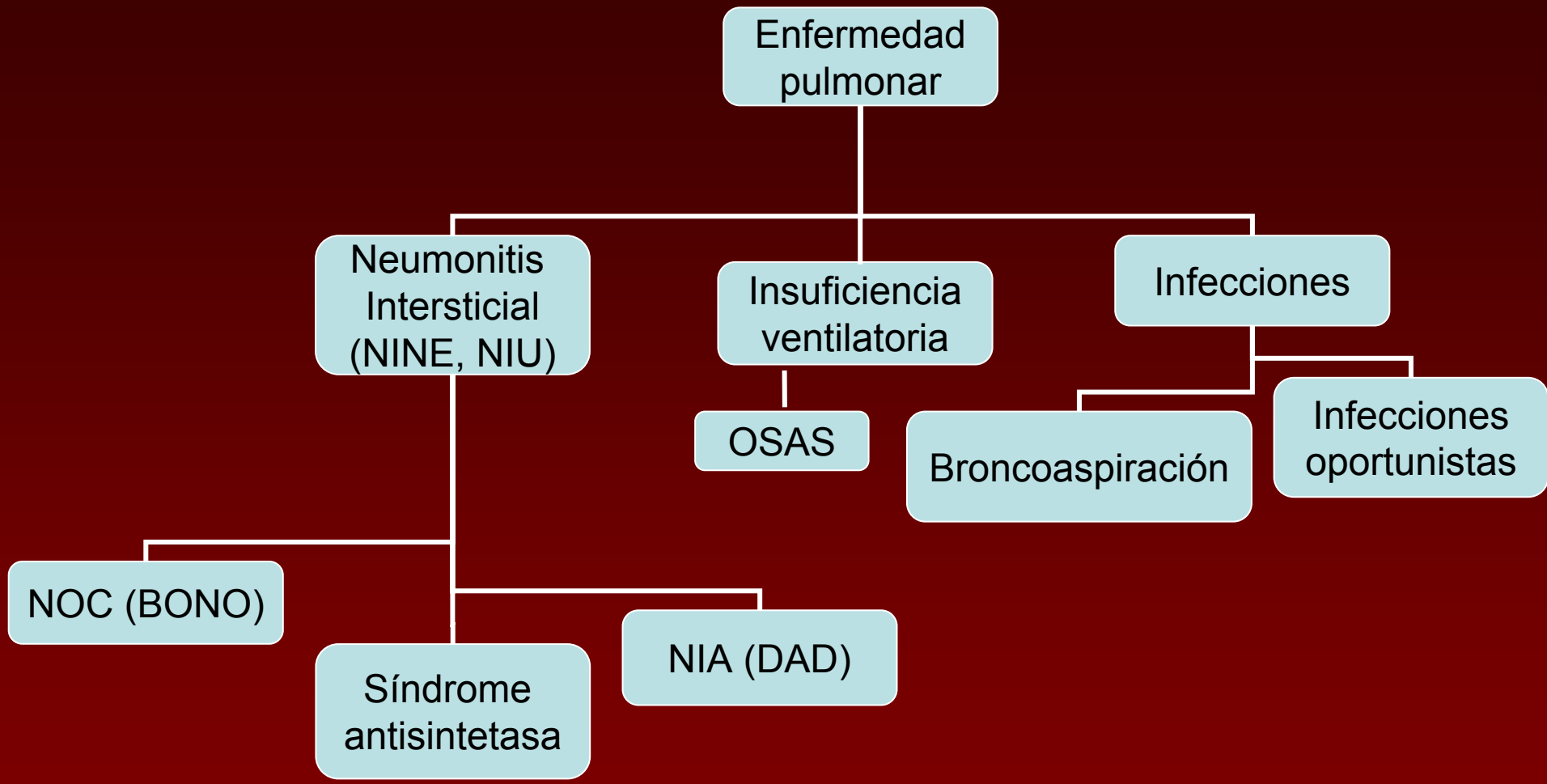
II Reunión en Enfermedades Autoinmunes Sistémicas

Afección pulmonar en las miopatías inflamatorias



Dr. Albert Selva O'Callaghan

Bilbao 2009, 25-26 junio



MIOSITIS Y PULMÓN

Fathi M, Lundberg I. Interstitial lung disease in polymyositis and dermatomyositis. Curr Opin Rheumatol. 2005; 12:701-706

Exploraciones Complementarias

PFR (CVF, FEV, DLCO, PIM, PEM)

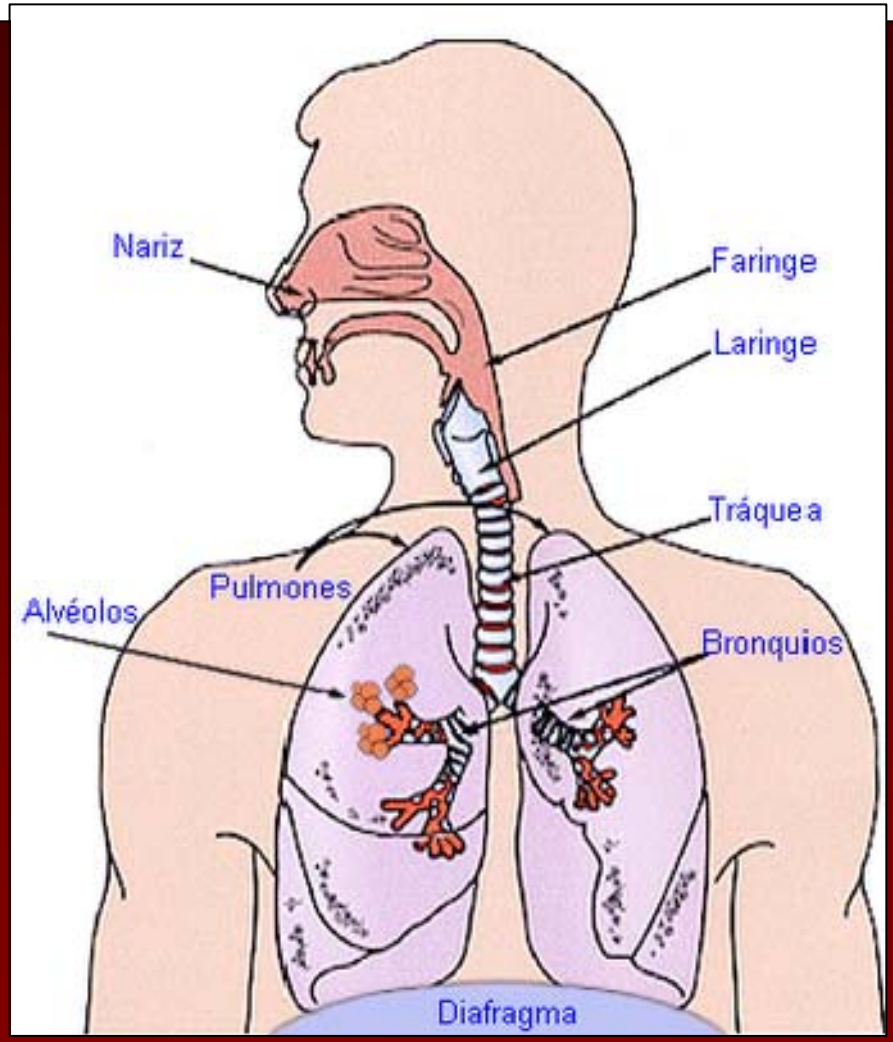
FBC (BAL, BAS, BTB)

TACAR

Ecocardiografía

PSG

Estudio inmunológico (antisintetasa, HLA)



Respirology (2008) **13**, 863–870 doi: 10.1111/j.1440-1843.2008.01363.x

ORIGINAL ARTICLE

Transbronchial biopsy is clinically useful in classifying patients with interstitial pneumonia associated with polymyositis and dermatomyositis

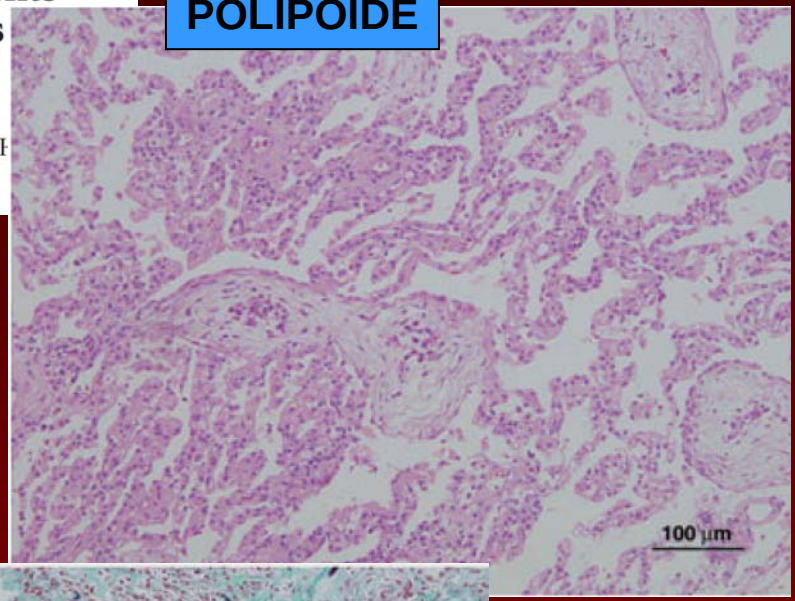
Hiroshi MOCHIMARU,[1,2,3] Masashi KAWAMOTO,[2] Tatsuji ENOMOTO,[1] Yoshinobu SAITOH, Shinji ABE,[1] Takahito NEI,[1] Yuh FUKUDA[2] AND Shoji KUDOH[1]

19 de 25 (78%) PM/DM, ILD [BTB]

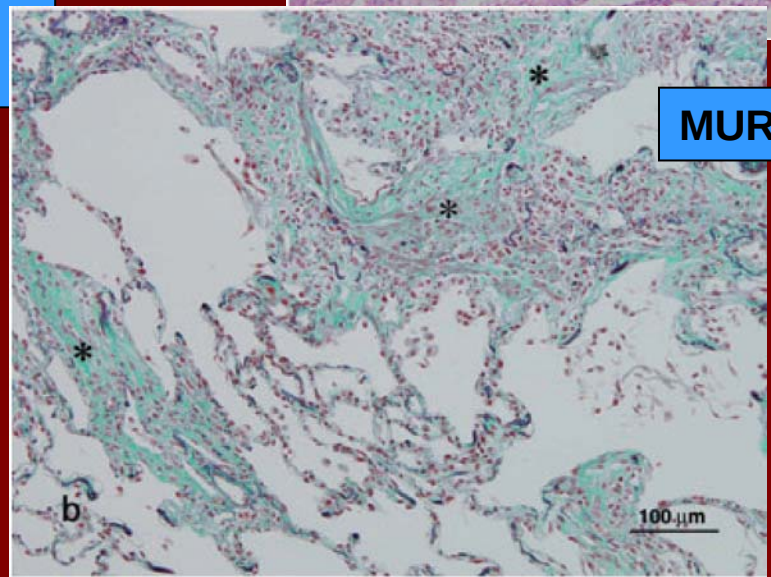
5 fibrosis polipoide (responden a GC)

14 fibrosis mural (peor pronóstico)

POLIPOIDE



MURAL

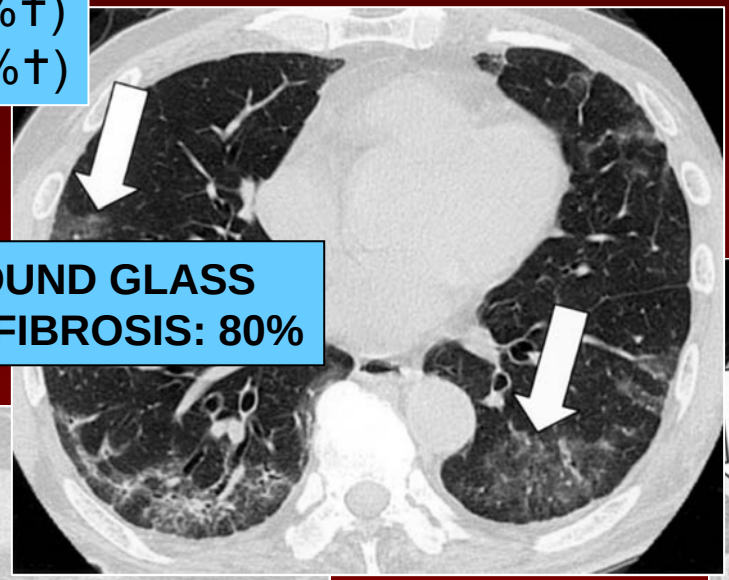


High-Resolution Computed Tomography Characterization of Interstitial Lung Diseases in Polymyositis/Dermatomyositis

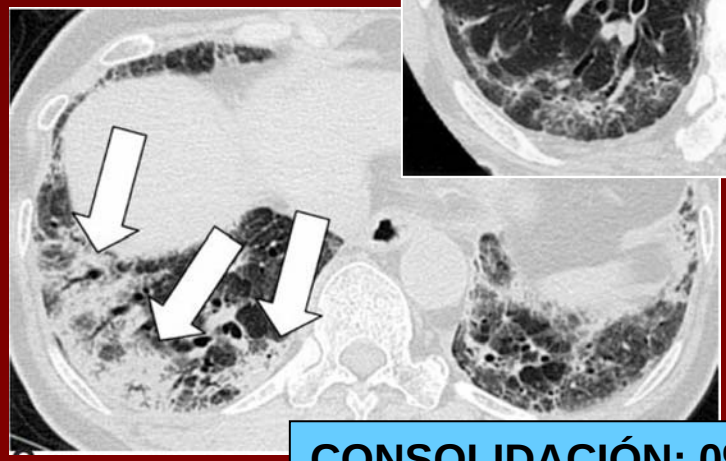
SHINICHIRO HAYASHI, MASAHIDE TANAKA, HIROMI KOBAYASHI, TAKAHIKO NAKAZONO, TOSHIMI SATOH, YUJI FUKUNO, NAOKO ARAGANE, YOSHIFUMI TADA, SHUICHI KOARADA, AKIHIDE OHTA, and KOHEI NAGASAWA

J Rheumatol 2008;35:260–9

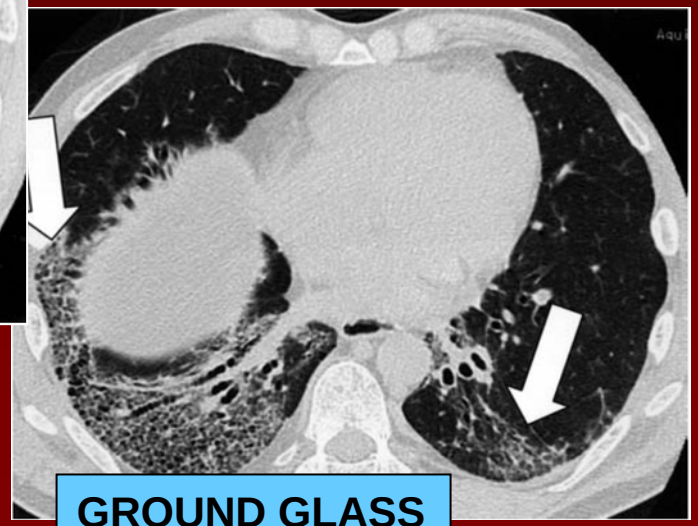
17 PM (12%†)
16 DM (44%†)



GROUND GLASS SIN FIBROSIS: 80%



CONSOLIDACIÓN: 0%



GROUND GLASS Y FIBROSIS: 20%

Manifestaciones clínicas

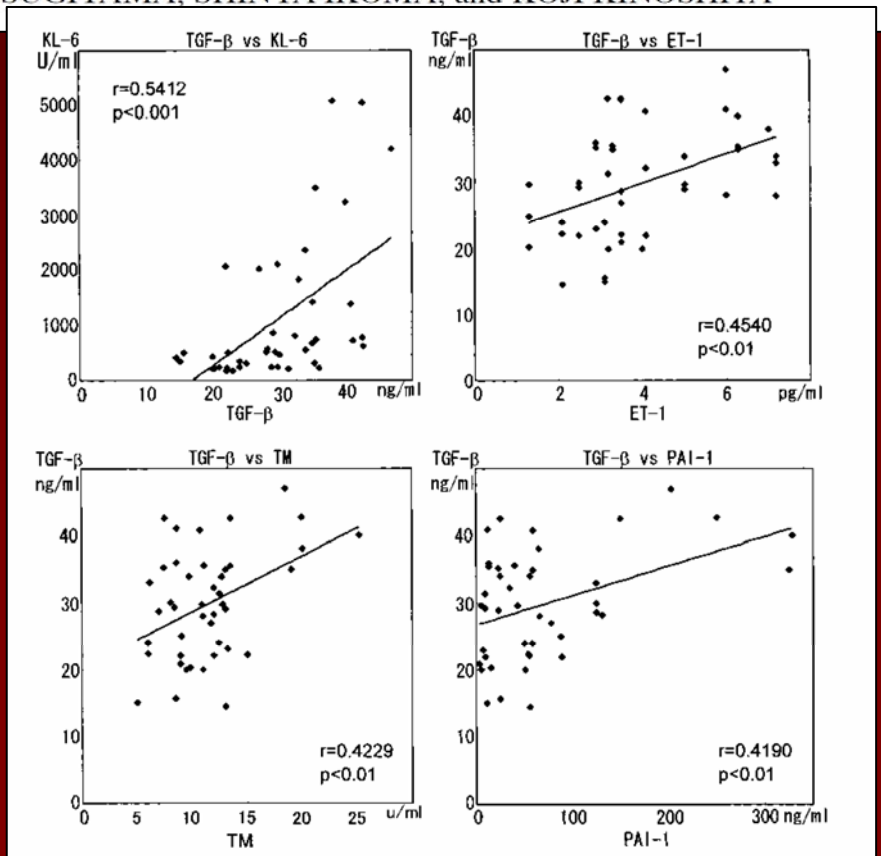
Autor	n/ afectación pulmonar	Neumonitis intersticial	Aguda	ASA	Miopatía restrictiva	Fuente
Kang EH	72	29 (40%)	6 (20%)	6/22 (27%)	-	Rheumatol 2005
Selva-O'Callaghan A	50/81 (61%)	32 (39%)	5 (10%)	17/50 (34%)	22 (18%)	Lupus 2005
Marie I	36/156 (23%)	36 (23%)	6 (16%)	11/36 (30%)	-	A&R 2002
Douglas WW	70	70	-	19/50 (38%)	-	AJRCCM 2001
Schnabel A	20/63 (32%)	20 (32%)	10 (50%)	15/20 (75%)	-	Semin Arthritis Rheum 2003

Etiopatogenia

Role of Endothelial Damage in the Pathogenesis of Interstitial Pneumonitis in Patients with Polymyositis and Dermatomyositis

MASANORI FUNAUCHI, HIDEKI SHIMADSU, CHISE TAMAKI, TOSHIAKI YAMAGATA, YUJI NOZAKI, MASAFUMI SUGIYAMA, SHINYA IKOMA, and KOJI KINOSHITA

(J Rheumatol 2006;33:903–6)

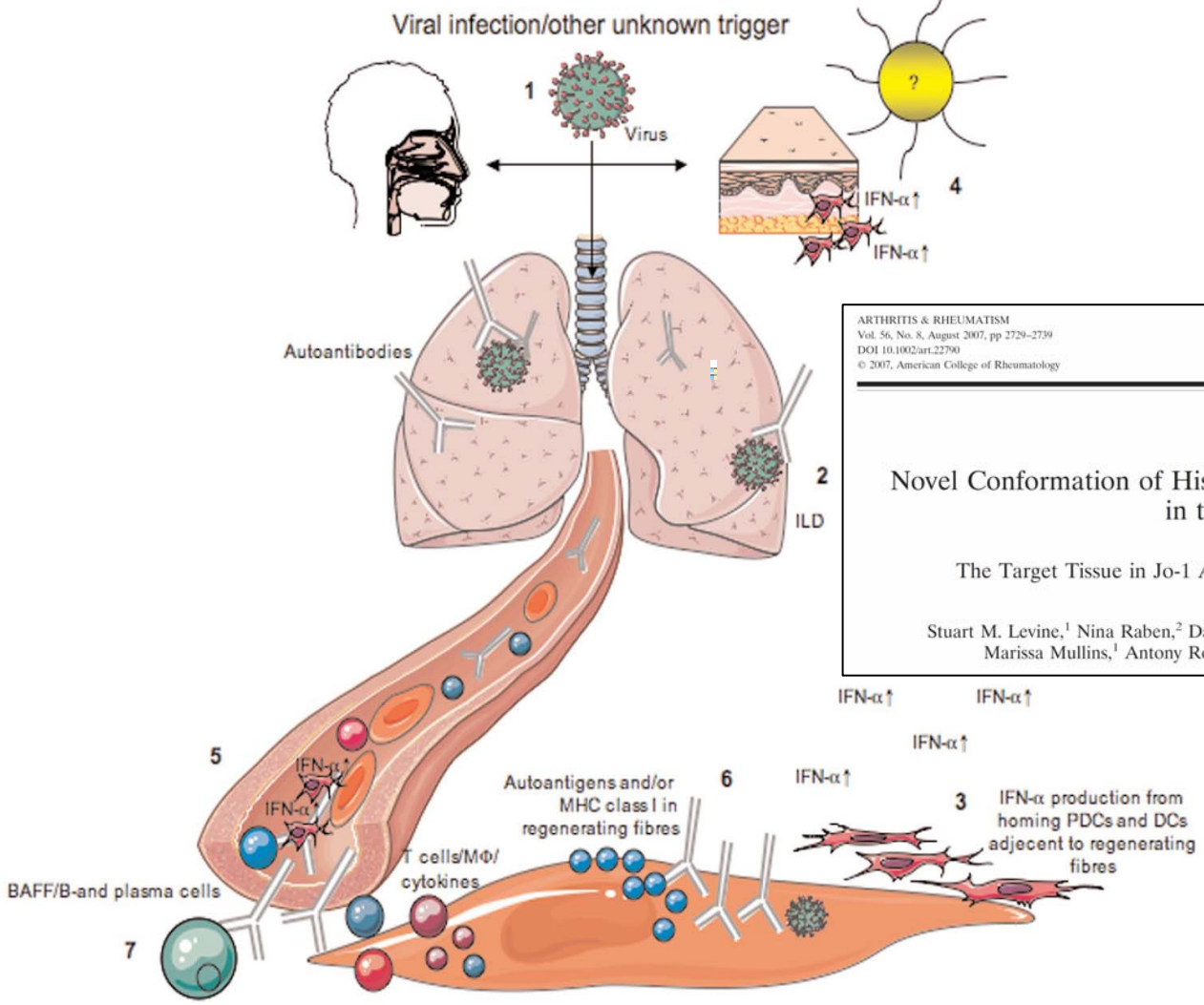


Review

Developments in the scientific and clinical understanding of inflammatory myopathies

Ingrid E Lundberg and Cecilia Grundtman

Arthritis Research & Therapy 2008, **10:**220 (doi:10.1186/ar2501)



ARTHRITIS & RHEUMATISM
Vol. 56, No. 8, August 2007, pp 2729–2739
DOI 10.1002/art.22790

Novel Conformation of Histidyl–Transfer RNA Synthetase in the Lung

The Target Tissue in Jo-1 Autoantibody–Associated Myositis

Stuart M. Levine,[1] Nina Raben,[2] Dan Xie,[3] Frederic B. Askin,[1] Rubin Tuder,[1] Marissa Mullins,[1] Antony Rosen,[1] and Livia A. Casciola-Rosen[1]

Evolución de la Neumopatía Intersticial

Arthritis & Rheumatism (Arthritis Care & Research)
Vol. 59, No. 5, May 15, 2008, pp 677–685
DOI 10.1002/art.23571

ORIGINAL ARTICLE

Interstitial Lung Disease in Polymyositis and Dermatomyositis: Longitudinal Evaluation by Pulmonary Function and Radiology

MARYAM FATHI,[1] JENNY VIKGREN,[2] MARIANNE BOIJSEN,[2] ULF TYLEN,[2] LENNART JORFELDT,[3] GÖRAN TORNLING,[1] AND INGRID E. LUNDBERG[1]

Estudio prospectivo con un seguimiento de 35 meses
18 de 23 PM/DM (78%) ILD (sólo 6 anti-Jo-1+)

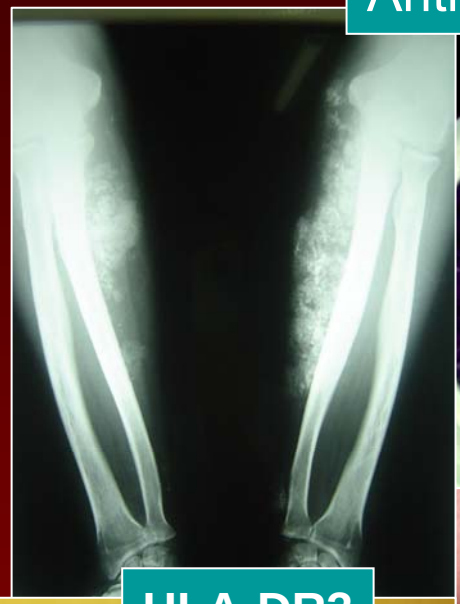
- 33% mejoraron
- 29% estabilización
- 28% deterioro
- 2 (10%) fallecieron

PFR se normalizaron a pesar de permanecer
la lesión estructural

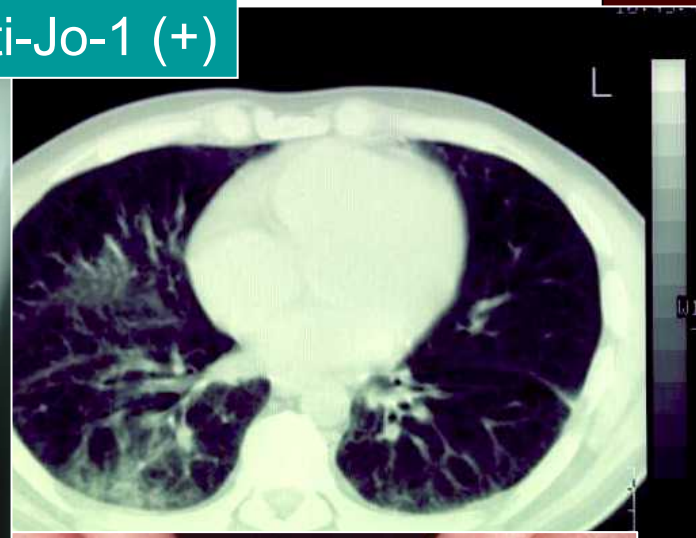
Síndrome antisintetasa (NOC, NIU, NINE)

Miositis (PM>DM)
Neumonitis intersticial
Artritis

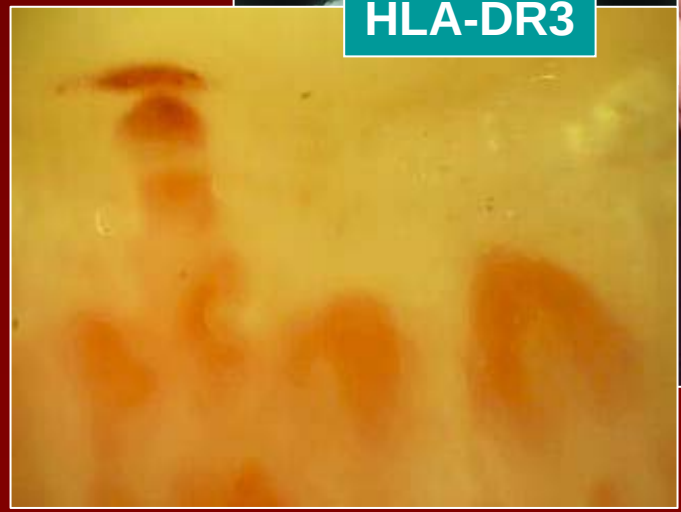
Manos de mecánico
Fiebre
F Raynaud
Calcinosis
Alteraciones capilaroscopicas

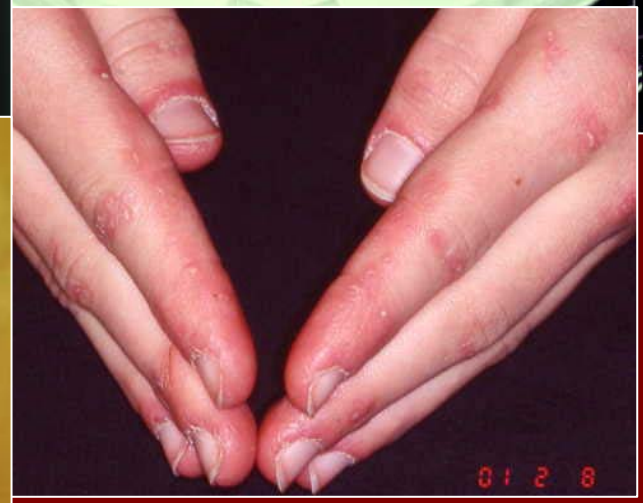


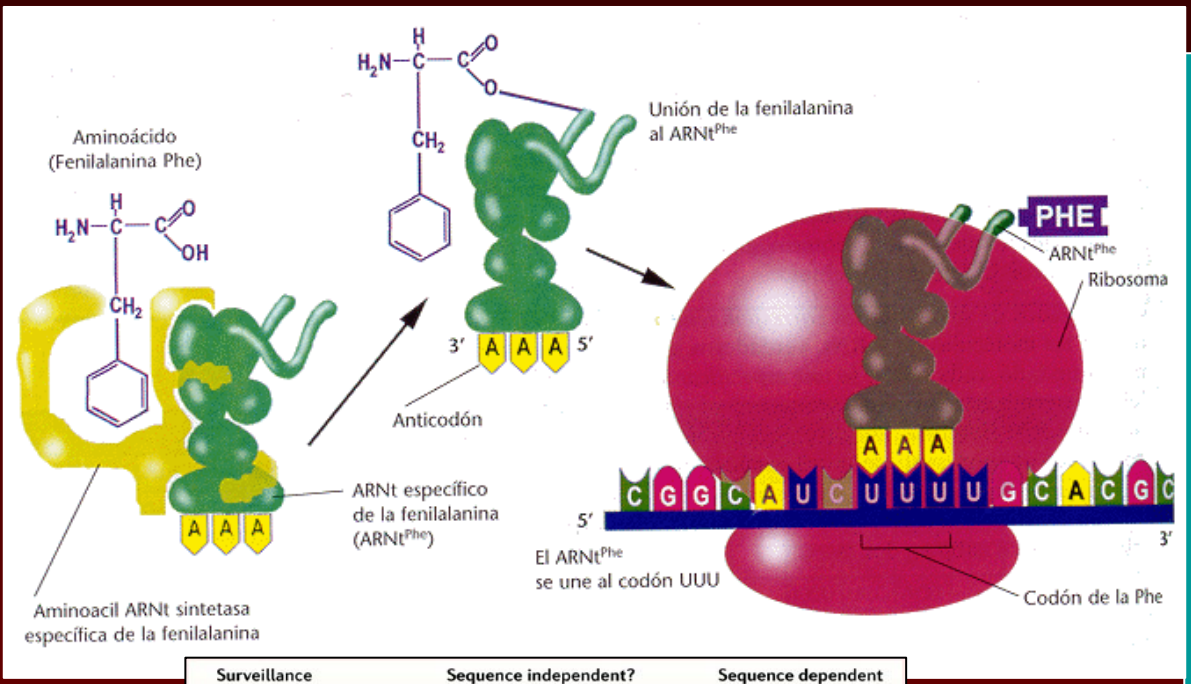
Anti-Jo-1 (+)

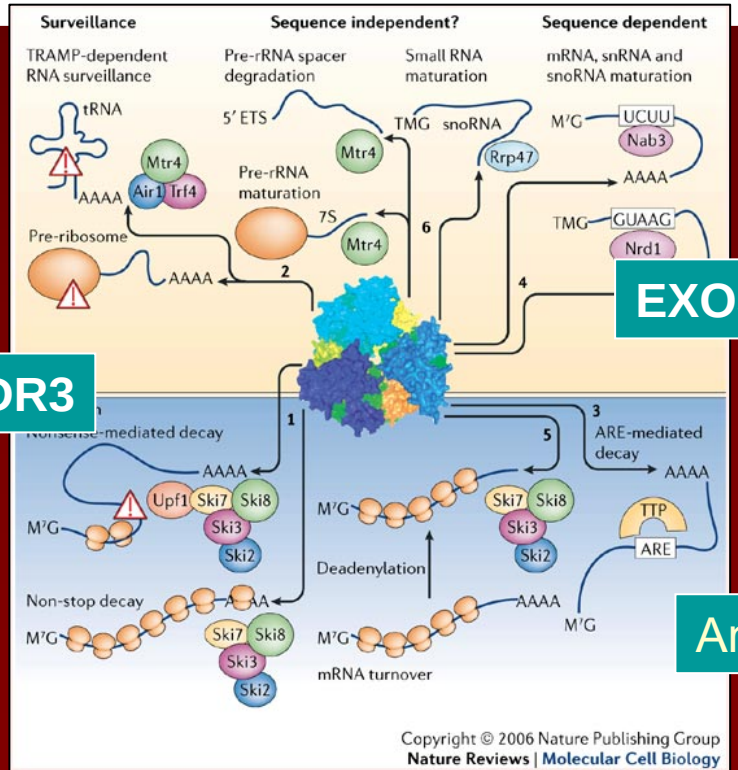


HLA-DR3









EXOSOMA

HLA-DR3

Anti-PM/Scl

- Anti-histidil-tRNA-sintetasa (anti-Jo-1) [20-30%]
- Anti-treonil-tRNA-sintetasa (anti-PL-7) [3-4%]
- Anti-alanil-tRNA-sintetasa (anti-PL-12) [3-4%]
- Anti-isoleucil-tRNA-sintetasa (anti-OJ) [<2%]
- Anti-glicil-tRNA-sintetasa (anti-EJ) [<2%]
- Anti-asparaginil-tRNA-sintetasa (anti-KS) [<2%]
- Anti-tirosil-tRNA-sintetasa (anti-YRS) [<2%]
- Anti-fenilalanil-tRNA-sintetasa (anti-Zo) [<2%]

Table 1
Overview of clinical associations of anti-PM/Scl antibodies

Study	Reichlin	Reimer	Genth	Oddis	Marguerie	Hausmanowa	Vandergheynst	Selva-O'callaghan
Reference	[6]	[16]	[36]	[17]	[27]	[30]	[22]	[21]
Year	1984	1988	1990	1992	1992	1997	2006	2006
Patient selection	PM	SSc	SSc/ANA	IIM/SSc/RP	SSc/IIM	IIM	ANoA	IIM
Anti-PM/Scl detection	ID	IP	IIF / ID	IIF/ID	ID	ID	IIF/ID	IP
Number of patients	20	8	12	23	32	20	14	10
Percentage females	–	88	92	–	–	–	93	40
Diagnosis (%)								
PM	23	–	–	4	3	–	7	10
DM	32	–	–	22	–	–	29	80
PM/SSc [a]	41	25	(42)	43	84	95	36	(40)
Scleroderma	5	75	83	30	13	–	21	–
Other	–	–	17	–	–	5	7	10
Symptoms [b] *(%)*								
Raynaud's phenomenon	50	88	92	65	100	70	71	50
Arthritis	50	50	58	83	97	95	86	80
Muscle involvement	95	43	50	78	88	100	79	–
Skin involvement	20	100	50	–	47	45	–	30
Lung involvement	35	67	42	30	78	60	86	60
Esopheagal involvement	–	43	36	–	78	–	21	50
Kidney involvement	–	25	0	–	–	5	21	–
Dry mouth/eyes	–	–	25	–	34	35	43	11

Caso/Sexo	Edad (años)	Diagnóstico	CPK (UI/l)	Neumopatía Intersticial	Manos mecánico	Artritis	Capilaroscopia Fenómeno Raynaud	ANA (IFI)	HLA
1 / ♀	60	DM (p)	558	CVF 54% Fibrosis	Si	Si	Alterada No	320 Nucleolar	DR11
2 / ♀	33	DM	2.560	CVF 52% NOC	Si	No	Alterada Si	640 Nucleolar	DR3
3 / ♂	76	DM	992	CVF 44% Fibrosis	Si	Si	Alterada Si	640 Nucleolar	DR3
4 / ♀	58	Sine miositis	67	CVF 57% Fibrosis	Si	Si	Alterada No	640 Nucleolar	DR3
5 / ♀	22	DM	2.096	CVF 50% Ground glass	Si	Si	Alterada Si	160 Moteado	DR3
6 / ♀	45	DM	669	CVF 38% Ground glass	No	Si	Alterada Si	640 Nucleolar	DR3
7 / ♂	15	DM	67	CVF 77% Ground glass	Si	Si	Alterada No	1.280 Nucleolar	DR3
8 / ♂	41	No biopsia	180	CVF 46% Fibrosis	Si	Si	No realizada No	1.280 Nucleolar	_
9 / ♂	51	PM	1.730	CVF 76% Fibrosis	Si	Si	Esclerodermia Si	1.280 Nucleolar	DR3
10 / ♀	54	DM	1.700	CVF 60% Ground glass	Si	Si	Esclerodermia Si	640 Nucleolar	DR3

Neumonitis intersticial (NOC)

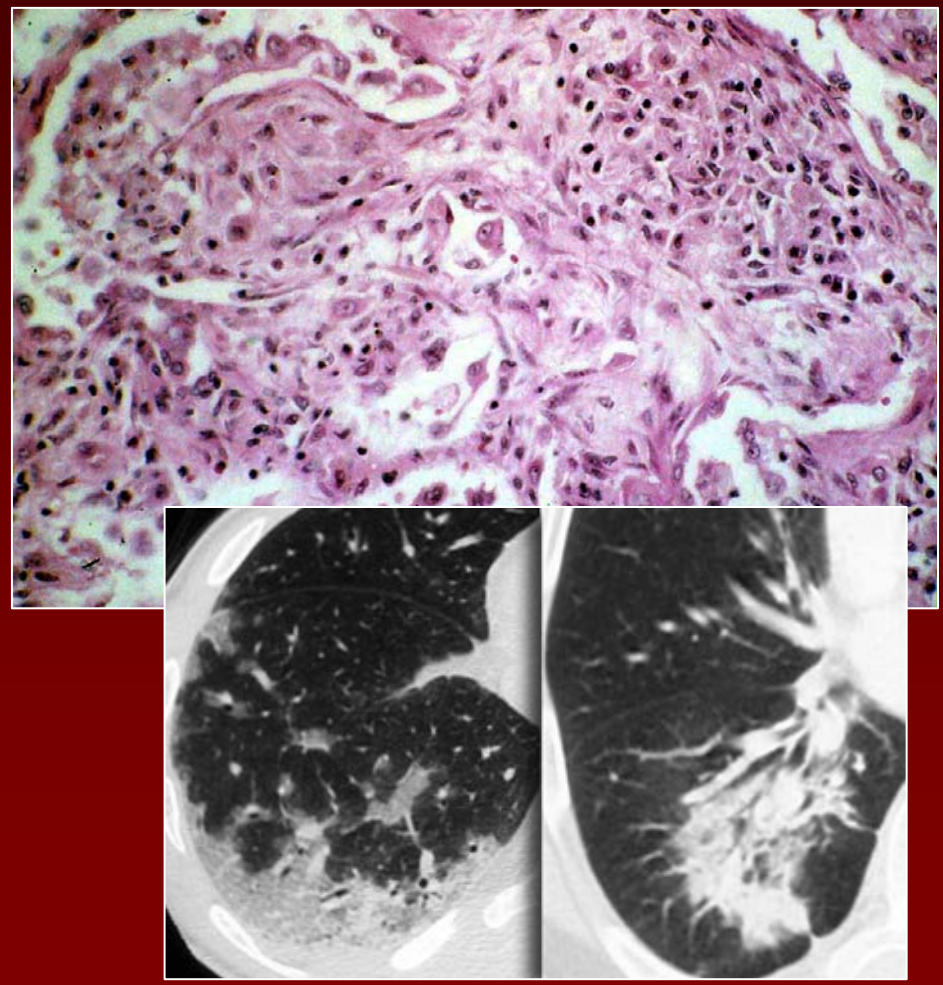
Importancia de la FBC (BAL, BAS, BTB)

PFR

Respuesta a tratamiento GC

1 mg/kg/d

Marcadores inmunológicos (antiaminoacil- tRNA sintetasa)



Neumonitis intersticial aguda (DAD)

Dermatomiositis (amiopática?)

Anti-sintetasa (-)

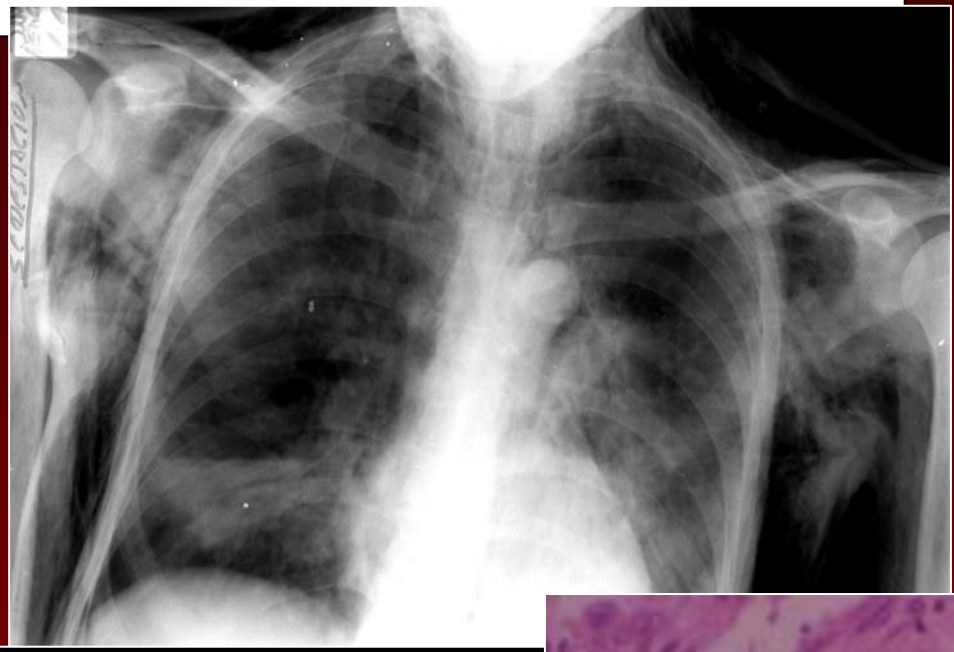
Curso fulminante (Hamman-Rich)

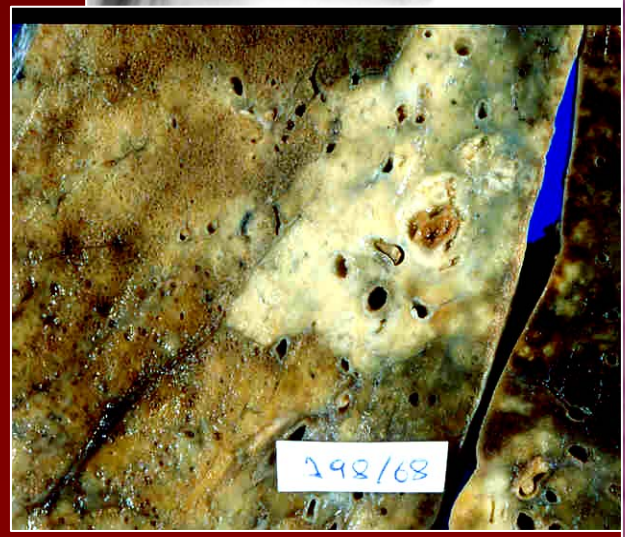
Neumomediastino

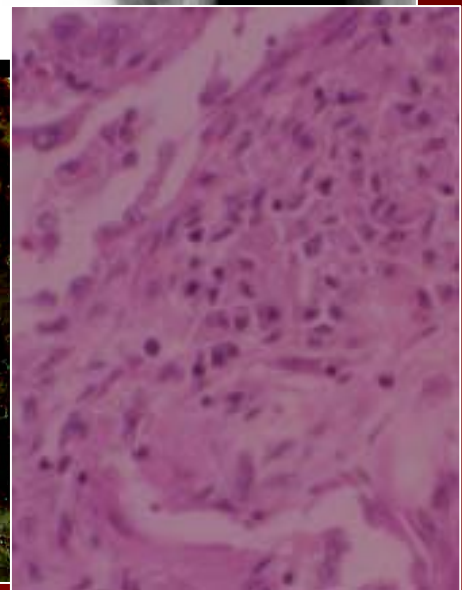
Mala respuesta al tratamiento

Trasplante pulmonar?

Daño alveolar difuso



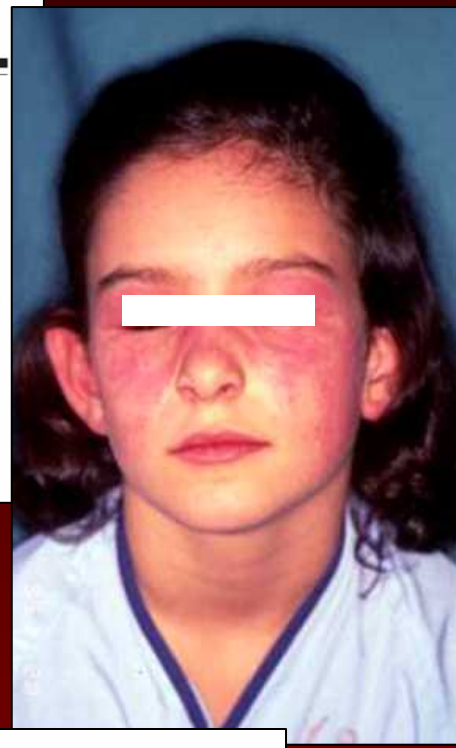


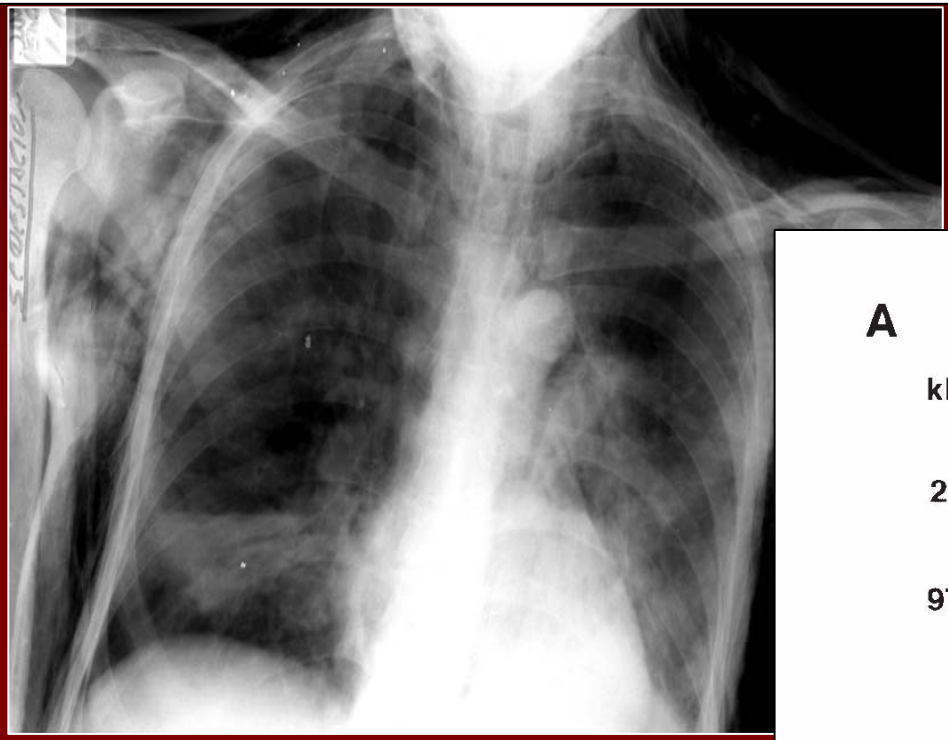


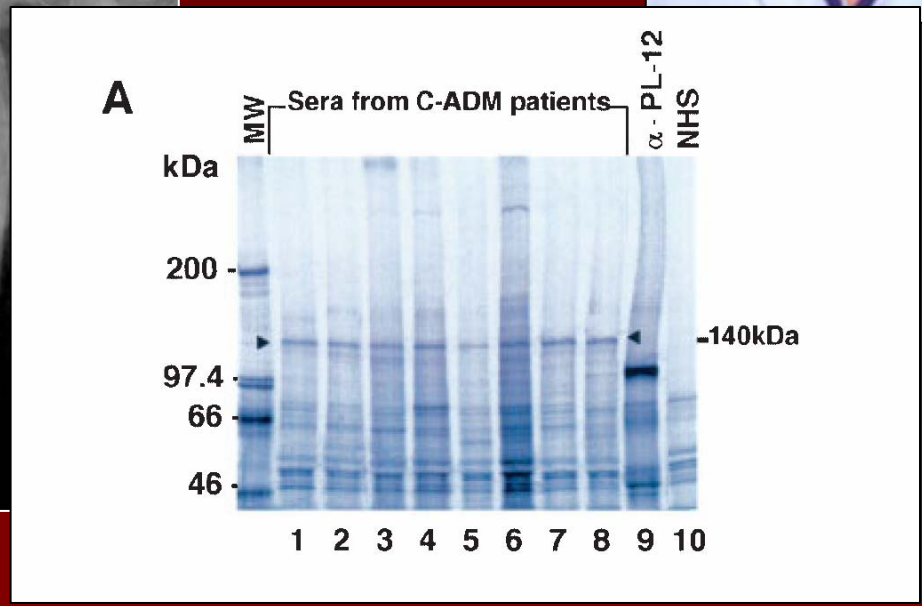
ARTHRITIS & RHEUMATISM
Vol. 52, No. 5, May 2005, pp 1571–1576
DOI 10.1002/art.21023

Autoantibodies to a 140-kd Polypeptide, CADM-140, in Japanese Patients With Clinically Amyopathic Dermatomyositis

Shinji Sato,[1] Michito Hirakata,[1] Masataka Kuwana,[1] Akira Suwa,[1] Shinichi Inada,[2] Tsuneyo Mimori,[3] Takeji Nishikawa,[1] Chester V. Oddis,[4] and Yasuo Ikeda[1]







Tjiu JW, et al. Digital infarcts showing microangiopathy in adult dermatomyositis suggest severe pulmonary involvement and poor prognosis. Br J Dermatol 2004; 150:1214—1216.

Nailfold Capillary Microscopy in Adults with Inflammatory Myopathy

Albert Selva-O'Callaghan, MD, PhD,* Vicente Fonollosa-Pla, MD, PhD,*
Ernesto Trallero-Araguás, MD,* Xavier Martínez-Gómez, MD,†
Carmen Pilar Simeon-Aznar, MD, PhD,*
Moisés Labrador-Horrillo, MD, PhD,* and
Miquel Vilardell-Tarrés, MD, PhD*

Semin Arthritis Rheum. 2008. *[Epub ahead of print]*

Table 2 Groups and Clinical Features of Myositis Patients Stratified by Capillary Score

Parameters	NC Score < 2 (n = 38) (<4 alterations)	NC Score = 2 (n = 15) (≥4 alterations)	P
Median age (y)	57.4 (44.8 to 69.5)	55.8 (37.3 to 62.6)	NS
Median age at onset (y)	49 (33.3 to 59.5)	47.5 (30.5 to 61.5)	NS
Median disease duration (y)	5.9 (3.2 to 11.6)	7.0 (2.1 to 13.7)	NS
MDAAT	0.3 (0.0 to 0.5)	0.5 (0.3 to 0.7)	<0.05
MDI severity	0.2 (0.1 to 0.3)	0.3 (0.2 to 0.4)	<0.05
Raynaud	8 (21)	8 (53)	<0.05
ILD	9 (23)	8 (53)	<0.05
Neoplasm	3 (8)	3 (20)	NS
MSA/MAA	16 (45)	8 (53)	NS

Data are expressed as the median (interquartile range) and count (percentage).
DM, dermatomyositis; ILD, interstitial lung disease; MDAAT, myositis disease activity assessment tool; MDI, myositis damage index; MSA/MAA, myositis specific/associated antibodies; NC, nailfold capillaroscopy; NS, nonsignificant; PM, polymyositis.

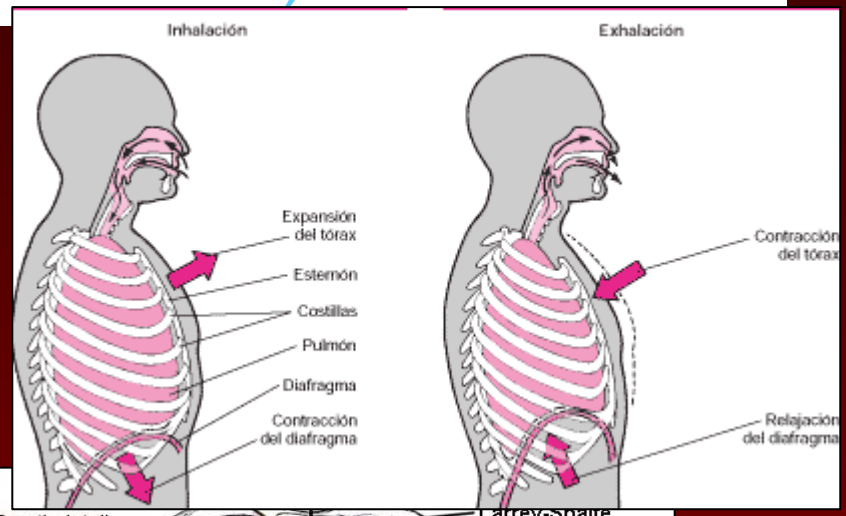
Insuficiencia ventilatoria (miopatía restrictiva)

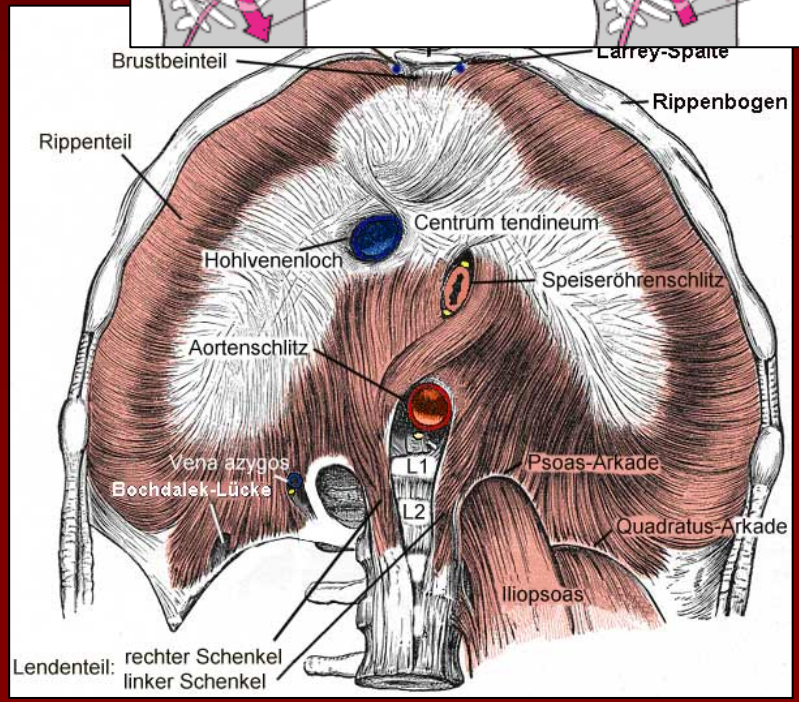
Grave que precisa ventilación con BIPAP (< 5%)

Miopatía restrictiva (20-30%)
curso paralelo a la enfermedad

Importancia de la determinación PIM y PEM

Responde a tratamiento inmunodepresor





Rheumatology 2000;39:914–916

Respiratory failure due to muscle weakness in inflammatory myopathies: maintenance therapy with home mechanical ventilation

A. Selva-O'Callaghan, L. Sanchez-Sitjes, X. Muñoz-Gall[1], T. Mijares-Boeckh-Behrens, R. Solans-Laque, J. Angel Bosch-Gil, F. Morell-Brotad[1] and Miguel Vilardell-Tarrés

Department of Internal Medicine and [1]Pneumology Service, Vall D'Hebron General Hospital, Barcelona,

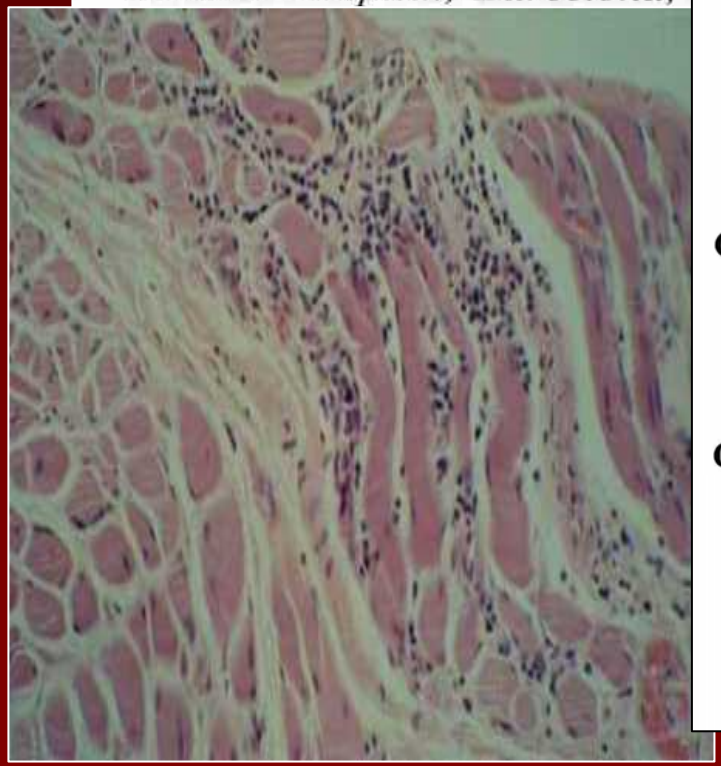
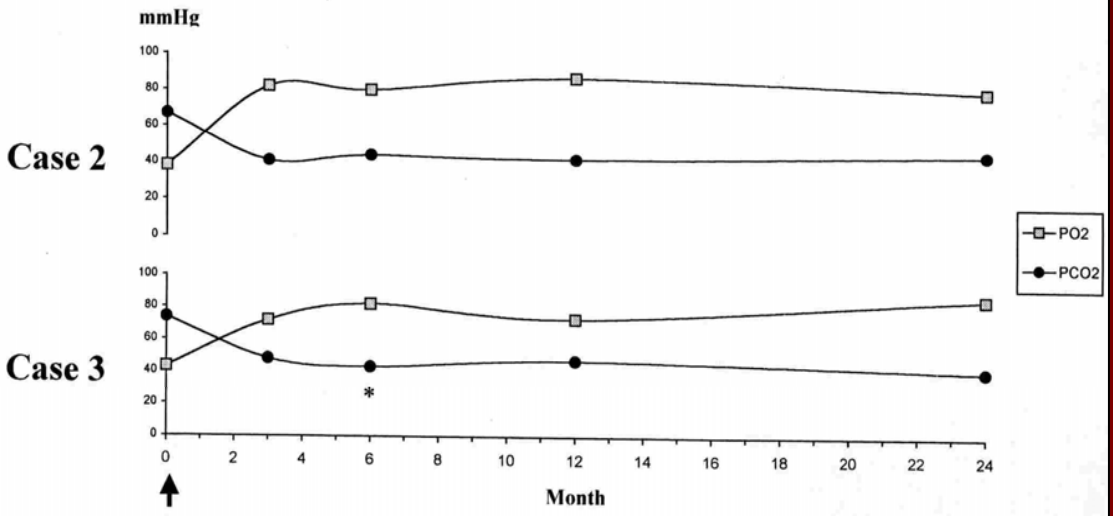


Figure 2: Evolution of arterial blood gases after mechanical ventilation was started in case 2 and case 3
(Arterial blood gases were tested while the patient was breathing air room without ventilation)



Muscle Nerve **39:** 144–149, 2009

OBSTRUCTIVE SLEEP APNEA IN PATIENTS WITH INFLAMMATORY MYOPATHIES

ALBERT SELVA-O'CALLAGHAN, MD, PhD,[1] GABRIEL SAMPOL, MD, PhD,[2,4] ODILE ROMERO, MD,[3,4]
PATRICIA LLOBERES, MD,[2,4] ERNESTO TRALLERO-ARAGUÁS, MD,[1] and MIQUEL VILARDELL-TARRÉS, MD, PhD[1]

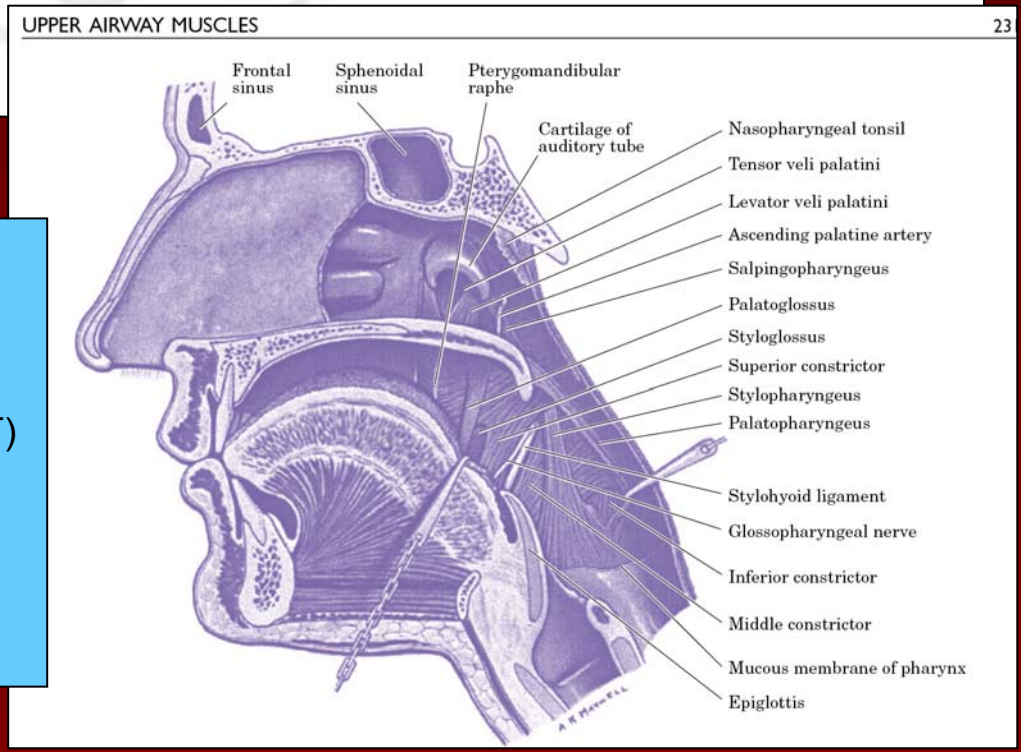
[1] Internal Medicine Department, Vall D'Hebron General Hospital, Universitat Autonoma Barcelona, C/Siracusa No. 12 Bis "A," Barcelona, Spain
[2] Pneumology Service, Vall D'Hebron General Hospital, Universitat Autonoma, Barcelona, Spain
[3] Clinical Neurophysiology Service, Vall D'Hebron General Hospital, Universitat Autonoma Barcelona, Barcelona, Spain
[4] Ciber, Enfermedades Respiratorias (Ciberes)

Accepted 18 September 2008

Estudio prospectivo y observacional en una cohorte 16 pacientes

Epworth Sleepiness Scale (ESS)
Myositis Disease Activity Assessment Tool (MDAAT)
Myositis Damage Index (MDI)

87% IAH > 5
7 CPAP, 4 bien tolerada y eficaz
Menos fatiga, pero no menos actividad



Tratamiento

Glucocorticoides

N-acetil cisteína 600 mg /8h
N Engl J Med 2005

Ciclofosfamida (pulsos) 0,7 gr /m^2
Rheumatology (Oxford) 2006
Lupus 2005

Tacrolimus / Ciclosporina
Lancet 1999
Arthritis Rheum 2005
J Rheumatol 2005
J Rheumatol 2008

Terapia biológica
Anti-TNF? Rituximab?

Trasplante Pulmonar

ARTHRITIS & RHEUMATISM
Vol. 52, No. 8, August 2005, pp 2439–2446
DOI 10.1002/art.21240

Treatment of Antisynthetase-Associated Interstitial Lung Disease With Tacrolimus

Margaret R. Wilkes, Susan M. Sereika, Noreen Fertig, Mary R. Lucas, and Chester V. Oddis

Combination Therapy with Corticosteroids, Cyclosporin A, and Intravenous Pulse Cyclophosphamide for Acute/Subacute Interstitial Pneumonia in Patients with Dermatomyositis

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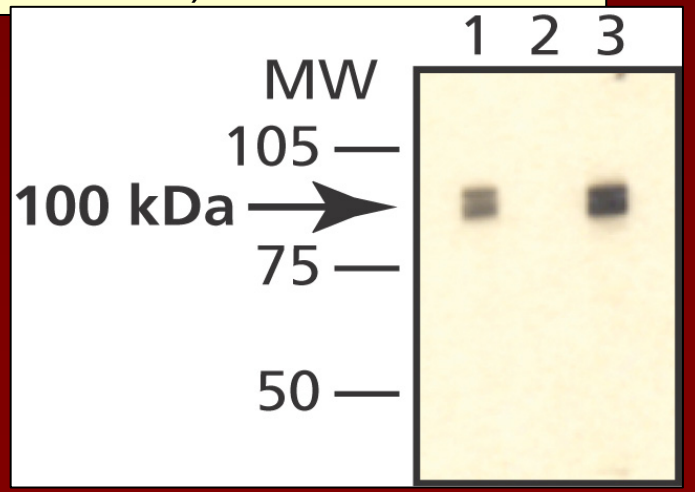
Tacrolimus-ILD-PM/Scl

En 4 de los 5 pacientes que recibieron tacrolimus, se produjo una mejoría de la CVF > 10% y en un caso al retirarlo por HTA, empeoró > 10%
Promedio de mejora de la CVF fue de 16% (IR 11,5-26,5)

p<0,05

De los 5 pacientes tratados alternativamente, sólo en 1 caso mejoró la CVF 10% tratado con GC y seguimiento < de 6 meses, en otro caso la CYF y posteriormente MTX la CVF se mantuvo estable, en el resto empeoró.

Promedio de mejora de la CVF fue de 2,5% (IR -7,5-36,5)



Conclusiones

1) La afectación pulmonar en la DM-PM es frecuente

2) Existen otras formas aparte de la neumopatía intersticial

3) Los anticuerpos antisintetasa no comportan un mal pronóstico

4) El estudio histológico no es determinante

5) Existen opciones terapéuticas útiles y que hay que conocer