Rheum for All: A Medley of Rheumatology Cases

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UMKC School of Medicine

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Case 1: 16-year-old female with:

- CC: "bilateral hand pain"
- HPI:
  - 1-month history pain in bilateral hand with stiffness
  - Started after a new weightlifting class at school
  - No swelling, no fever, rashes, abdominal pain
- PMx/FHx/SHx:
  - Obesity, Hypertension, Hashimoto thyroiditis (on Synthroid)
  - No notable family history
  - Lives with her brother, parents live in Mexico

Case 1: 16-year-old female with:

- PE:
  - EENT appropriate, with oral ulcer
  - Heart and pulses appropriate
  - Lungs appropriate
  - Limitation of MCPs (2-4) bilaterally, all other joints appropriate
  - No rashes

Thoughts? Differential?

Labs

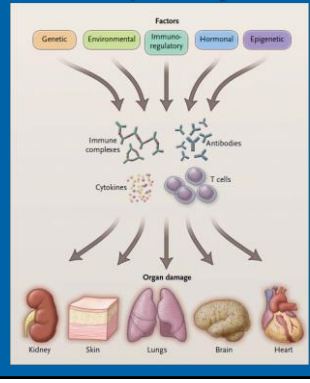
4.4	10.0 / 29.4	228
139 / 4.0	108 / 26	15 / 0.71
		76

AST: 11
ALT: 8
Alb: 3.1

ANA: > 1280
DsDNA: >1280
Smith: positive
RNP: positive
C3: 32
C4: < 8

Urinalysis
3+ protein
2+ blood
2-10 WBC
20-50 RBC
Pro/Cre ratio 1.0

Systemic Lupus Erythematosus



Tsokos G. N Engl J Med. 2011

Systemic Lupus Erythematosus (SLE)

- 20% of all lupus diagnosed prior to 18 years
- Female > Male (4:1), higher in adults (9:1)
- Rare before age 5
- Anti-dsDNA and anti-Smith most seen
- Renal disease common and affects morbidity
- CNS involved in 10-30%
- Skin – Malar, discoid, alopecia, photosensitivity
- Arthritis, Serositis, cytopenias

Harry O, et al. J Pediatr. 2018

SLE Criteria

Entry criterion
Antinuclear antibodies (ANA) at a titer of ≥1:80 on HEp-2 cells or an equivalent positive test (ever)

↓

If absent, do not classify as SLE
If present, apply additive criteria

↓

Additive criteria
Do not count a criterion if there is a more likely explanation than SLE.
Occurrence of a criterion on at least one occasion is sufficient.
SLE classification requires at least one clinical criterion and ≥10 points.
Criteria need not occur simultaneously.
Within each domain, only the highest weighted criterion is counted toward the total score§.

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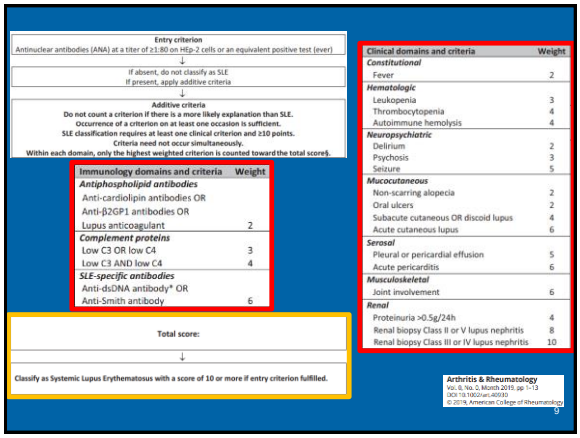

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Criteria need not occur simultaneously.
Within each domain, only the highest weighted criterion is counted toward the total score§.

Immunology domains and criteria	Weight
Antiphospholipid antibodies	
Anti-cardiolipin antibodies OR Anti-β2GP1 antibodies OR Lupus anticoagulant	2
Complement proteins	
Low C3 OR low C4	3
Low C3 AND low C4	4
SLE-specific antibodies	
Anti-dsDNA antibody* OR Anti-Smith antibody	6

Total score:

↓

Classify as Systemic Lupus Erythematosus with a score of 10 or more if entry criterion fulfilled.

Clinical domains and criteria	Weight
Constitutional	
Fever	2
Hematologic	
Leukopenia	3
Thrombocytopenia	4
Autoimmune hemolysis	4
Neuropsychiatric	
Delirium	2
Psychosis	3
Seizure	5
Mucocutaneous	
Non-scarring alopecia	2
Oral ulcers	2
Subacute cutaneous OR discoid lupus	4
Acute cutaneous lupus	6
Serosal	
Pleural or pericardial effusion	5
Acute pericarditis	6
Musculoskeletal	
Joint involvement	6
Renal	
Proteinuria >0.5g/24h	4
Renal biopsy Class II or V lupus nephritis	8
Renal biopsy Class III or IV lupus nephritis	10

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Hematologic Disorder

- Leukopenia
  - (< 4,000 K/ml)
- Thrombocytopenia
  - (<100,000 K/ml in the absence of offending drug)
- Hemolytic anemia
  - (low haptoglobin, elevated LDH, positive Coomb's)

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Renal biopsy Class III or IV lupus nephritis	10

10

Mucocutaneous Acute cutaneous lupus

Mucocutaneous	
Non-scarring alopecia	2
Oral ulcers	2
Subacute cutaneous OR discoid lupus	4
Acute cutaneous lupus	6

- Acute cutaneous lupus
- Rash on cheeks and nose, butterfly shape
- Flat or raised erythema
- Spares nasolabial folds
- Photosensitive
- No scarring
- Hyper-/hypo pigmentation with resolution

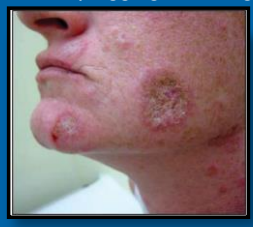
Walling et al.Am J Clin Dermatol.2009
Kuhn et al.J Autoimmun.2014

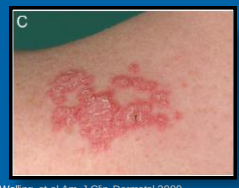
Oral Ulcers

Hard palate involvement is classic: hyperemia, petechiae, or ulceration, often painless

Discoid lupus

- Occurs in 20% of SLE patients; 5-10% develop SLE
- Well demarcated, discoid plaque, peripheral scale, central hypopigmentation/atrophic leads to scarring, follicle plugging, scarring alopecia

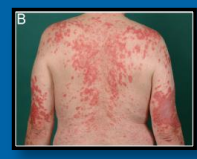


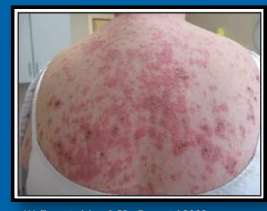


Walling et al.Am J Clin Dermatol.2009
Kuhn et al.J Autoimmun.2014
Okon et al. Best Pract & Res Clin Rheum. 2013
Rothfield et al. Clin Dermatol.2006

Subacute cutaneous lupus

- Photosensitive, no scarring, pigmentation changes with resolution
- Scaly annular and papulosquamous plaques
- Rare below waist





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Walling et al.Am J Clin Dermatol.2009
Kuhn et al.J Autoimmun.2014
Okon et al. Best Pract & Res Clin Rheum. 2013
Sontheimer et al. Arch Dermatol Res.2009

Renal

Renal	
Proteinuria >0.5g/24h	4
Renal biopsy Class II or V lupus nephritis	8
Renal biopsy Class III or IV lupus nephritis	10

- Lupus nephritis is important predictor of long-term survival
- More common in African and Asian
- Diagnose with kidney biopsy
- Random Protein/Creatinine ratio (normal < 0.2)

Class I	Minimal mesangial lupus nephritis
Class II	Mesangial proliferative lupus nephritis
Class III	Focal lupus nephritis†
Class IV	Diffuse segmental (IV-S) or global (IV-G) lupus nephritis‡
Class V	Membranous lupus nephritis§
Class VI	Advanced sclerosing lupus nephritis

Harry O, et al. J Pediatr. 2018
Weening J, et al. J Am Soc Nephrol and Kidney Int. 2004

Lupus Approved Medications

1. Hydroxychloroquine (Plaquenil)
2. Corticosteroids
3. Aspirin
4. Belimumab (Benlysta)- first new drug approved in 50 years.
   - Antibody against BAFF/BLyS (B-cell activating factor/ B-lymphocyte stimulator)

Also a lot of medications used off-label

Case 2: 3-year-old female with:

CC: Fever

HPI:

- Fever with rash x 14 days
- Multiple limited swollen joints

PE:

- Diffuse rash
- Lymphadenopathy
- Hepatosplenomegaly
- Polyarthritis

Labs

- Anemia, thrombocytosis, elevated ESR/CRP, elevated ferritin

Thoughts? Differential?

Labs

10.3	10.1	647
	34.6	

140	97	10
4.2	26	0.25

97

EBV: negative
Hepatitis A, B, C: negative
CMV: negative
Ehrlichia: Negative
Parvovirus: negative
Rickettsia: negative
RVP: negative

AST: 37
ALT: 37
Alb: 3.8
LDH: 1025
CRP: 5.6
ESR: 53

ANA: negative
ASO: negative
Ferritin: 2430

Systemic Onset Juvenile Idiopathic arthritis (sJIA)

Arthritis in any number of joints together with a fever of at least 2 weeks' duration that is documented to be daily (quotidian) for at least 3 days and is accompanied by one or more of the following:

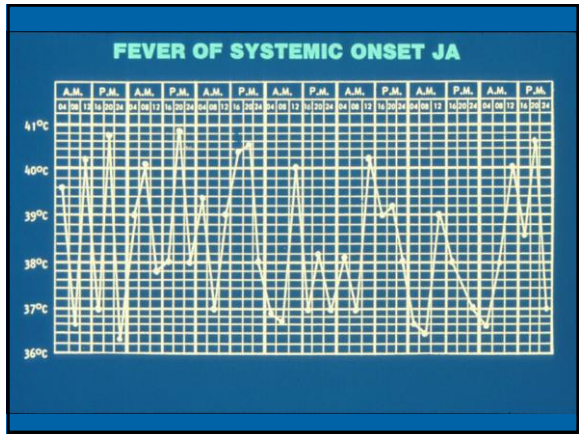
- Evanescent rash
- Generalized lymphadenopathy
- Enlargement of liver or spleen
- Serositis

Exclusions:

- Psoriasis or a history of psoriasis in the patient or a first-degree relative
- Arthritis in an HLA-B27–positive male beginning after his sixth birthday
- Ankylosing spondylitis, enthesitis-related arthritis, sacroiliitis with inflammatory bowel disease, reactive arthritis, or acute anterior uveitis, or a history of one of these disorders in a first-degree relative
- The presence of IgM RF on at least two occasions at least 3 months apart

HLA, Human leukocyte antigen; *IgM*, immunoglobulin M.

Petty et al. Text of Pediatric Rheumatology. 7th ed.



Systemic Onset JIA (sJIA)

- **5-15% of total # JIA patients**
- **High, intermittent fever (Quotidian)**
- **Characteristic rash 80%**
- **Male = Female**
- **Onset commonly before 5 years**
- **Arthritis may lag behind systemic features**
  - **(months to years)**
- **ANA, RF negative**
- **Anemia, leukocytosis, thrombocytosis**
- **Absent eye disease**
- **Macrophage activation syndrome**

Petty et al. Text of Pediatric Rheumatology. 7th ed.
Behrens E. et al. J Rheumatol. 2008

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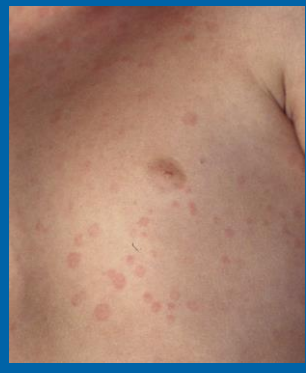

Rash:
Maculopapular
Nonpruritic
Urticarial-like

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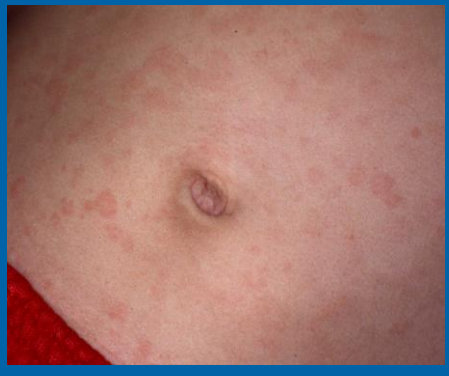

sJIA Treatment

- NSAIDs (indomethacin)
- Steroids (oral and IV)
- Anakinra – IL-1 antagonist
- Canakinumab – IL-1β antagonist
- Tocilizumab – IL-6 antagonist
- Cyclosporine
- Tofacitinib

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Remember…
Juvenile Idiopathic Arthritis

- *Umbrella* diagnosis heterogeneous group of chronic arthridities
- Diagnosis of exclusion
- Diagnosis: historical/objective findings
- Laboratory tests: classification, exclusion, prognosis, **BUT** do not establish the diagnosis

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Case 2.5:
3-year-old female with sJIA:

CC: Fever

HPI:

- Discharge home on anakinra
- 2 weeks later developed fever
- Persistent, high fever x 5 days
- No swollen joints or rash

PE:

- Lymphadenopathy
- Hepatosplenomegaly
- Polyarthritis, improved, but present

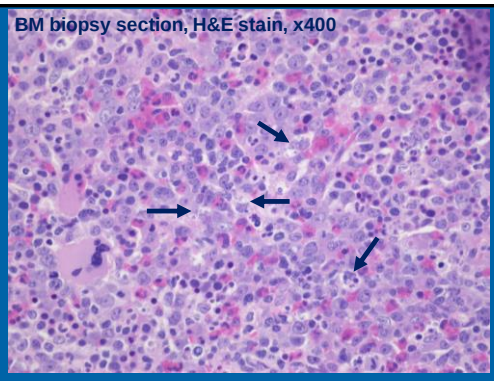
Labs

22.9	7.0 / 24.6	212

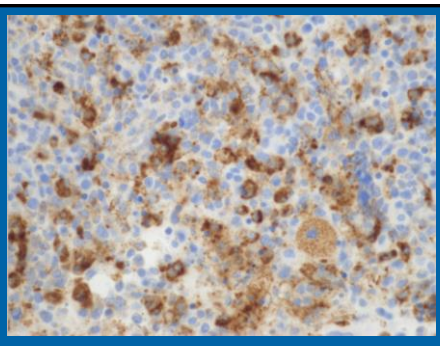
132	97	10	
4.2	26	0.25	97

EBV: negative	AST: 177	ANA: negative
Hepatitis A, B, C: negative	ALT: 37	ASO: negative
CMV: negative	Alb: 2.7	sIL-2r: 9618
Ehrlichia: negative	LDH: 4977	Triglycerides: 182
Parvovirus: negative	CRP: 7.8	Ferritin: 23697
Rickettsia: negative	ESR: 36	D-Dimer: > 20
RVP: negative		

BM biopsy section, H&E stain, x400



Hypercellular marrow, increased histiocytes which show hemophagocytosis (arrows)

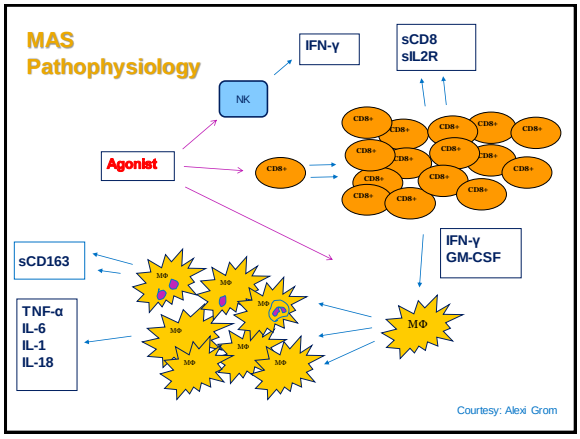


- CD68 IHC stain, x400
- Hightlights increased histiocytes and hemophagocytosis

Macrophage Activation Syndrome (MAS)

- Caused by excessive activation & proliferation of T cells and macrophages (exaggerated immune response)
- Macrophages exhibit hemophagocytic activity
- Predominately histiocytes and T cells (CD8+)
- Clinical signs due to "cytokine storm"
  - T cells: IFN-γ, IL-2, GM-CSF
  - Macrophage: IL-6, IL-1, TNF-α, IL-18
- Can quickly become fatal
  - Mortality up to 30%

Silverman et al. *J Pediatr*. 1983
Hadchouel et al. *J Pediatr*. 1985
Billiau, et al. *Blood*. 2005



MAS Pathophysiology

- Defect in cytotoxic cell function
  - **Normal** = cytotoxic cells induce apoptosis of immune cells at conclusion of immune response
  - **Abnormal** = cytotoxic cells that fail to provide appropriate apoptotic signals for removal of immune cells
    - Leads to overabundance and excessive activation

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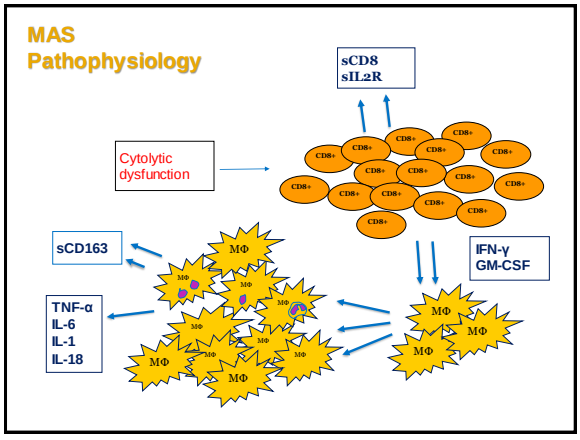
Verbsky et al. *Ann Med.* 2006



MAS Clinical Findings

- Acutely ill with persistent fever
- Hepatosplenomegaly
- Hemorrhagic rash
  - Bruising, purpura, mucosal bleeding
- Enlarged lymph nodes
- CNS dysfunction
  - Mental status changes
  - Seizures

MAS Laboratory Findings

- Cytopenias
  - Sharpe fall in
    - WBC (neutropenia)
    - Hemoglobin
    - Platelets (early sign)
    - ESR (despite high CRP)
- Elevated ferritin
- Elevated sIL-2R
- Elevated sCD163
- Coagulopathy
  - Elevated d-dimer
  - Alterations in clotting time
    - Increase PT, PTT
  - Decrease in fibrinogen

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And More Laboratory Findings

- Abnormal liver function tests
  - Elevated AST, ALT
  - Elevated bilirubin
  - Ammonia is normal
- Hypertriglyceridemia
  - Due to elevated TNF-α (decrease lipoprotein lipases)
- Hypoalbuminemia
- Hyponatremia

Classification Criteria for MAS (in sJIA)

- A febrile patient with known or suspected sJIA is classified as having MAS if:

Ferritin > 684 ng/ml

- *And* any 2 of the following:

Platelet count ≤ 181 x10⁹/liter

AST > 48 units/liter

Triglycerides > 156 mg/dl

Fibrinogen ≤ 360 mg/dl

Ravelli, et al. *Arthritis Rheumatol*. 2016

Treatment Options

- High dose steroids
  - Methylprednisolone IV (30 mg/kg) daily x 3-5 days
  - Then IV (2 mg/kg) divided 2-3 times daily
- Cyclosporine (CyA)
  - PO or IV (2-5 mg/kg) divided twice daily
- Etoposide
- Biologics (anakinra)
- Rituximab

Case 3: 17-month-old Hispanic female with fever

HPI:

- Abdominal pain and diarrhea x 4 days, then resolved
- Fever persisted
- Poor oral intake
- No joint complaints
- No cough, congestion, runny nose
- Father ill with fever, headache, myalgias

PE:

- Fussy
- Eczematous rash antecubital and popliteal
- No other PE abnormalities

Thoughts? Differential?

Labs

1.4	10.3 / 29.6	14
135 / 3.2	108 / 22	3 / 0.23
		84

AST 447
ALT 169
ESR 10
CRP 13.6
LDH 4204
Ferritin 2291
D-dimer 14.12

Stool studies: negative
Rickettsia: negative
Ehrlichia: negative
Enterovirus: negative
Adenovirus: positive
SARS-CoV-2: positive
SARS-CoV antibody: negative

BNP: 828
Triglycerides: 363
ANA: negative
Uric Acid: 2.9

SARS-CoV-2 and Multisystem Inflammatory Syndrome in Children (MIS-C)

(1) An individual aged < 21 years with:
(2) Clinical criteria:
- A minimum 24-h history of subjective or objective fever ≥ 38.0 °C AND
- Severe illness necessitating hospitalization AND
- Two or more organ systems affected (i.e., cardiac, renal, respiratory, hematologic, gastrointestinal, dermatologic, neurological)

(3) Laboratory evidence of inflammation
- One or more of the following: an elevated CRP, ESR, fibrinogen, procalcitonin, D-dimer, ferritin, LDH, or IL-6; elevated neutrophils or reduced lymphocytes; low albumin

(4) Laboratory or epidemiologic evidence of SARS-CoV-2 infection
- Positive SARS-CoV-2 testing by RT-PCR, serology, or antigen OR
- COVID-19 exposure within 4 weeks prior to onset of symptoms

(5) No alternative diagnosis

CDC Health Alert Network. Multisystem Inflammatory Syndrome in Children (MIS-C) Associated with Coronavirus Disease 2019 (COVID-19). Available online: https://emergency.cdc.gov/han/2020/han00432.asp

SARS-CoV-2 and Multisystem Inflammatory Syndrome in Children (MIS-C)

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CDC Health Alert Network. Multisystem Inflammatory Syndrome in Children (MIS-C) Associated with Coronavirus Disease 2019 (COVID-19). Available online: https://emergency.cdc.gov/han/2020/han00432.asp

Additional Evaluation

- Chest x-ray: normal
- Echocardiogram:
  - moderate pericardial effusion
  - Normal coronary vessels
- Bone marrow biopsy – pancytopenia
- Peripheral smear – no blasts

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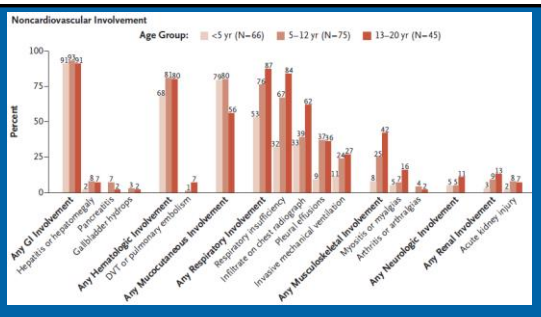
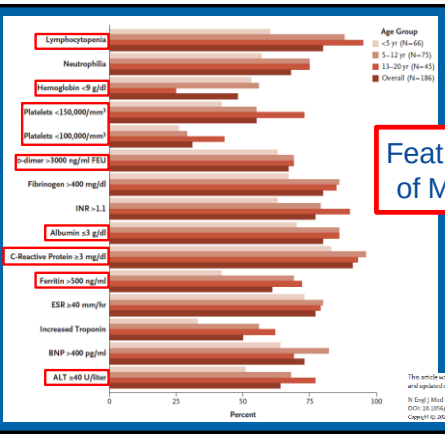
Demographics Patients with SARS-CoV-2 Infection

Characteristic	Laboratory Confirmation of SARS-CoV-2 Infection (N=131)	
	RT-PCR Positive (N=73)†	Antibody Test Positive, RT-PCR Negative or Unknown (N=58)
Male sex — no. (%)	43 (59)	36 (62)
Median age (interquartile range) — yr	9.1 (4.8–14.2)	9.1 (4.1–11.7)
Age group — no. (%)		
<1 yr	6 (8)	0
1–4 yr	13 (18)	19 (33)
5–9 yr	21 (29)	14 (24)
10–14 yr	17 (23)	18 (31)
15–20 yr	16 (22)	7 (12)
Race and ethnic group — no. (%)‡		
White, non-Hispanic	13 (18)	8 (14)
Black, non-Hispanic	17 (23)	18 (31)
Hispanic or Latino	29 (40)	12 (21)
Other race, non-Hispanic	4 (5)	1 (2)
Unknown	11 (15)	19 (33)
Underlying conditions		
Previously healthy — no. (%)§	49 (67)	43 (74)
At least one underlying condition, excluding obesity — no. (%)	24 (33)	15 (26)



- 80% with Cardiac involvement
  - Elevated troponin, BNP, pericarditis, coronary change



Features of MAS

Treatment

Table 2. Clinical Characteristics of the Patients According to the Number of Kawasaki's Disease–like Features Present.*

Characteristic	Patients with 4 or 5 Features (N=38)	Patients with 2 or 3 Features plus Laboratory Findings (N=36)	Other (N=112)†	All Patients (N=186)
Intravenous immune globulin — no. (%)	38 (100)	35 (97)	71 (63)	144 (77)
Median day of illness on which treatment was received (IQR)	6 (6–8)	7 (6–8)	6 (5–8)	6 (5–8)
Second dose received — no. (%)	16 (42)	9 (25)	14 (12)	39 (21)
Systemic glucocorticoid — no. (%)	20 (53)	18 (50)	53 (47)	91 (49)
Interleukin-6 inhibitor — no. (%)‡	1 (3)	1 (3)	12 (11)	14 (8)
Interleukin-1Ra inhibitor — no. (%)**	5 (13)	6 (17)	13 (12)	24 (13)
Anticoagulation therapy — no. (%)††	14 (37)	18 (50)	55 (49)	87 (47)

- After treatment initiation**
  - Laboratory findings are slow to improve
  - Clinical improvement before laboratory improvement

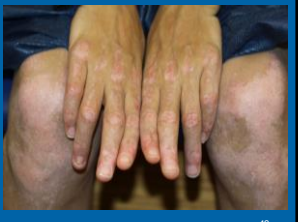
Case 4:
8-year-old female with fatigue and rash

HPI:

- Fatigue for 4 months, can't keep up with others
- Difficulty getting dressed
- Nonpainful/Nonpruritic rash
- Daily cough

PE:

- Rash on hands, face, knees
- Diffuse weakness
  - Shoulders, hips, core worst
- Arthritis: MCPs, knees



Thoughts?
Differential?

Labs

8.4	11.3 / 34.4	418

139	108	15
4.0	26	0.71

76

AST 564

ALT 233

ANA > 1280

DsDNA negative

RNP & Smith negative

C3 102

C4 41

ESR 25

CRP 1.5

CK 13,870

Aldolase 29.2 (nl < 14.5)

Juvenile dermatomyositis

- Females > Males
- Peak incidence 3-7 years, then again in early teenage years
- Rash
  - Gottron papules – small hand joints, knee, elbows
  - Malar rash
  - Shawl sign
  - Heliotrope rash
- Proximal muscle weakness
- GI tract dysfunction – dysphagia, dysphonia
- Arthritis

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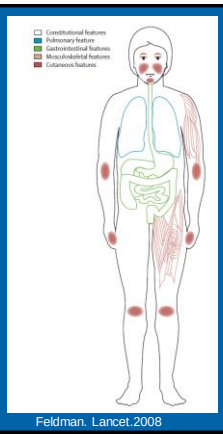

Feldman. Lancet.2008

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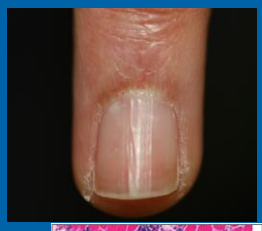

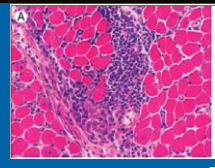

Feldman. Lancet.2008

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Table 2 The European League Against Rheumatism/American College of Rheumatology (EULAR/ACR) classification criteria for adult and juvenile idiopathic inflammatory myopathies (IIMs) 2017

When no better explanation for the symptoms and signs exists, these classification criteria can be used

Variable	Score points	
	Without muscle biopsy	With muscle biopsy
Age of onset		
Age of onset of first symptom assumed to be related to the disease ≥18 years and <40 years	1.3	1.5
Age of onset of first symptom assumed to be related to the disease ≥40 years	2.1	2.2
Muscle weakness		
Objective symmetric weakness, usually progressive, of the proximal upper extremities	0.7	0.7
Objective symmetric weakness, usually progressive, of the proximal lower extremities	0.8	0.5
Neck flexors are relatively weaker than neck extensors	1.9	1.6
In the legs, proximal muscles are relatively weaker than distal muscles	0.9	1.2

Probable:
≥ 5.5 without biopsy
≥ 6.7 with biopsy

Definite:
≥ 7.5 without biopsy
≥ 8.7 with biopsy

To cite: Lundberg IE, Tjärnlund A, Bottai M, *et al*. *Ann Rheum Dis* Published Online First: [*please include* Day Month Year]. doi:10.1136/annrheumdis-2017-211468

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Table 2 The European League Against Rheumatism/American College of Rheumatology (EULAR/ACR) classification criteria for adult and juvenile idiopathic inflammatory myopathies (IIMs) 2017

When no better explanation for the symptoms and signs exists, these classification criteria can be used

Variable	Score points	
	Without muscle biopsy	With muscle biopsy
Skin manifestations		
Heliotrope rash	3.1	3.2
Gottron's papules	2.1	2.7
Gottron's sign	3.3	3.7
Other clinical manifestations		
Dysphagia or oesophageal dysmotility	0.7	0.6

Probable:
≥ 5.5 without biopsy
≥ 6.7 with biopsy

Definite:
≥ 7.5 without biopsy
≥ 8.7 with biopsy

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Table 2 The European League Against Rheumatism/American College of Rheumatology (EULAR/ACR) classification criteria for adult and juvenile idiopathic inflammatory myopathies (IIMs) 2017

When no better explanation for the symptoms and signs exists, these classification criteria can be used

Variable	Score points	
	Without muscle biopsy	With muscle biopsy
Laboratory measurements		
Anti-Jo-1 (anti-histidyl-tRNA synthetase) autoantibody present	3.9	3.8
Elevated serum levels of creatine kinase (CK)* *or* lactate dehydrogenase (LD)* *or* aspartate aminotransferase (ASAT/AST/SGOT)* *or* alanine aminotransferase (ALAT/ALT/SGPT)*	1.3	1.4
Muscle biopsy features—presence of:		
Endomysial infiltration of mononuclear cells surrounding, but not invading, myofibres		1.7
Perimysial and/or perivascular infiltration of mononuclear cells		1.2
Perifascicular atrophy		1.9
Rimmed vacuoles		3.1

Probable:
≥ 5.5 without biopsy
≥ 6.7 with biopsy

Definite:
≥ 7.5 without biopsy
≥ 8.7 with biopsy

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Treatment

Table 3. Initial treatment plans for the first 4 weeks

Intravenous methylprednisolone
30 mg/kg/day (maximum 1 gm) once a day for 3 days. May continue 1 × per week (optional)

Methotrexate
Subcutaneous unless only oral possible: lesser of 15 mg/m^2 or 1 mg/kg (maximum 40 mg) once weekly

Prednisone
2 mg/kg/day (maximum 60 mg) once daily × 4 weeks, then follow schedule in Table 4

Intravenous immunoglobulin
2 gm/kg (maximum 70 gm), every 2 weeks × 3, then monthly (optional intravenous methylprednisolone × 1 with each dose)

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Arthritis Care & Research
Vol. 64, No. 4, April 2012, pp 546–553
DOI 10.1002/acr.20695

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Questions??

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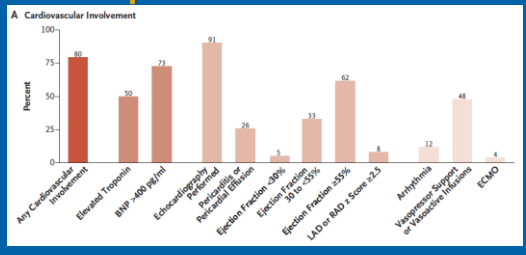
MAGNET

Back up



- 186 patients
  - 38 (20%) with 4/5 features of KD
  - 36 (19%) with 2/3 features of KD