ENFOQUE DEL NIÑO CON PATOLOGÍA MUSCULAR AUTOINMUNE

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UNIVERSIDAD SAN SEBASTIAN SEDE PATAGONIA PUERTO MONTT

59° CONGRESO CHILENO DE PEDIATRÍA PUERTO VARAS OCTUBRE 2019

Hereditarias

Distrofias musculares

Distrofinopatías (Duchennne y Becker)

Distrofias de cinturas

Distroafia facioescapulohumeral

Distrofia oculofaríngea

Distrofias musculares congénitas

Miopatías distales

Miopatías miofibrilares

Miopatías metabólicas

Miopatías lipídicas

Glucogénesis musculares

Mitocondriopatías

Síndromes miotónicos distróficos

Síndromes miotónicos no distróficos

Miotomía congénita (Thomsen, Becker)

Miotonías del canal del sodio

Paramiotonía congénita

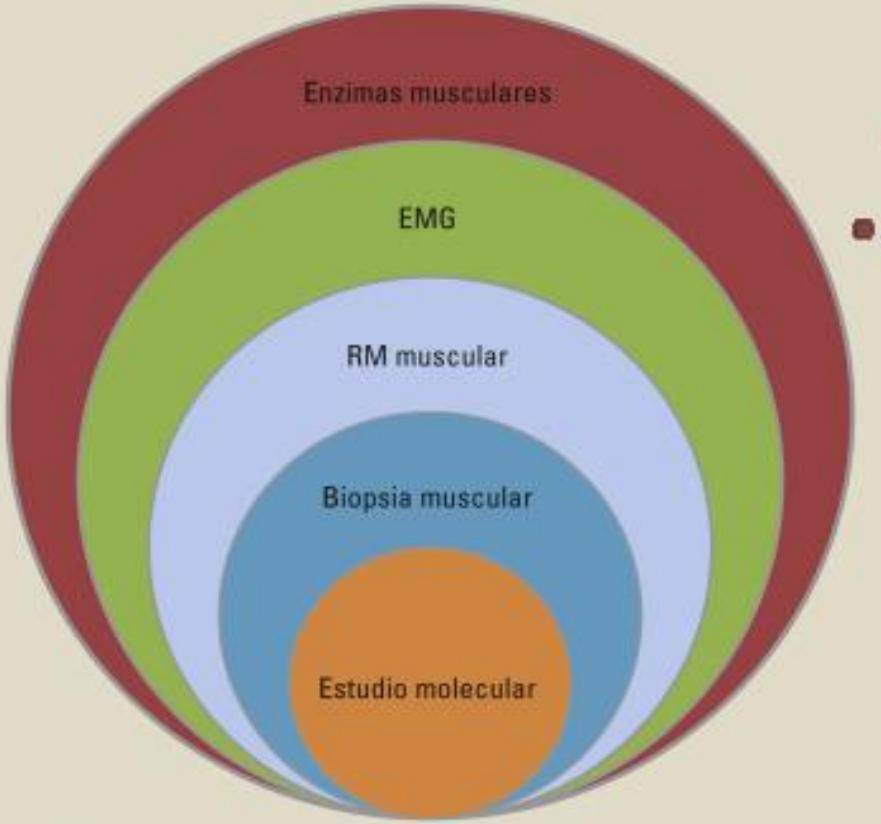
Parálisis periódicas

Miopatías congénitas

Antecedentes personales y familiares

+

Examen físico



Adquiridas

Miopatías inflamatorias idiopáticas

Dermatomiositis

Polimiositis

MCI*

Miopatías necrosante inmunomediada

Miositis focal y de causas infecciosas

Miopatías endocrinológicas

Miopatías en enfermedades sistemáticas

Miopatías tóxicas medicamentosa

Miopatías del enfermo crítico

Miofascitis macrofágica

Carnitina libre en sangre y orina
Acilcarnitina en sangre
Biopsia de piel (cultivo de fibroblastos)
Frotis de sangre periférica

Curva de lactato y aminio
Actividad de alfa-glucosidasa en sangre seca
Anticuerpos específicos o asociados a miositis
Estudio hormonal según clínica

POLIMIOSITIS Y DERMATOMIOSITIS

- Enfermedades de causa desconocida, de carácter inmune, manifestadas por lesión inflamatoria muscular asociada a necrosis de células musculares.
- En USA 3 casos por 100.000.
- Polimiositis
  - Enfermedad rara
  - Afecta al 45-60% adultos y < 5% niños.
- Dermatomiositis
  - Adultos y niños menores de 18 años.
  - La incidencia anual estimada JDM es 0.19–4.1 casos por millón de niños

AMBAS más frecuentes en mujeres.

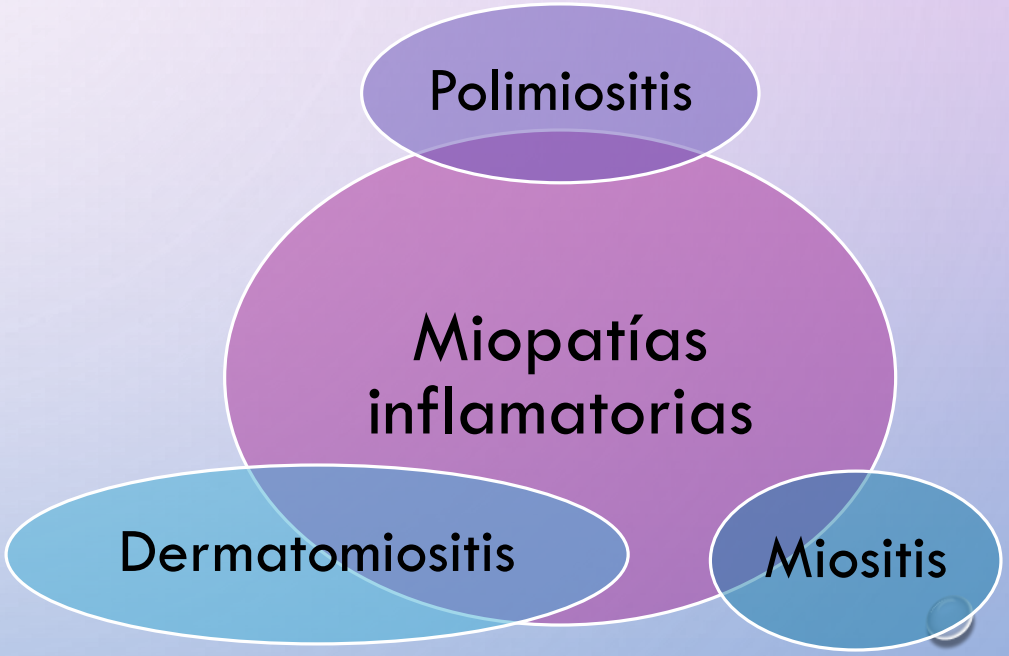


Table 2 Summarises the most notable genetic acquisitions on IIM

	Increased IIM risk [27, 31–34]	Reduced IIM risk [30, 34]
HLA	-B*0801	DQA1*0201
	-A*0101	DQA1*0101
	-DRB1*0301	DQA1*0102
	-DQA1*0501	DR4
	-DPB1*0101	DR7
	-DQA1*0301	
	-DQB1*02	
	-DRB1*15021 (Japanese patients)	
Other genes		Correlated features
	IL-1: polymorphism IL-1α-889CC [31]	Higher risk for calcinosis
	PTPN22 gene: variant R620W [35]	Associated with juvenile and adult idiopathic inflammatory myopathy
	TNF-α gene: variant TNF308A [37]	Higher risk for calcinosis and ulcerations

IIM inflammatory idiopathic myositis, *HLA* human leukocyte antigen, *PTPN22* protein tyrosine phosphatase N22, *TNF* tumour necrosis factor, *IL-1* interleukin-1

Ilaria Pagnini
Clinic Rev Allerg Immunol
DOI 10.1007/s12016-015-8512-9

Criterios Diagnósticos

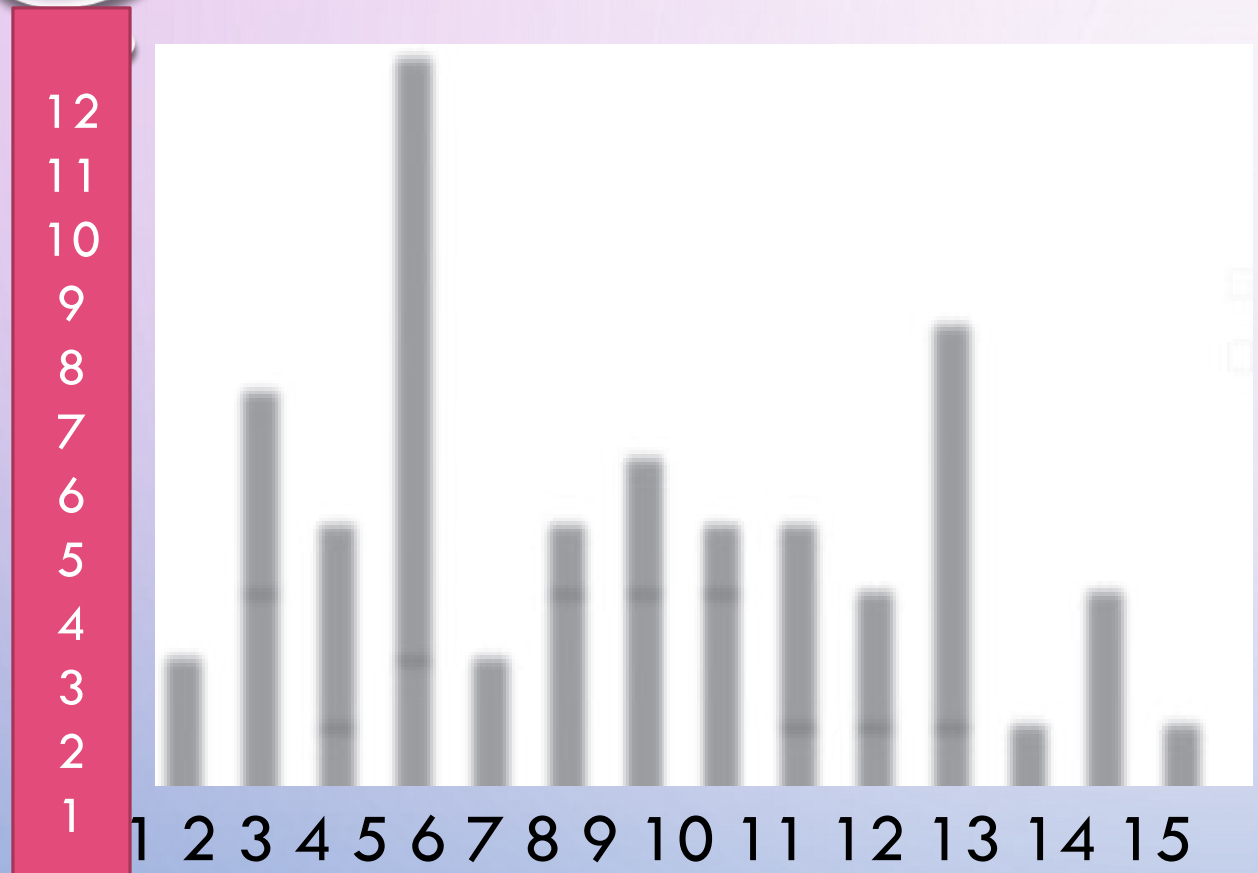
TABLA 1.
Clasificación de las enfermedades musculares inflamatorias idiopáticas

Tipo I	Polimiositis idiopática primaria
Tipo II	Dermatomiositis idiopática primaria
Tipo III	Polimiositis o dermatomiositis asociada con neoplasias malignas
Tipo IV	Polimiositis o dermatomiositis juvenil
Tipo V	Polimiositis o dermatomiositis asociada con enfermedades del tejido conectivo (síndromes de sobreposición)
Tipo VI	Miositis de cuerpos de inclusión
Tipo VII	Misceláneas: miositis de cuerpos de inclusión, miositis eosinofílica y miositis nodular localizada

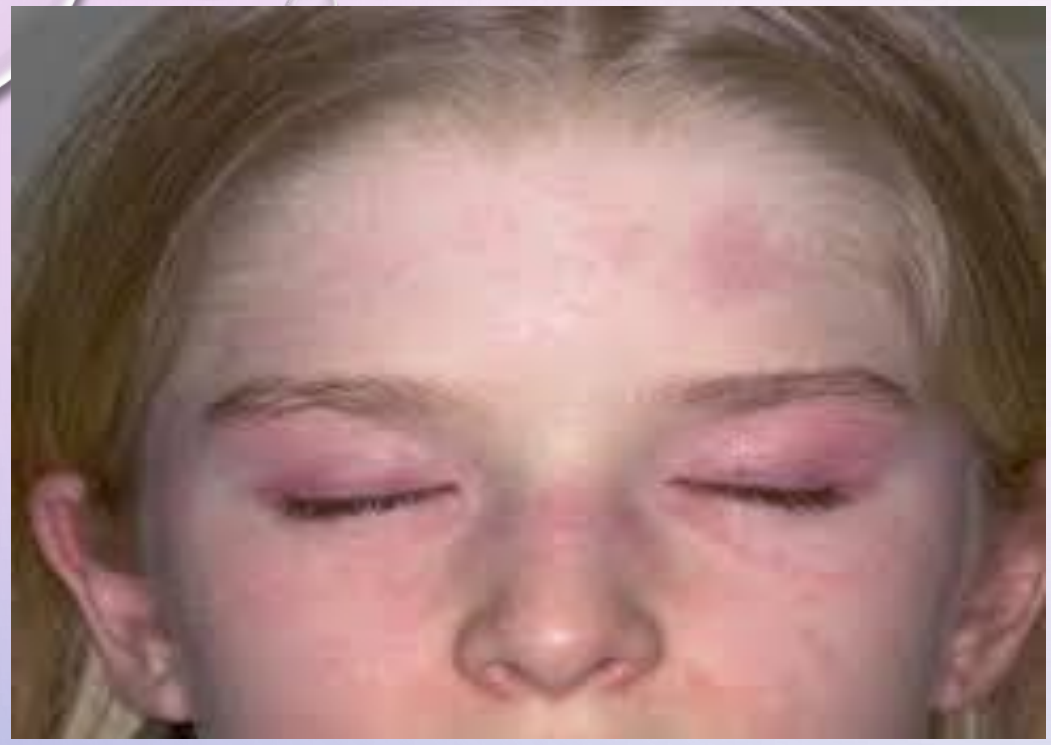
Clasificación de Bohan y Peter (1975)

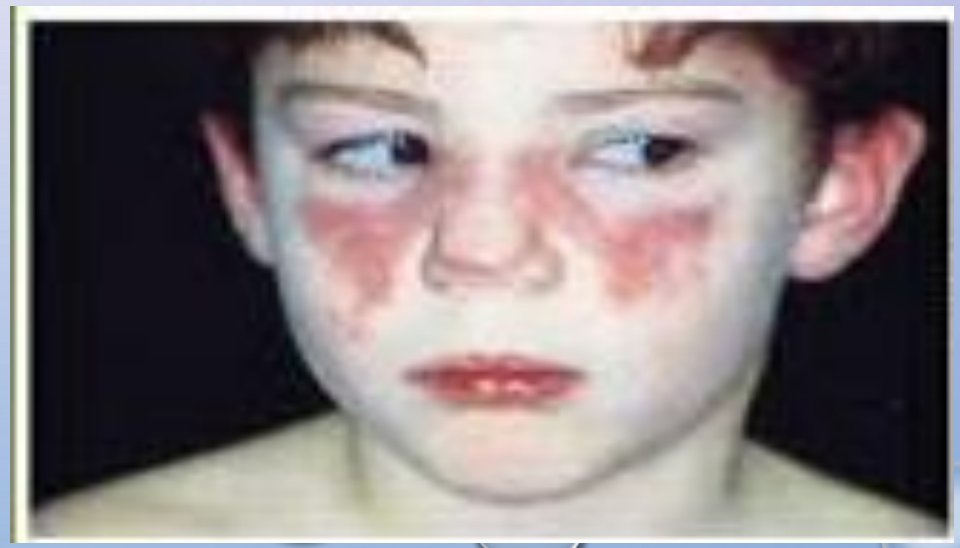
CRITERIOS DE	BOHAN Y PETER 1975
CRITERIO	DEFINICIÓN
1. Debilidad muscular proximal.	Debilidad muscular en la cintura escapular o pélvica.
2. Enzimas sarcoplásmicas elevadas.	Elevación de las enzimas musculares en suero (CK-Aldolasas - aspartate aminotransferase - alanine aminotransferase - lactate dehydrogenase)
3. Cambios miopáticos en la electromiografía.	Potenciales de acción característicos de miopatía.
4. Biopsia muscular.	Inflamación crónica con degeneración y reparación de fibras musculares.
5. Pápulas de Gotron o Heliotropo.	Pápulas planas en nudillos y rash hiperémico alrededor de dedos, párpados y cara.

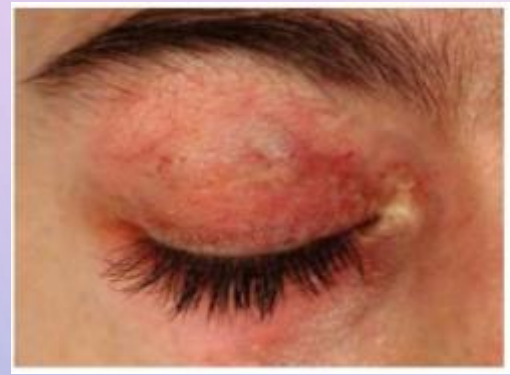
Nuevos criterios 2016
Typical findings on muscle magnetic resonance imaging (MRI) and ultrasonography.
Nailfold capillaroscopy abnormalities
Calcinosis
Dysphonia

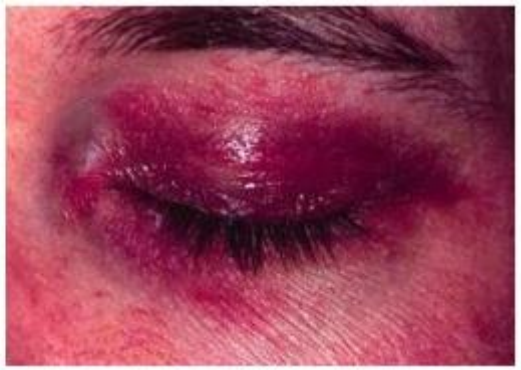


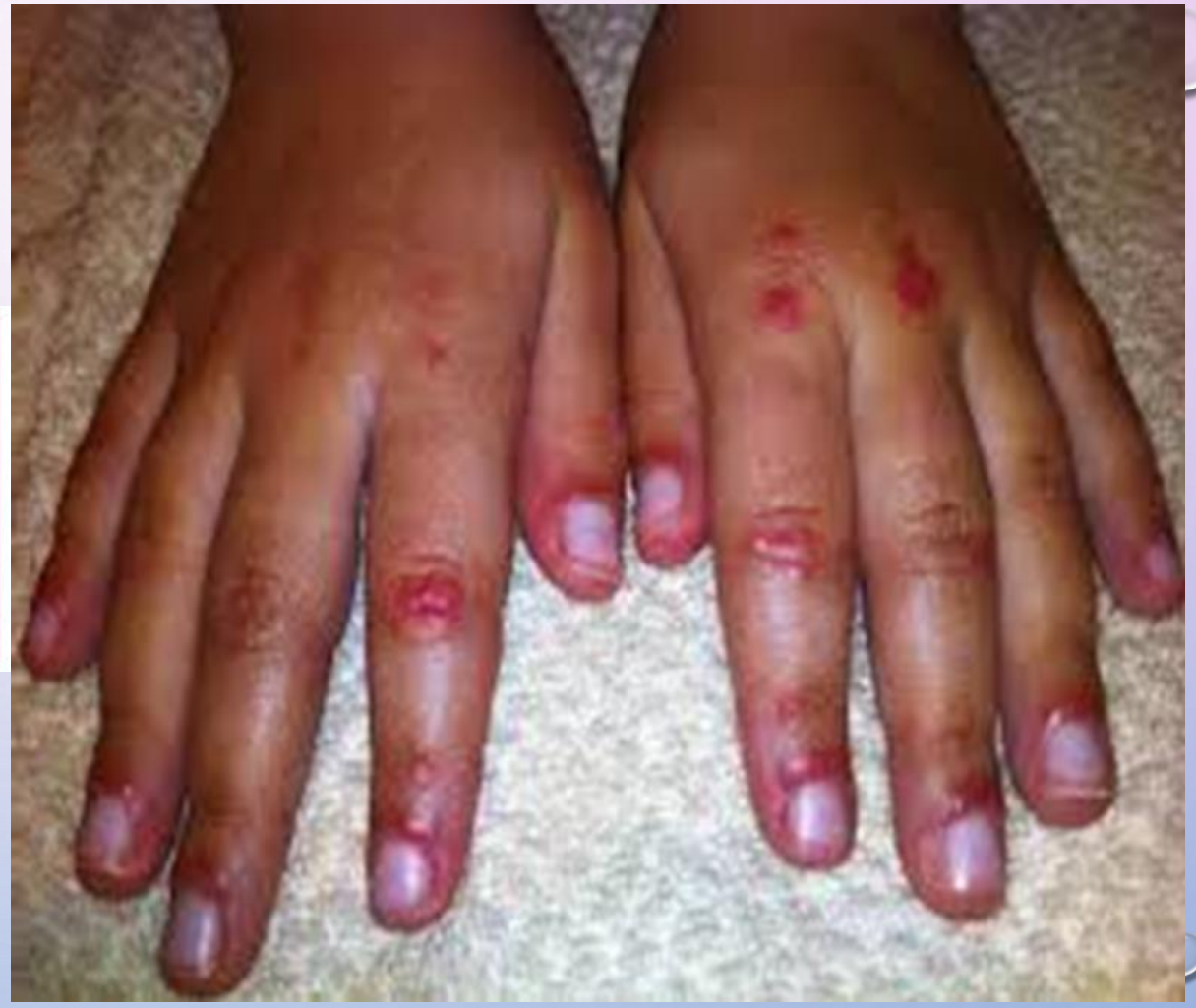
Peter J. GOWDIE, International Journal of Rheumatic Diseases 2013; 16: 561–567











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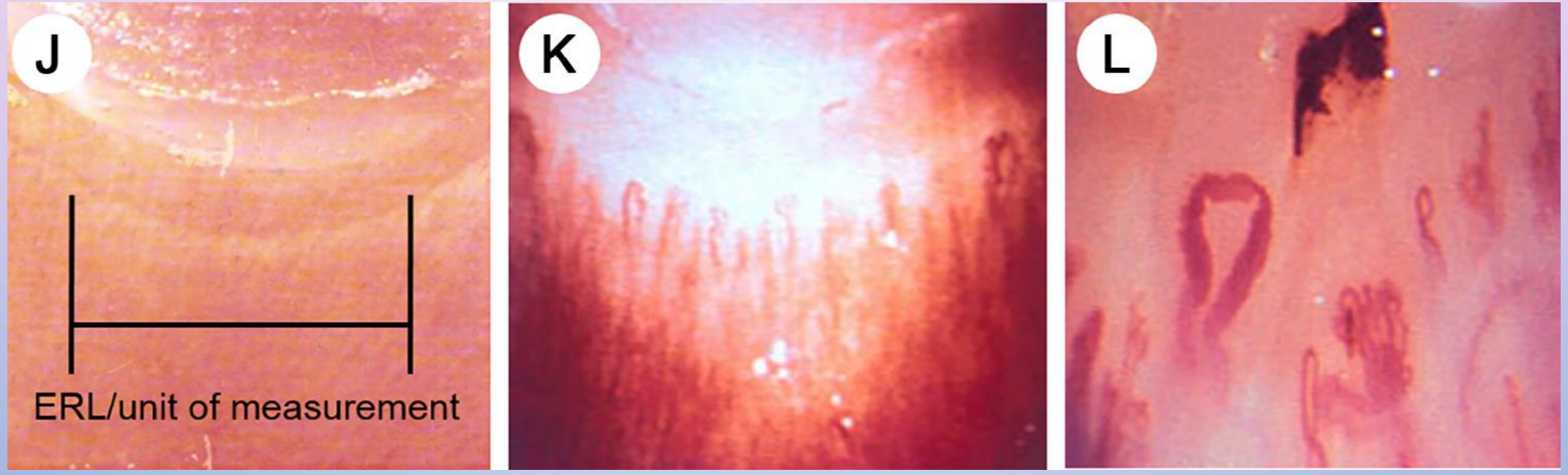
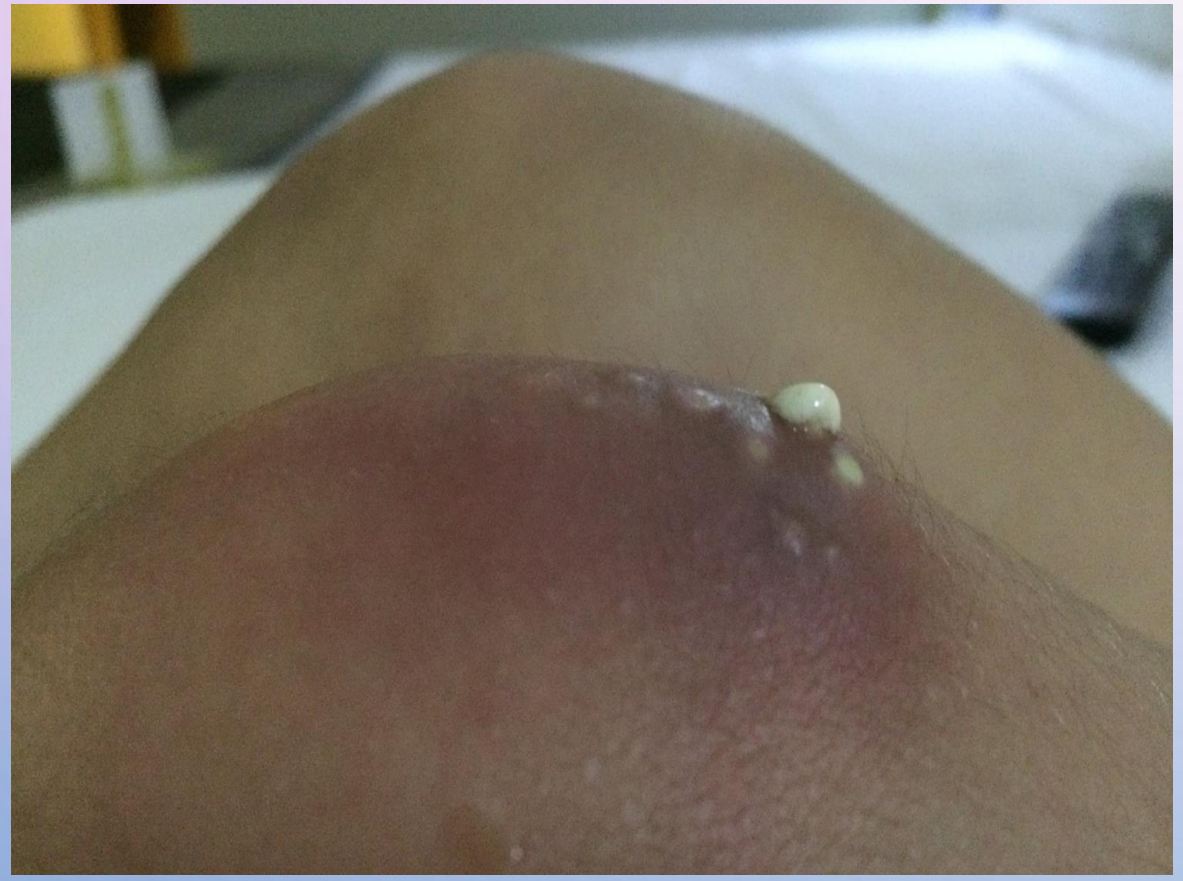
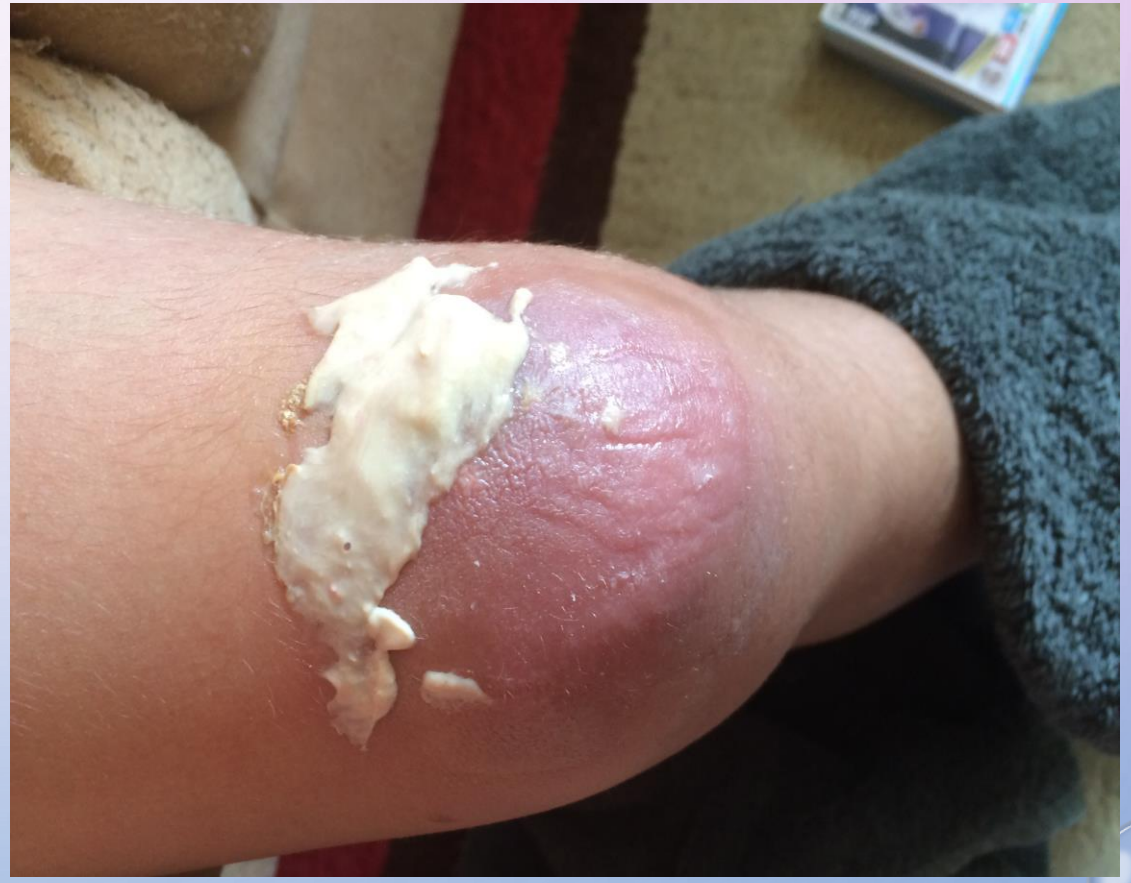
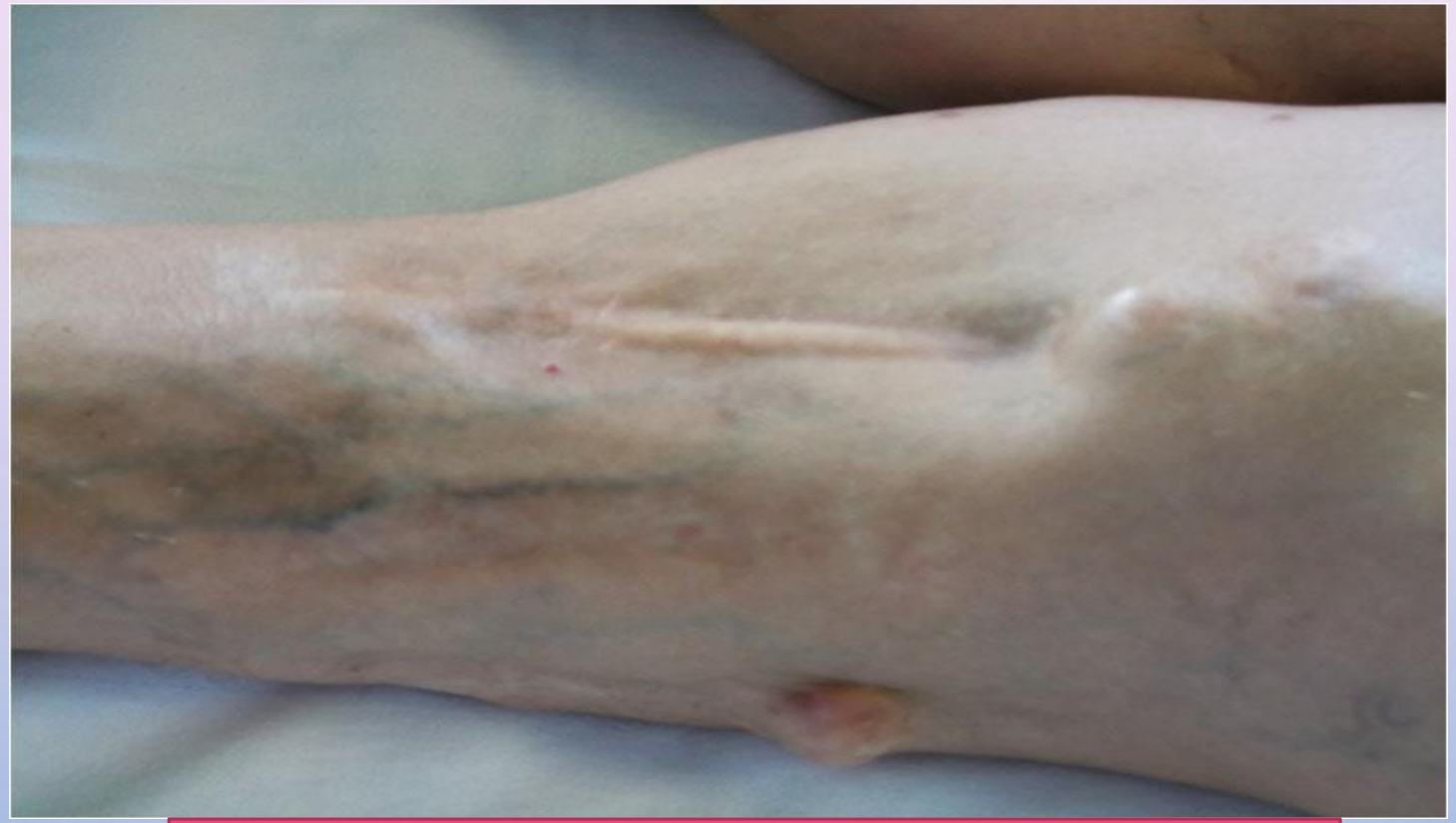


Pachmann J of Pediatrics 2017



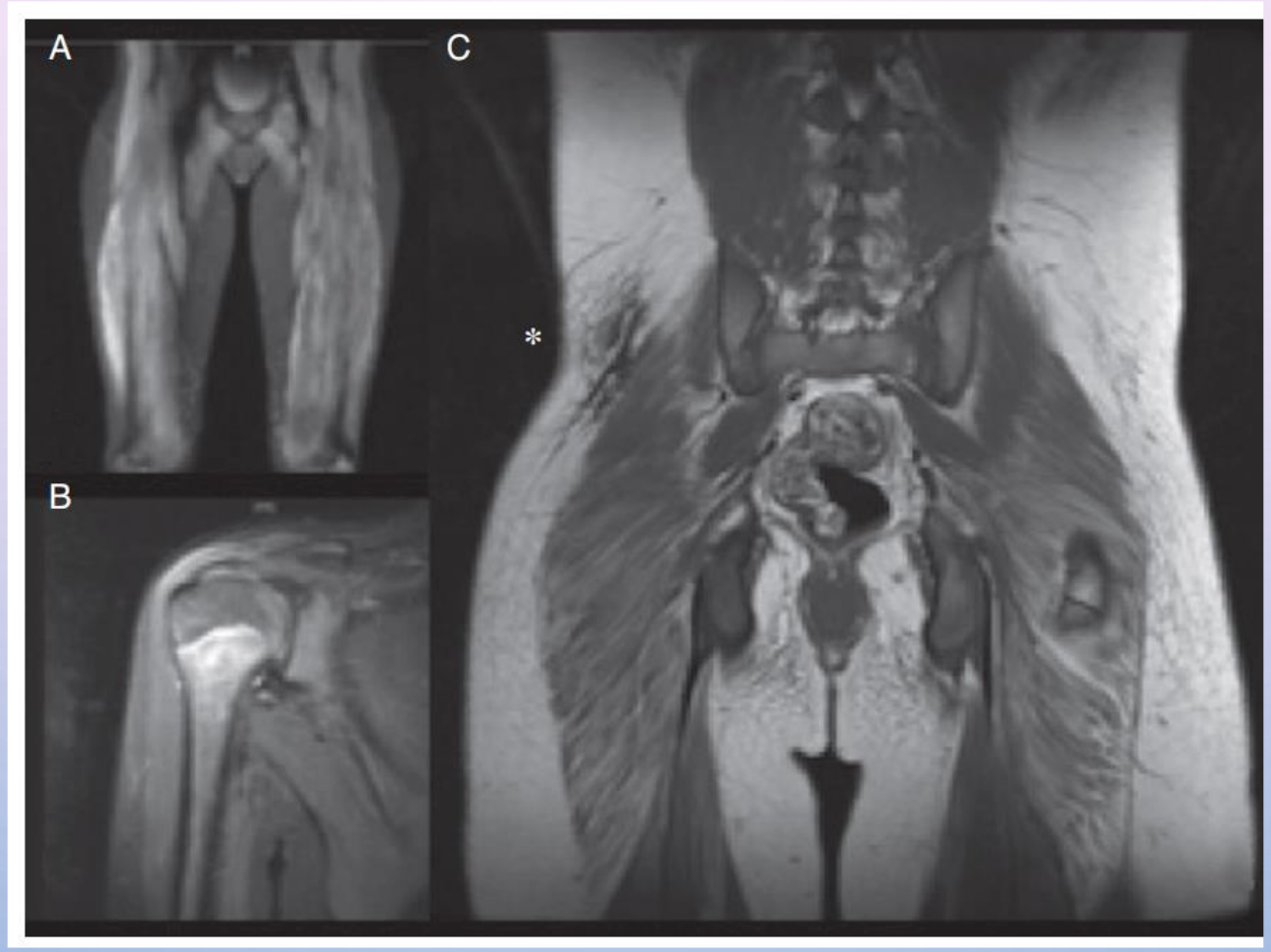




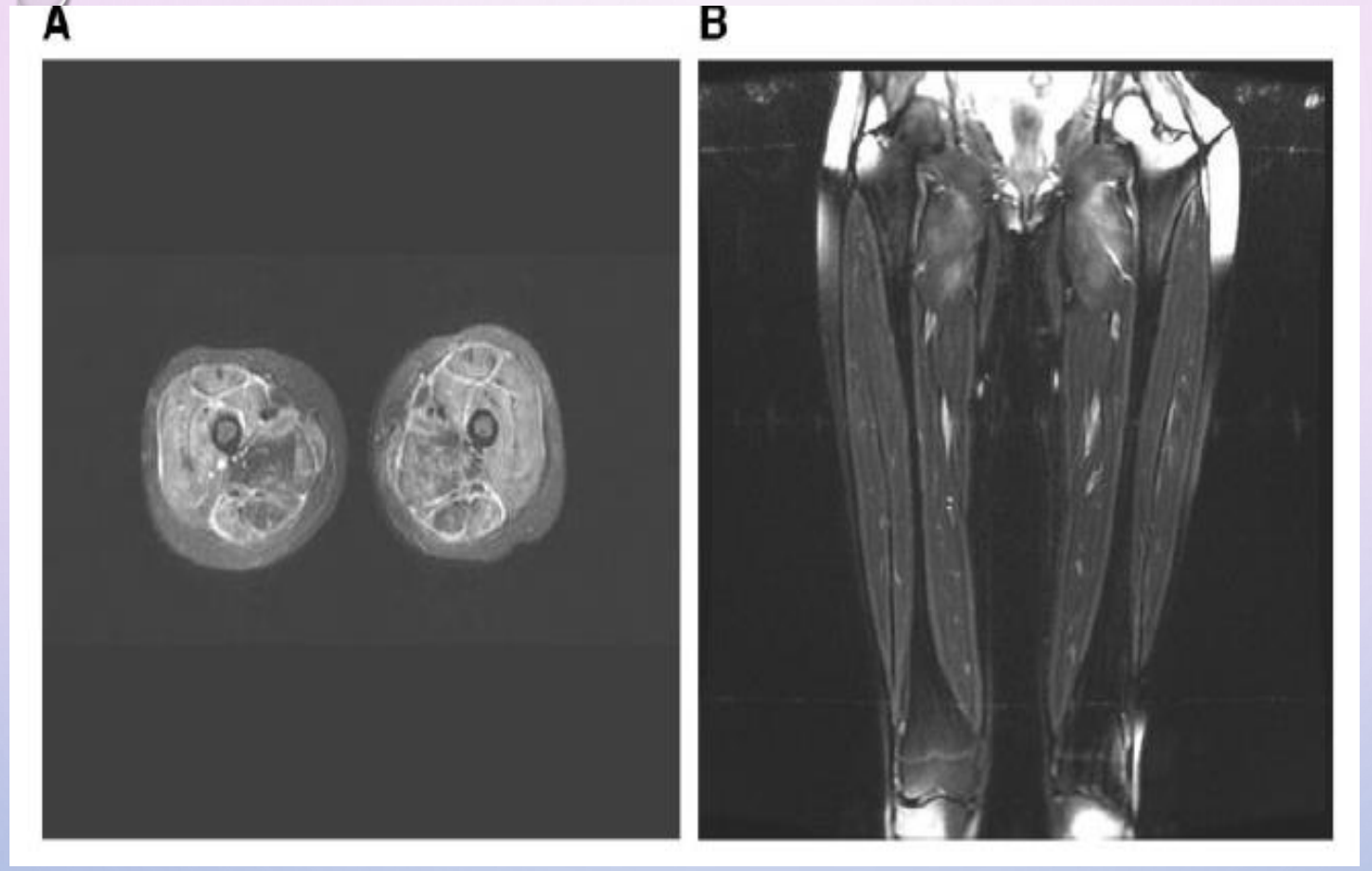
lipodystrophy at the popliteal fossa in a dermatomyositis

Signo de Gower

- Debilidad muscular
- Apoyo de manos



aña O, et al. La resonancia magnética muscular y el tratamiento agresivo precoz
Actas Dermosifiliogr. 2017. http://dx.doi.org/10.1016/j.ad.2017.07.003



Mandela Thyoka

doi:10.1093/rheumatology/key144

2018

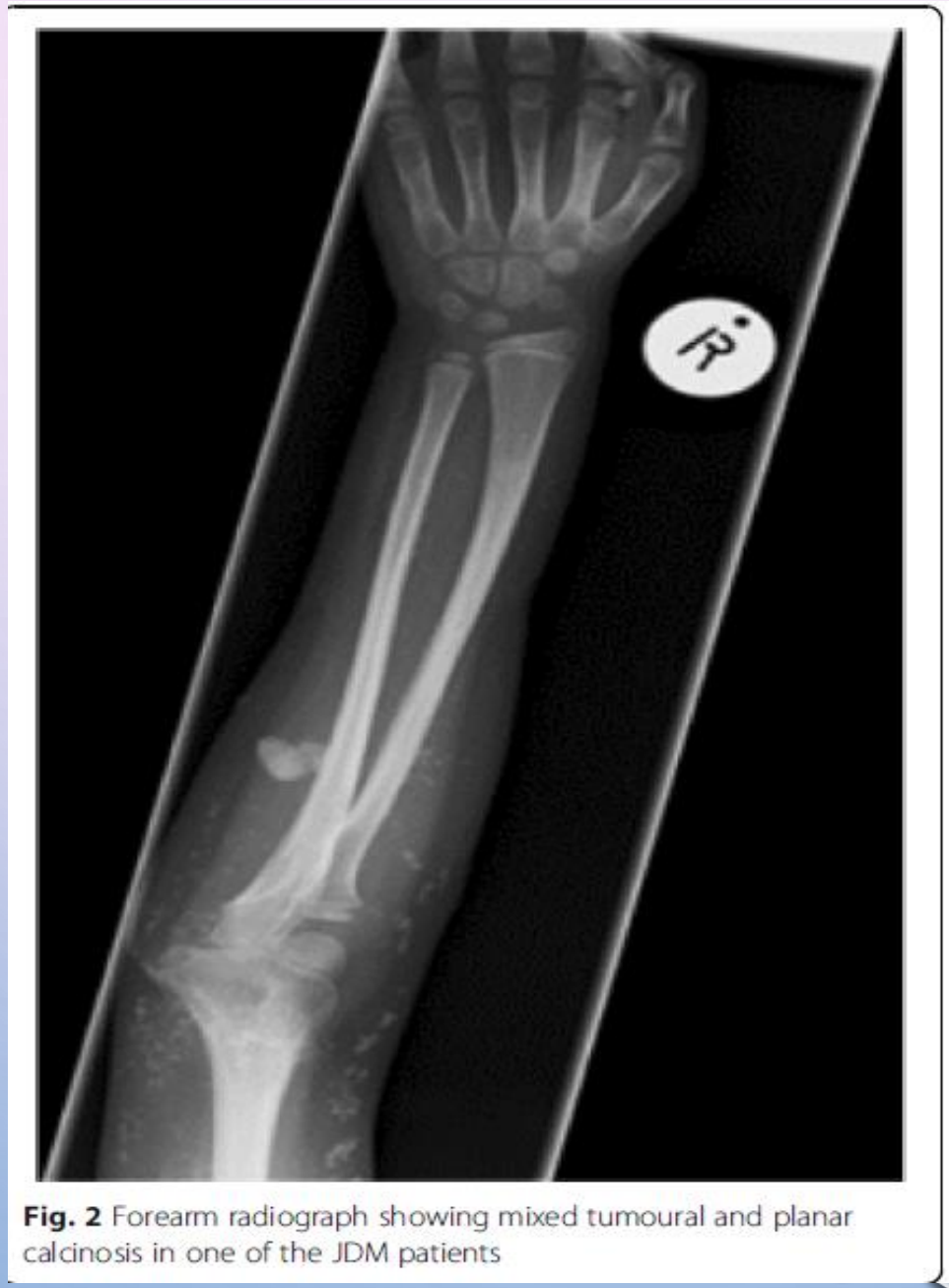


Fig. 2 Forearm radiograph showing mixed tumoural and planar calcinosis in one of the JDM patients

Table 1 Clinical features of juvenile dermatomyositis at onset and accrued throughout course of illness

	At presentation† (%)	Over disease course† (%)
Lethargy	45/45 (100)	47/47 (100)
Weakness	54/57 (95)	56/57 (98)
Gottron's papules	51/56 (91)	53/57 (93)
Myalgia/arthralgia	46/51 (90)	
Gower's sign	40/45 (89)	43/49 (88)
Malar rash	30/38 (79)	36/51 (71)
Heliotrope rash	36/49 (73)	38/55 (69)
Nailfold changes	26/38 (68)	38/49 (78)
Weight loss/anorexia	25/41 (61)	30/48 (65)
Contractures	17/29 (59)	28/45 (62)
Poikiloderma	8/15 (53)	11/26 (42)
Dysphonia	14/31 (45)	
Mouth ulcers	11/29 (38)	20/38 (53)
Arthritis	15/43 (35)	18/49 (37)
Fever	16/45 (36)	
Dysphagia	11/44 (25)	
Dysphonia or dysphagia		21/49 (43)
Skin ulceration	3/13 (23)	4/17 (24)
Lymphadenopathy	8/35 (23)	10/39 (26)
Gastrointestinal involvement	6/38 (16)	8/40 (20)
Pulmonary involvement	4/34 (12)	6/36 (17)
Hepatomegaly	5/41 (12)	6/46 (13)
Calcinosis	0/13 (0)	8/45 (18)

Table 2 Results of investigations at diagnosis in descending order of frequency of use.

	No. of patients with abnormal result at diagnosis (%)
Creatine kinase	37/57 (65)
Aspartate aminotransferase	29/33 (88)
Alanine aminotransferase	19/33 (58)
Lactate dehydrogenase	23/25 (92)
Aldolase	10/10 (100)
Erythrocyte sedimentation rate	46/55 (84)
Electromyogram	4/4 (100)
Magnetic resonance imaging	28/29 (97)
Muscle biopsy	24/29 (83)
Antinuclear antibodies	33/52 (63)
Rheumatoid factor	1/18 (6)
Extractable nuclear antigen	1/29 (3)
Double-stranded DNA	0/15 (0)

Peter J. GOWDIE, International Journal of Rheumatic Diseases 2013; 16: 561–567 AUSTRALIA

Clínica 115 pacientes Canadá

Sign or symptom	JDM patients at presentation,%	JDM patients at any time,%
Constitutional features		
•Fever	16	Not applicable
•Adenopathy	8	20
•Anorexia	18	Not applicable
Cutaneous features		
•Gottron's rash	91	Not applicable
•Heliotrope rash	83	Not applicable
•Malar/facial rash	42	Not applicable
•Nailfold capillary changes	80	91
•Calcinosis	3	30
•Lipodystrophy	0	14
Musculoskeletal features		
•Myalgia/arthralgia	25	Not applicable
•Arthritis	6	58
•Dysphonia or dysphagia	24	41
Gastrointestinal involvement	5	22
Pulmonary involvement	11	32

Michelle Batthish & Brian M. Feldman Curr Rheumatol Rep (2011) 13:216–224

Table 1 Disease manifestations among the JDM cases in Cape Town

	Cumulative clinical manifestations (%)	Clinical manifestations at last review
Skin		
Calcinosis	11 (44)	8 (32)
Skin ulcers	11 (44)	1
Oedema (Generalized, Periorbital or limb)	11 (44)	0
Alopecia	3 (12)	3 (12)
Lipodystrophy	1 (4)	1 (4)
Musculoskeletal		7 (28)
Muscle tenderness	13 (52)	0
Arthritis	7 (28)	2 (8)
Contractures	5 (20)	5 (20)
Osteoporosis with fracture	1 (4)	1 (4)
Endocrine	9 (36)	9 (36)
Growth failure	8 (32)	8 (32)
Diabetes mellitus	1 (4)	1 (4)
Adrenal insufficiency	1 (4)	0
Respiratory	3 (12)	3 (12)
Interstitial lung disease (ILD)	2 (8)	2 (8)
Dysphonia	2 (8)	0
Pulmonary hemorrhage	1 (4)	1 (4)

	Cumulative clinical manifestations (%)	Clinical manifestations at last review
Gastrointestinal		
Dysphagia	5 (20)	0
Abdominal pain or bleeding	3 (12)	0
Ocular	2 (8)	2 (8)
Cataracts	2 (8)	2 (8)
Cardiovascular		
Abnormal capillaroscopy	15 (60)	NA
Raynaud's phenomenon	6 (24)	NA
Cardiomyopathy	1 (4)	0
Nervous system	3 (12)	1 (4)
Seizures; neuropathy	2 (8)	0
Sensorineural hearing loss	1 (4)	1 (4)
Infections		
Fungal (skin)	7 (28)	NA
Bacterial (staph aureus 2, gram negative 1)	3 (12)	NA
TB	2 (8)	NA

50 pacientes Turquía 2017

Table 2 Laboratory investigation results

Investigation	Reference range	Median (IQR)	No. positive/ No. tested	Percentage
Enzymes				
CK	26–145 U/L	1074.0 (222.8–3397.5)	22/24	91.7
Aldolase	3.0–12.0U/L	12.6 (7.7–15.9)	3/5	60
LDH	142–261U/L	445.0 (277.0–493.5)	14/19	73.7
AST	0–41 U/L	74.0 (35.25–140.5)	16/22	72.7
ALT	5–25 U/L	48.0 (26.5–82.0)	18/23	78.3
Autoantibody				
Anti-Jo1	<7 EliA U/ml	0.3 (0.3–0.45)	0/6	0
Anti-RNP	<7 EliA U/ml	0.3 (0.3–0.5)	0/9	0
Inflammatory markers				
ESR		20 (12–35)	11/19	57.9
CRP		2.6 (<1–6)	2/10	20
vWF activity		77 (62–112 %)	2/4	50

Table 1 Demographic and clinical findings of 50 patients with juvenile dermatomyositis

Demographics	
Gender [*n* (%)]	
Female	35 (70%)
Male	15 (30%)
Age at the onset of symptoms [mean ± SD, years]	6.1 ± 4.1
Min–max	1.5–16.0
Age at the onset of diagnosis [mean ± SD, years]	6.6 ± 4.1
Min–max	2.0–16.0
Clinical findings	
Heliotrope rash [*n* (%)]	50 (100%)
Gottron papule [*n* (%)]	48 (96%)
Muscle weakness [*n* (%)]	45 (90%)
Erythematous rash [*n* (%)]	44 (88%)
Calcinosis [*n* (%)]	19 (38%)
Distribution of calcinosis [*n* (%)]	
Upper extremity	1 (5.3%)
Lower extremity	1 (5.3%)
Upper and lower extremity	6 (31.6%)
Trunk	2 (10.5%)
Diffuse	9 (47.4%)

Kenan Barut1
Clin Rheumatol 2017 DOI 10.1007/s10067-016-3530-4

Table 2 Laboratory findings at the disease onset of the patients

Hemoglobin [mean ± SD, g/dL]	11.5 ± 1.4
Min–max	9.0–16.0
Leukocyte count [mean ± SD,/mm^3]	9.616 ± 3.393
Min–max	3.800–16.300
Thrombocyte count [mean ± SD,/mm^3]	332.250 ± 121.179
Min–max	139.000–812.000
Erythrocyte sedimentation rate [mean ± SD, mm/h]	35 ± 22.1
Min–max	6.0–107.0
Creatine kinase [mean ± SD, IU/L]	223,326 ± 329,201
Min–max	20–13.050
C-reactive protein [*n* (%)]	
Positive	19 (38%)
Negative	31 (62%)
Antinuclear antibody [*n* (%)]	
Positive	34 (68%)
Negative	16 (32%)
Anti-jo 1 negativity [*n*]	30
ENA negativity [*n*]	17
The presence of EMG findings [*n*]	27
The presence of muscle biopsy findings [*n*]	14

EMG electromyography, *ENA* extractable nuclear antigen

TABLE 3. *Summary of Physical Exam, Laboratory, and Ancillary Testing Results*

	All JDM patients	Symptomatic	Asymptomatic	Clinically amyopathic
Number of patients	46	26	20	10
Skin features, *n* (%)				
Heliotrope rash	29 (63)	17 (65)	12 (60)	5 (50)
Gottron papules	32 (70)	17 (65)	15 (75)	8 (80)
Gottron sign	36 (78)	18 (69)	18 (90)	8 (80)
Nailfold changes	33 (72)	18 (69)	15 (75)	6 (60)
Muscle symptoms, *n* (%)				
Weakness	26 (57)	26 (100)	0 (0)	0 (0)
Fatigue	31 (67)	24 (92)	7 (35)	3 (30)
Dysphagia	6 (13)	5 (19)	1 (5)	0 (0)
Shortness of breath	4 (9)	4 (15)	0 (0)	0 (0)
Change in voice	5 (11)	3 (12)	2 (10)	0 (0)
Muscle examination, *n* (%)				
MMT ≤ 4	32 (70)	24 (92)	8 (40)	0 (0)
Neck of hip flexion	22 (48)	16 (62)	6 (30)	0 (0)
Positive gower sign	15/34 (44)	14/17 (82)	1/17 (6)	0/7 (0)
Weakness detected by at least one modality	35 (76)	25 (96)†	10 (50)	0 (0)
Abnormal laboratory results, *n* (%)				
ESR	19/34 (56)	14/20 (70)	5/14 (36)	4/8 (50)
CK	27/46 (59)	19/26 (73)	8/20 (40)	2/10 (20)
Aldolase	33/45 (73)	23/25 (92)	10/20 (50)	2/10 (20)
AST	28/46 (61)	23/26 (85)	5/20 (25)	1/10 (10)
ALT	26/46 (57)	21/26 (81)	5/20 (25)	2/10 (20)
LDH	30/46 (65)	22/26 (85)	8/20 (40)	4/10 (40)
ANA	24/37 (65)	9/17 (53)	15/20 (75)	6/10 (60)
Abnormal diagnostic studies, *n* (%)				
MRI	31/42 (74)	21/24 (88)	10/18 (56)	4/9 (44)
Muscle biopsy	5/7 (71)	5/5 (100)	0/2 (0)	0/1 (0)
EMG	2/4 (50)	1/1 (100)	1/3 (33)	0/1 (0)
Skin biopsy	8/9 (89)	1/2 (50)	7/7 (100)	4/4 (100)

Edward J. Oberle, Pediatric Dermatology 1–8, 2016

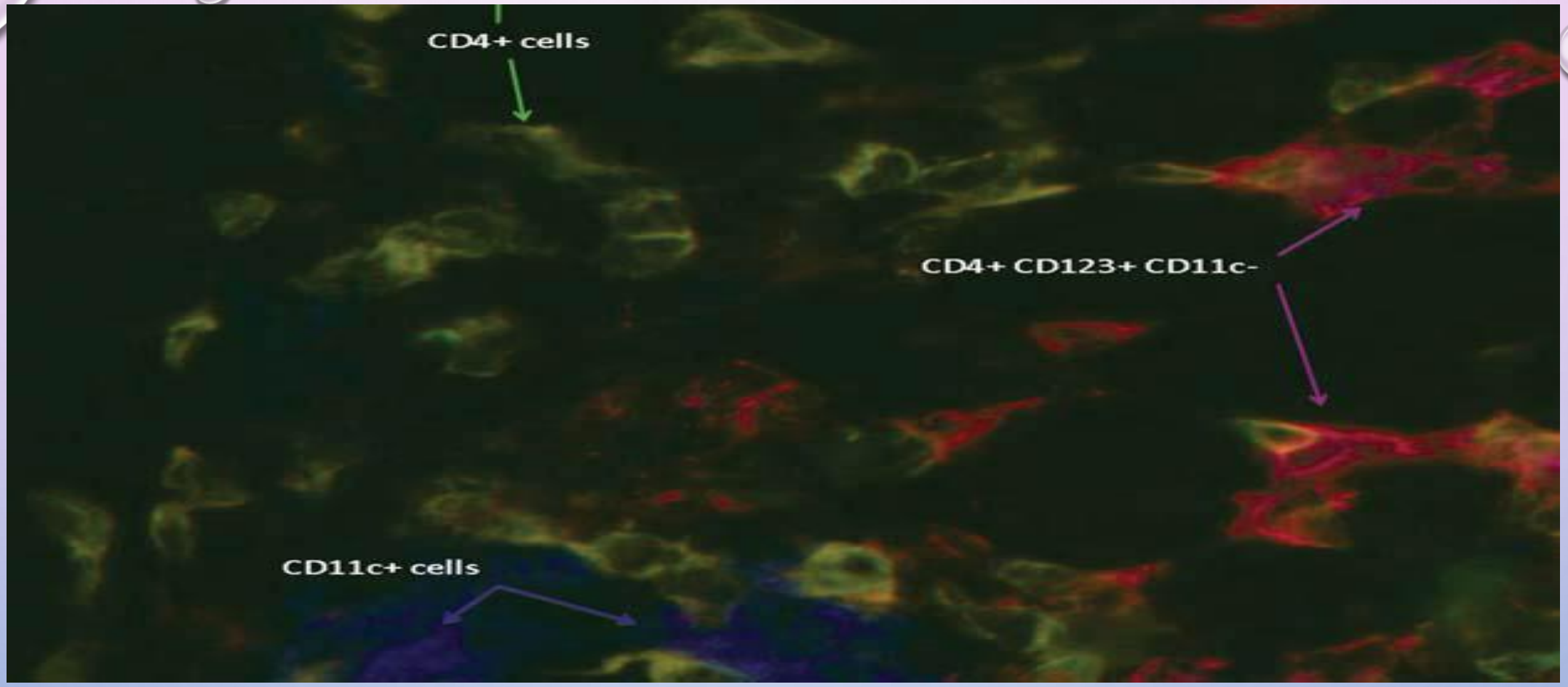
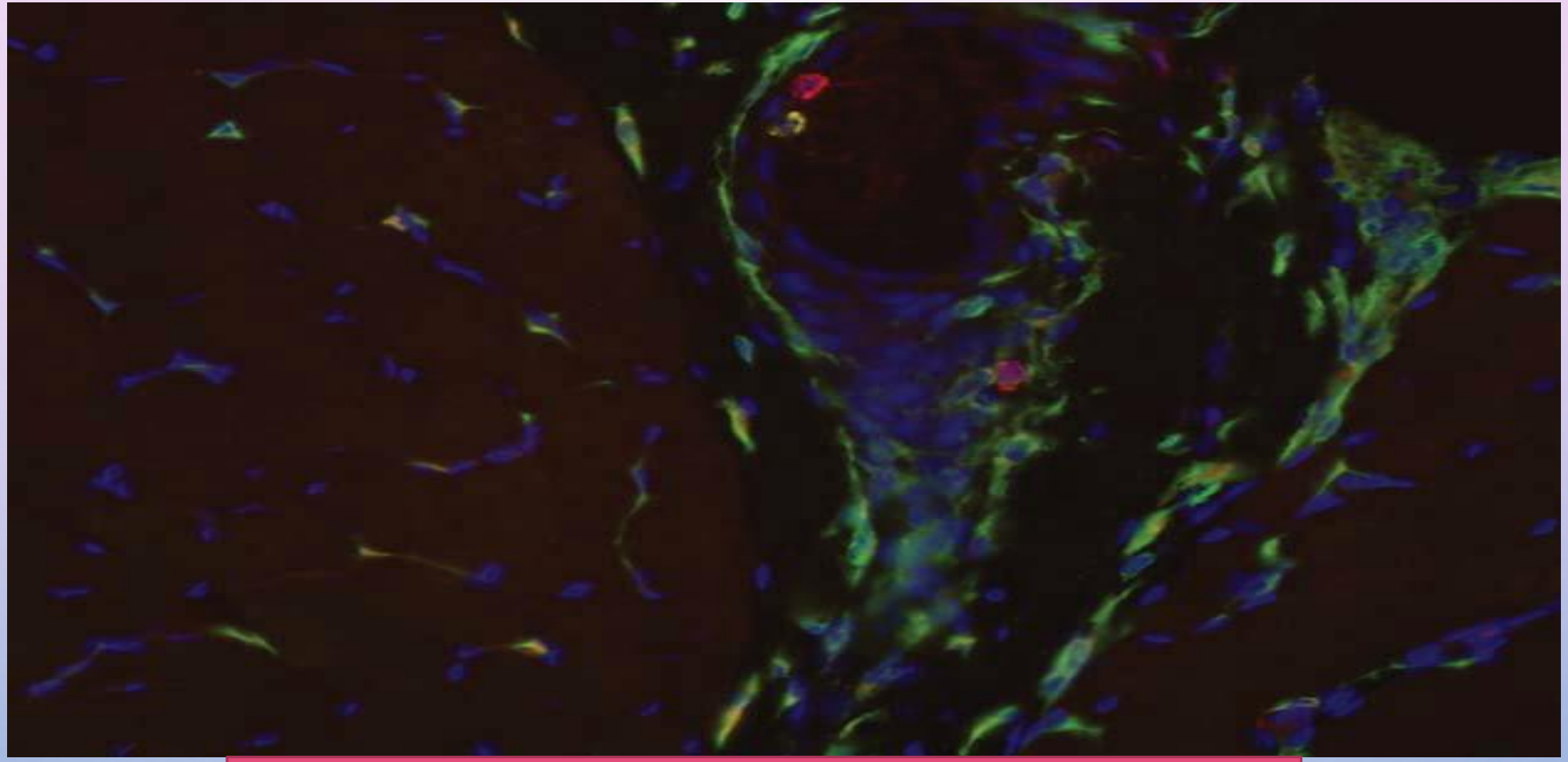
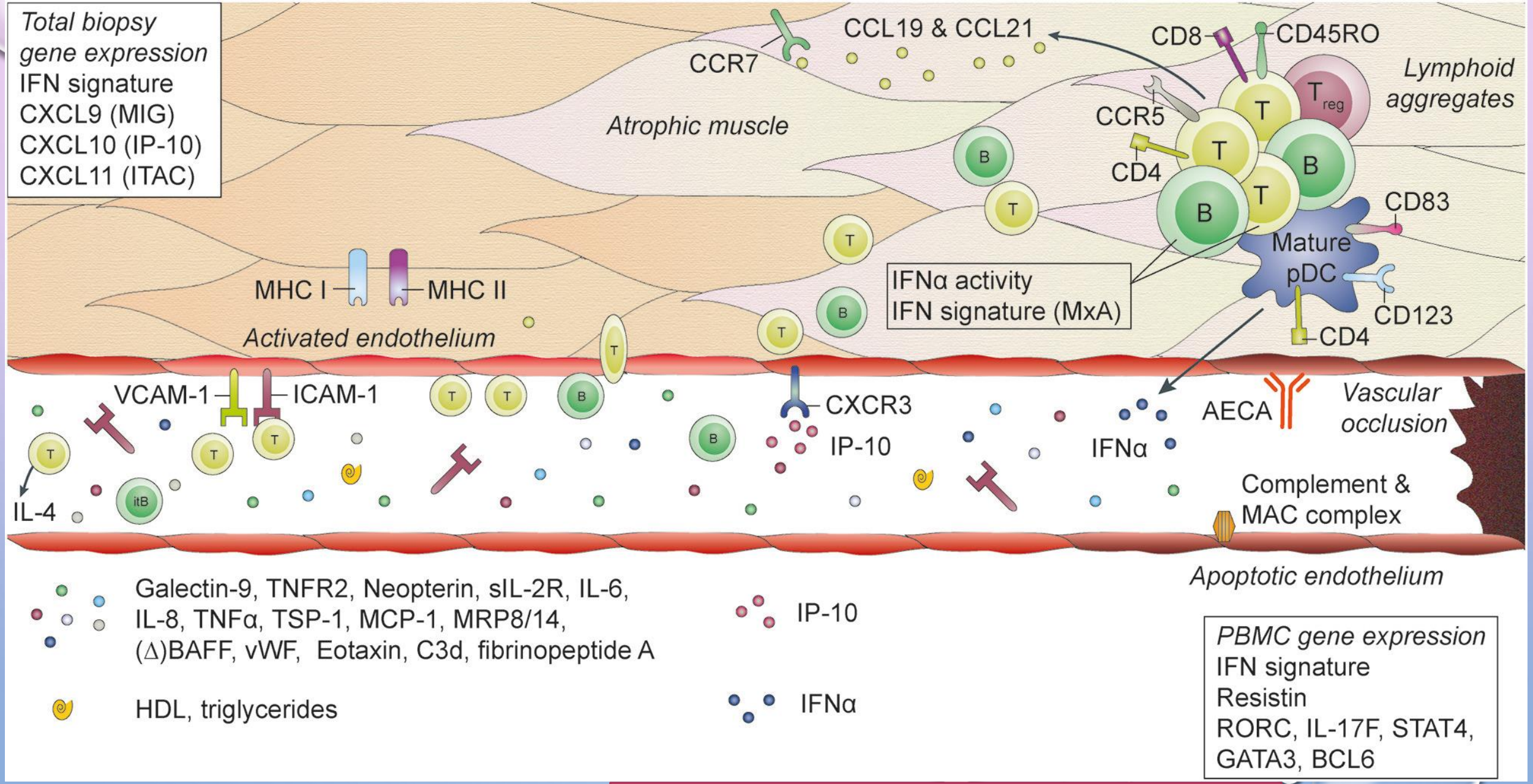


FIGURE 1. Lymphoid aggregation in muscle tissue from a newonset juvenile dermatomyositis subject stained for CD4
Þ cells (green), CD4 þCD123þ CD11c plasmacytoid dendritic cells (magenta) and CD11c þ myeloid dendritic cells (blue).



Inflammation surrounding a vessel in a new-onset juvenile dermatomyositis subject's muscle biopsy with CD4 þ cells (green) and immature CD4 þ D45RA þ immature T cells(magenta).



Wiencke Frontiers in immunology dec 2018

Antibody association with JDM	Target autoantigen	Clinical or laboratory association	Year	Investigators
ANA		HLA	2007	Wedderburn
Antisynthetase antibodies (ARS)	Cytoplasmic aminoacyltRNA synthetase	High frequency of interstitial lung disease	2007	Matsushita
Anti–Jo-1	Histidyl-tRNA synthetase	High frequency of interstitial lung disease and mechanic hands	1994 2006 2009	Rider O'Hanlon Chinoy
Anti–PL-12	Alanyl-transfer RNA synthetase		1994	Rider
Anti-SRP	Anti–signal recognition particle	Acute-onset severe disease with dysphagia and cardiac disease	2008	Rider Rouster-Stevens
Anti–Mi-2	Helicase protein	Cutaneous disease with milder muscle disease	1994 2006	Rider O'Hanlon
Anti-MJ or anti-p140	NXP-2	JDM with calcinosis	1997 2009	Oddis Gunawardena
Anti-p155/140	TIF1-c	Severe cutaneous involvement	2006	Targoff
CADM-140	Intracytoplasmic MDA5	Rapidly progressive interstitial lung disease	2005	Sato
Anti–PM-Scl	Exosome-associated proteins	Scleroderma overlap with DMJ	2007 2009	Wedderburn Chinoy, Schilders

TABLE 1. *Myositis-Specific Autoantibodies*

Autoantibody (alternate names)	Target autoantigen	Clinical associations	Frequency in juvenil dermatomyositis
Anti-synthetase autoantibodies (anti-ARS)	Cytoplasmic aminoacyl-tRNA synthetase	Antisynthetase syndrome: myositis, interstitial lung disease, arthritis, mechanic's hands, fever, Raynaud phenomenon	<3%
Anti-Jo-1	Histidyl-tRNA synthetase		
Anti-PL-7	Threonyl-tRNA synthetase		
Anti-PL-12	Alanyl-tRNA synthetase		
Anti-OJ	Isoleucyl-tRNA synthetase		
Anti-EJ	Glycyl-tRNA synthetase		
Anti-KS	Asparaginyl-tRNA synthetase		
Anti-Ha	Tyrosyl-tRNA synthetase		
Anti-Zo	Phenylalanyl-tRNA synthetase		
Anti-Mi-2	Nuclear helicase	Classic cutaneous features Milder muscle disease Better response to therapy	5–15%
Anti-SRP	Signal recognition particle	Polymyositis Severe muscle involvement Necrotizing myopathy	<1%
Anti-TIF1-γ (anti-p155/140)	Transcriptional intermediary factor 1-γ (TIF1-γ, ectodermin)	Severe cutaneous disease Malignancy in adults but not children	20–30%
Anti-MJ (anti-p140)	Nuclear matrix protein (NXP-2)	Subcutaneous calcinosis	27%
Anti-MDA5 (anti-CADM-140)	RNA helicase encoded by melanoma differentiation-associated gene 5 (MDA5)	Rapidly progressive interstitial lung disease in Asian adults	Not reported

Table. Clinical Associations: MSAs and MAAs in juvenile-onset myositis

Autoantibodies	Target autoantigen	Prevalence in patients with juvenile-onset myositis	Clinical associations
Common MSAs are found in 45%-55% of patients with juvenile-onset myositis			
Anti-Mi2	NuRD complex	3%-4%[39,40]	'Classic' dermatomyositis. Responds well to standard therapies. Favorable prognosis[5,46]
Anti-TIF 1g (p155/140, TRIM33)	TIF 1-γ	18%-35%[39,40]	Severe cutaneous disease. Rashes in photo-exposed pattern. Chronic disease course. Lipodystrophy[41-45]
Anti-NXP2 (p140, MJ)	NXP 2	15%-22%[39,40]	Calcinosis. More severe muscle disease. Gastrointestinal bleeding, ulcers and dysphagia. Worse disease outcome and functional status[42,44,46]
Anti-MDA5 (CADM-140)	MDA 5	6%[39]	More common in east Asia where associated with clinically amyopathic myositis, rapidly progressive interstitial lung disease and a high mortality. In Caucasian populations associated with mild muscle disease, interstitial lung disease, arthritis, and ulceration[48-50]
Rare but clinically important MSAs are found in 5%-8% of patients with juvenile-onset myositis			
Antisynthetases			Antisynthetase syndrome: myositis, interstitial lung disease, fever, mechanics hands, Raynaud's phenomenon and arthritis; occurs in older children. Increased mortality[42]
- Jo-1	- Histidyl	2%-3%[39,40]	
- PL12	- Alanyl	2%-3%[39,40]	
- PL7	- Theronyl	2%-3%[39,40]	
- OJ	- Isoleucyl	2%-3%[39,40]	
- EJ	- Glycyl	2%-3%[39,40]	
- KS	- Asparaginyl	2%-3%[39,40]	
- Zo	- Phenylalanyl	2%-3%[39,40]	
- Ha	- Tyrosyl	2%-3%[39,40]	
Anti-SRP	SRP	2%[39,40]	Necrotizing autoimmune myositis. Severe weakness. Cardiac involvement. Occurs in older children. May be refractory to standard treatment[42]
Anti-HMGCR	HMGCR	1%[39]	Necrotizing autoimmune myositis[52,53]
Anti-SAE	SAE	1%[39]	Initially amyopathic disease with muscle involvement occurring later
MAAs are found in 16%-20% of patients with juvenile-onset myositis. Some may occur in conjunction with a myositis-specific autoantibody			
Anti-PmScl	Exsome-associated PM- Scl-75; PM-Scl-100; C1D[55]	5%[39]	Overlap syndromes[42]
Anti-U1RNP	U1RNP[56]	2%[40]	Overlap syndromes[42]
Anti-Ro52	Ro52[57]	5%[40]	Overlap syndromes. May be found in conjunction with other MSA, particularly antisynthetases[42]

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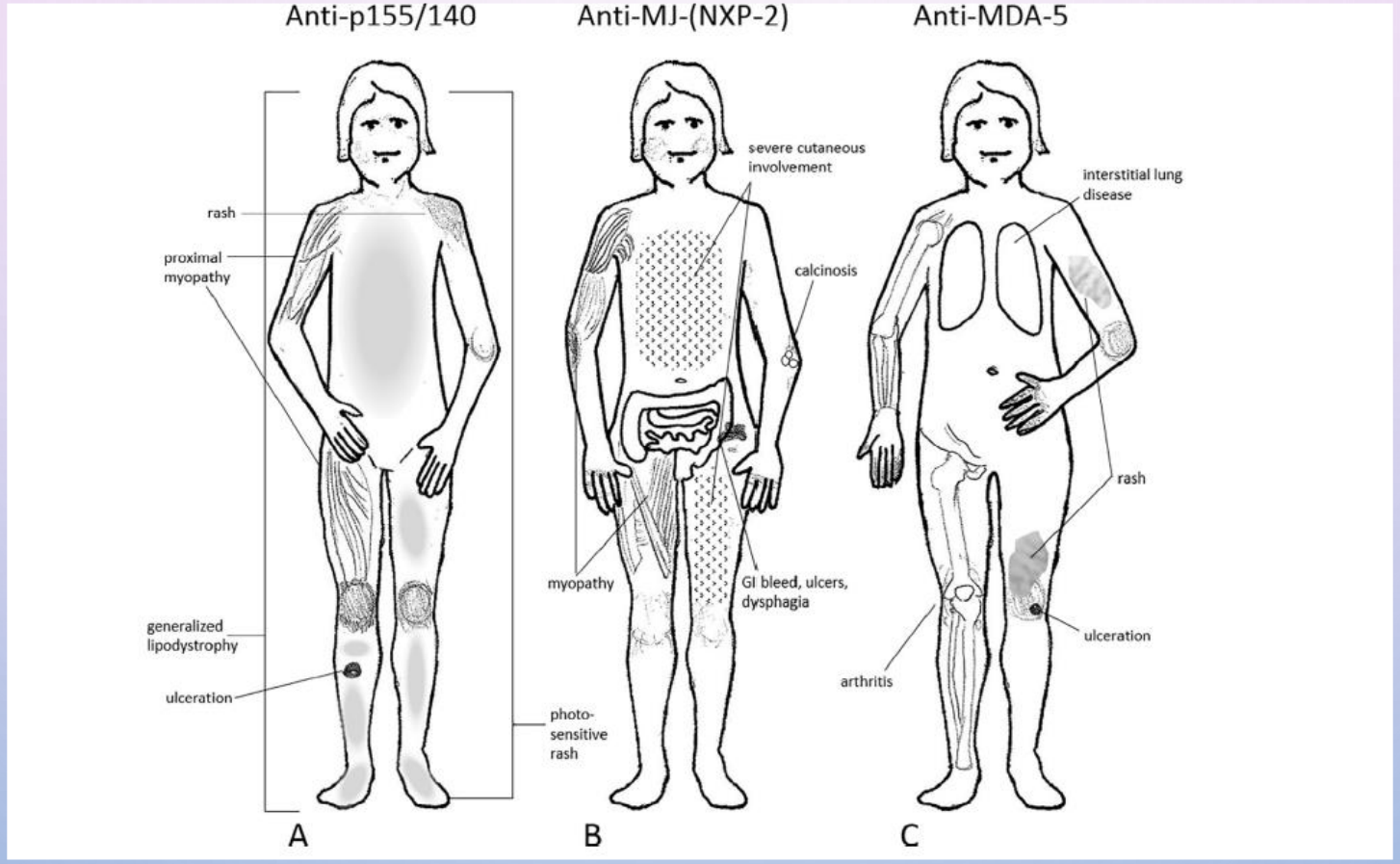
Common Myositis-Specific Antibodies in Juvenile Dermatomyositis



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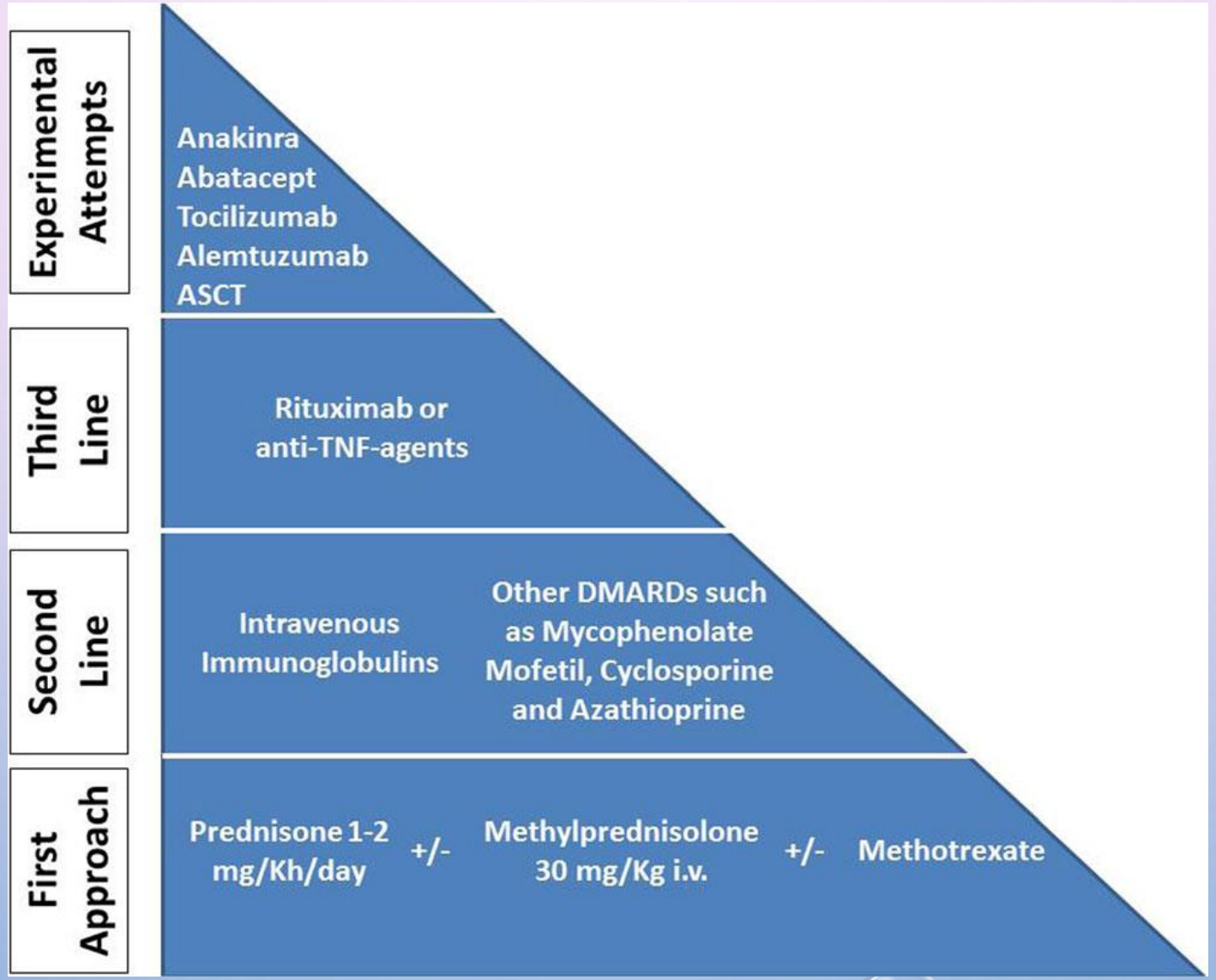


different treatment lines from the initial JIIM

Ilaria Pagnini
Clinic Rev Allerg Immunol DOI 10.1007/s12016-015-8512-9

Table 3 Therapy in patients with juvenile dermatomyositis (n = 57) at diagnosis and throughout disease course

	At diagnosis (%)	Throughout course (%)
Oral steroids	48 (84)	53 (93)
High dose pulse intravenous steroids	42 (74)	47 (82)
Methotrexate	27 (47)	36 (63)
Intravenous immunoglobulin	4 (7)	18 (32)
Cyclosporine	2 (4)	10 (18)
Cyclophosphamide	0 (0)	1 (2)

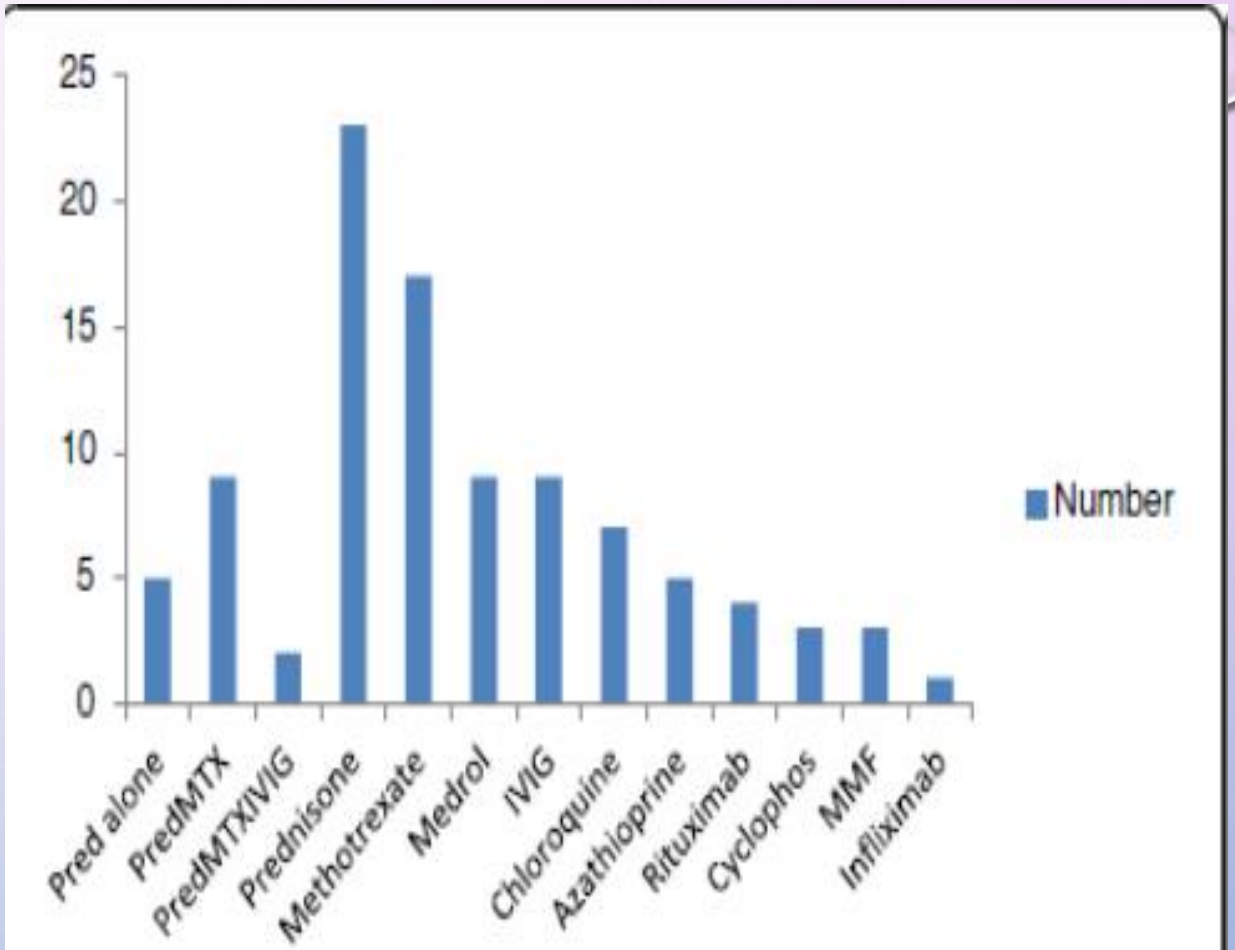


Fig. 3 Drugs used in management of Cape Town JDM patients. Abbreviations: Cyclophos: cyclophosphamide; Medrol: intravenous methylprednisolone; Pred: Prednisone PredMTX: Prednisone and methotrexate; IVIG: Intravenous immunoglobulin

Formas de seguimiento y estratificación de gravedad UK 2018
van Dijkhuizen et al. Arthritis Research & Therapy (2018) 20:180

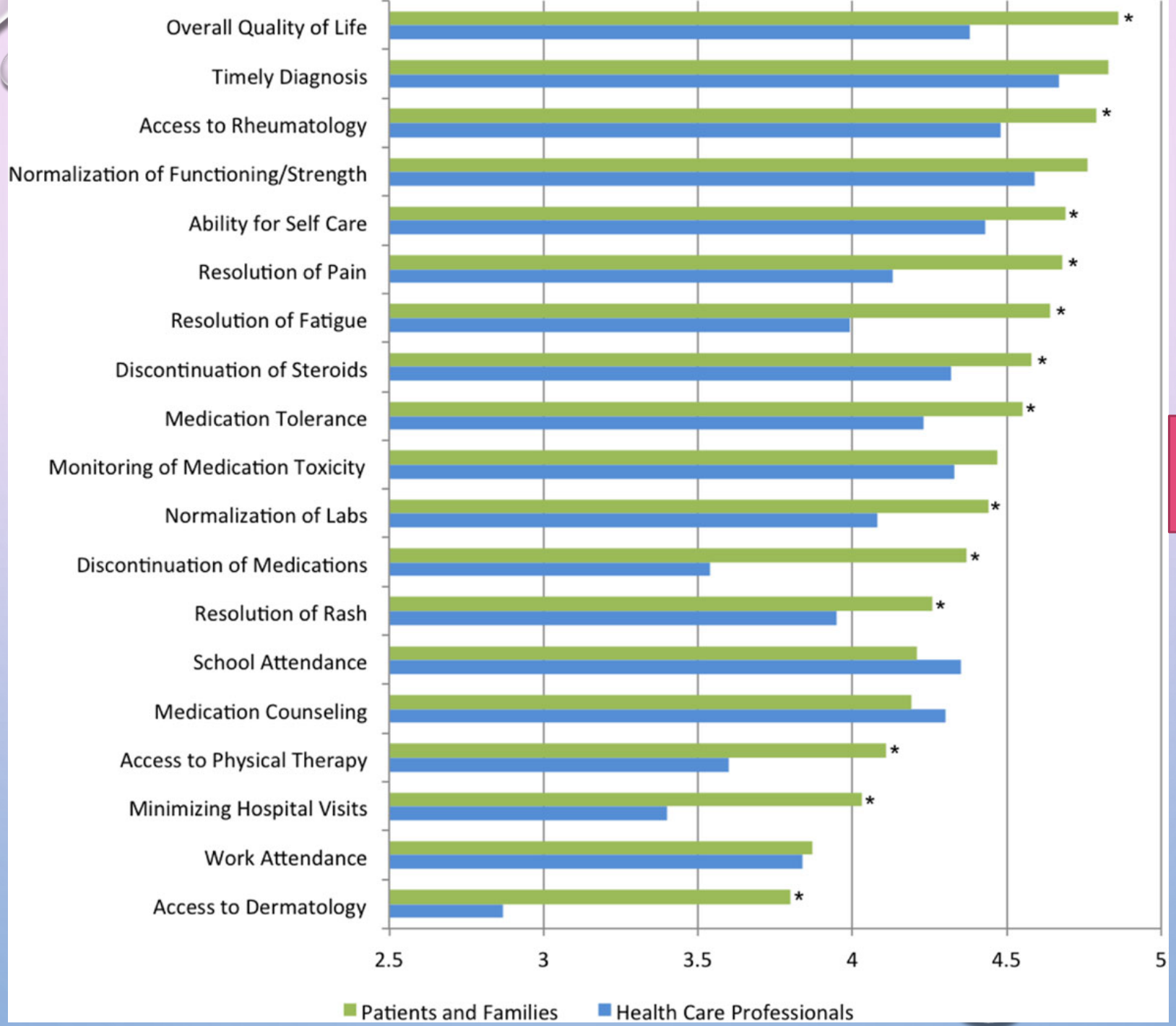
Table 1 Baseline table

Parameter	Included *N* = 340	Excluded *N* = 73
Female, *n* (%)	236 (69.4)	54 (74.0)
Age at diagnosis, years	7.4 (4.5, 10.5)	7.3 (4.1, 11.1)
Disease duration at diagnosis, years	0.3 (0.2, 0.6)	0.3 (0.2, 1.0)
Time after diagnosis at enrollment, years	0.2 (0.1, 1.1)	2.3 (0.4, 5.4)
Duration of follow up, years	4.1 (1.6, 7.1)	1.2 (0.1, 2.6)
Disease activity at enrollment:		
CK, U/L	103 (64, 440)	98 (45, 256)
CMAS, points	41 (21, 50)	46 (37, 52)
MMT8, points	65 (45, 80)	80 (64, 80)
PGA, cm	3 (1.3, 6.0)	2.3 (0.5, 4.0)

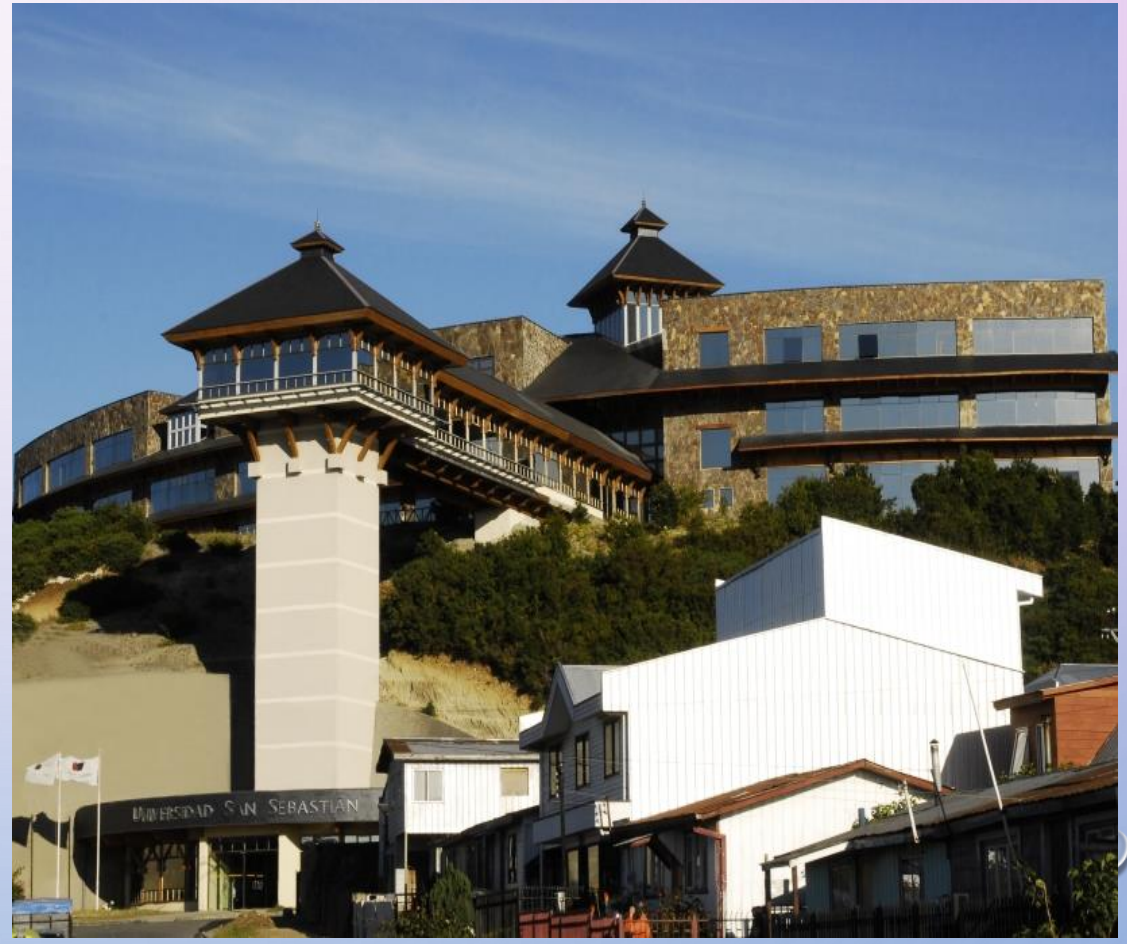
Table 2 JDM disease activity core set and disease damage core set

JDM disease activity core set		
	Item(s) used to measure the domain	
Domain	PRINTO/PRCSG [49]	IMACS [50]
Global assessment by physician	VAS or Likert scale	VAS or Likert scale
Muscle strength assessment	CMAS and MMT	MMT
Laboratory assessment: muscle enzymes	Creatine phosphokinase, LDH, aldolase, AST/ALT	At least 2 of the following: creatine phosphokinase, LDH, aldolase, AST/ALT
Functional ability assessment	CHAQ	CHAQ or CMAS
Global assessment by parents/patients	VAS or Likert scale	VAS or Likert scale
Global JDM disease activity tool	DAS and MDAA	–
JDM disease damage core set [49]		
Domain	Item(s) used to measure the domain	
Global assessment by physician	VAS or Likert scale	
Functional ability assessment	CHAQ	
Growth and development	Height and weight	
	Menses	
	Tanner puberty stage	
Global JDM damage tool	MDI	
Muscle strength assessment	CMAS	

ALT alanine aminotransferase; *AST* aspartate aminotransferase; *CHAQ* Childhood Health Assessment Questionnaire; *CMAS* Childhood Myositis Assessment Scale; *DAS* Disease Activity Score; *IMACS* International Myositis Assessment and Clinical Studies group; *JDM* juvenile dermatomyositis; *LDH* lactate dehydrogenase; *MDAA* Myositis Disease Activity Assessment; *MDI* Myositis Damage Index; *MMT* Manual Muscle Testing; *PRCSG* Pediatric Rheumatology Collaborative Study Group; *PRINTO* Pediatric International Trials Organization; *VAS* visual analogue scale



Tory et al. Pediatric Rheumatology (2018) 16:28



Gracias por su atención