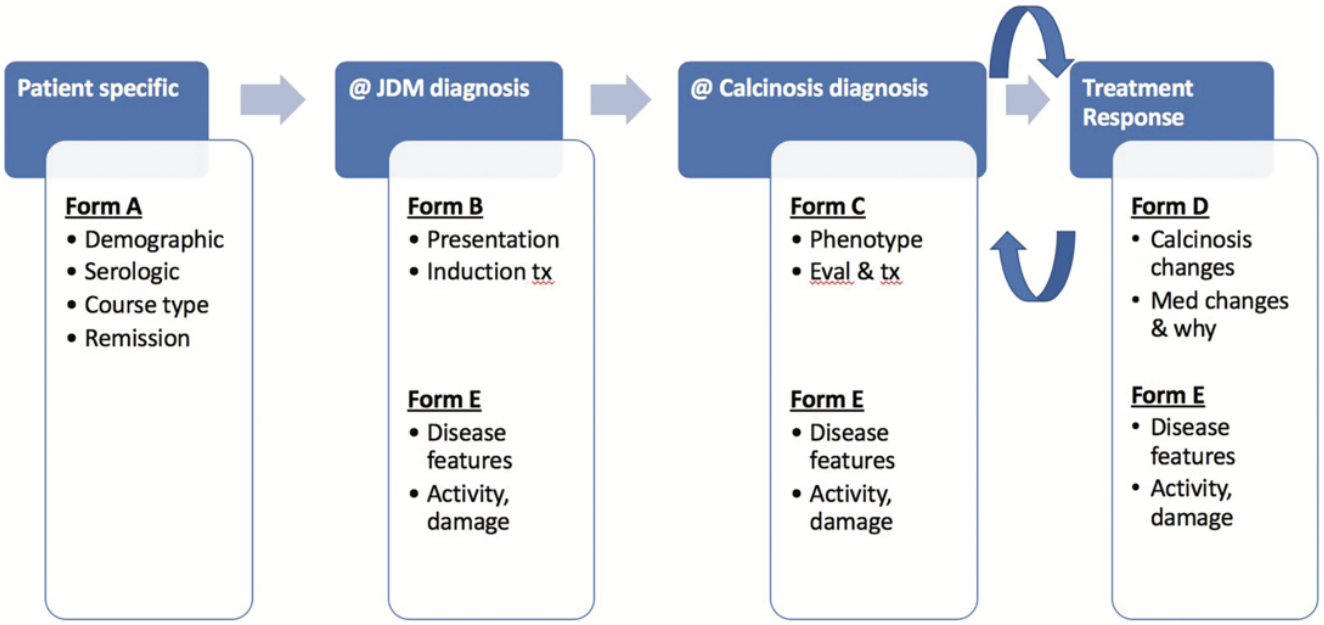
The following schematic represents the necessary data capture for each patient submitted. We recommend completing this form for each patient as you chart review, as it will correspond to the REDCap submission. We recommend using a new data collection form(s) for each unique patient as applicable to ensure accurate data is entered into the online REDCap database and to reference if there are any anomalies.

Please note the different clinical details obtained from each timepoint of the clinical course. The number of treatment iterations will determine how many times Form D will be completed.

Please note the use of the accompanying excel spreadsheet that will allow you to accurately report select dates/time intervals without sharing them with the research team.

Any questions, please contact Amir Orandi, Orandi.amir@mayo.edu



Form A: Patient Specific Details

Instructions: Answer the following demographical questions

1. Sex: M___ Female___
2. Race **(select all that apply)**
   a) Asian
   b) Black, African American, African or Afro-Caribbean
   c) Hispanic, Latino or Spanish origin
   d) Middle Eastern/North African
   e) Native American, American Indian or Alaskan Native
   f) Native Hawaiian or other Pacific Islander
   g) White
   h) Other _____
   i) Unknown
3. Date of initial JDM diagnosis: _____ (MM/DD/YYYY, enter in spreadsheet)
4. Date of most recent follow-up visit at your institution _____ (MM/DD/YYYY, enter in spreadsheet)
5. If deceased, date of death _____ (MM/DD/YYYY, enter in spreadsheet)
6. Height, and weight of the patient at time of JDM diagnosis: _____ cm _____ kg (enter in spreadsheet)
7. For each myositis specific or associated antibodies, select all that were **positive (+), negative (-), or not-tested (NT)** during any time of their disease course.

	+	-	NT		+	-	NT		+	-	NT		+	-	NT
Jo-1				SRP				U1RNP				P155/140 (TIF1g)			
PL7				HMGCR				U2RNP				MJ (NXP2)			
PL12				PM-ScL				U3RNP				caDM140 (MDA5)			
EJ				Ku				RNA-Pol				Mi-2			
OJ				Ro60				Th/To				Other			

8. Select the location of myositis antibody testing **(select all that apply)**
   a) Oklahoma Medical Research Foundation (OMRF)
   b) Other ____
   c) Unknown
9. For each antibody, select all that were **positive (+), negative (-), or not-tested (NT)** during any time of their disease course.

	+	-	NT		+	-	NT		+	-	NT		+	-	NT
ANA				SSB				Anti-dsDNA				Other			
RF (confirmed)				Anti-Sm				Anticardiolipin (confirmed)							
Anti-CCP				Anti-RNP				Beta-2-Glycoprotein I (confirmed)							
SSA				Anti-ScL 70				Lupus anticoagulant							

								(confirmed)									

Answer the following questions based on the patient's overall disease course:

1. Clinical treatment response:
   a) Did the patient ever achieve a *complete clinical response* (resolution of all clinical and biochemical signs of active JDM) for at least six months while <u>on</u> JDM medications? **(yes/no).**
      Date when complete clinical response was achieved: _____ (MM/DD/YYYY, enter in spreadsheet)
   b) Did the patient ever achieve *clinical remission* (resolution of all clinical and biochemical signs of active JDM) for at least six months while <u>off</u> all JDM medications? **(yes/no).**
      Date when remission achieved: _____ (MM/DD/YYYY, enter in spreadsheet)
2. Type of disease course – **select best fitting option.**
   a) Monocyclic: Achieved clinical remission (resolution of clinical and biochemical signs of disease) and off all medications 2 years from diagnosis.
   b) Polycyclic: Recurrence of disease activity after a definite period of clinical remission (definition above).
   c) Chronic continuous: Persistence of disease activity and/or medication use for 2 or more years following diagnosis.
   d) Not applicable due to total disease duration less than 2 years.

3. Was this patient's case ever published? If so, please include article information or PMID
   _____

Form B: At JDM Diagnosis

Instructions: Answer the following questions regarding the patient's initial diagnosis of JDM.

****Form E: JDM Disease activity, must also be completed for this time point****

1. Approximate date initial JDM symptoms were observed: _____ (MM/DD/YYYY, enter in spreadsheet)

2. Date that treatment for JDM was first initiated: _____ (MM/DD/YYYY, enter in spreadsheet)

3. Was calcinosis present at the time of initial JDM diagnosis? **(yes/no)**

4. Did the patient have amyopathic/hypomyopathic/skin predominant disease? **(yes/no)**
   [Defined as having no functional limitations or weakness based on history, strength assessment or CHAQ with muscle enzymes ≤ 1.2 times the upper limit of normal].

5. What was the 25-OH Vitamin D level at the time of diagnosis?
   - ☐ _____ ng/mL OR nmol/L _____
   - ☐ Not tested

6. Was Vitamin D and/or calcium supplementation prescribed **in the first 3 months** following diagnosis **(yes/no)**

7. What immunosuppressive treatment was prescribed **in the first 3 months** following diagnosis:
   - ☐ IV methylprednisolone **(yes/no)**
     If yes, was the maximal dose greater than 2 mg/kg/day? **(yes/no)**
     If yes, did the patient receive IV methylprednisolone at any point after the first 7 days of JDM treatment? **(yes/no)**

   - ☐ Oral prednisone **(yes/no)**
     - ☐ What was the maximal oral prednisone dose (mg/kg/d)?

○ ≤1 mg/kg/d	○ >1 to ≤1.5 mg/kg/d	○ >1.5 to <2 mg/kg/d	○ ≥2 mg/kg/d

   - ☐ Steroid-sparing immunosuppressant **(yes/no – select all that apply)**

• Methotrexate	• Leflunomide	• Azathioprine	• Mycophenolate (CellCept)	• MPA (Myfortic)
• Sirolimus	• Tacrolimus	• Thalidomide	• Lenalinomide	• Cyclosporine
• Hydroxychloroquine	• Sulfasalazine	• CYC (po)	• CYC (IV)	• Colchicine
• Other _____	•	•	•	•

*CYC = cyclophosphamide
*If cyclosporine was used, what was the titrated drug level? _____ ng/mL

   - ☐ IVIG

   - ☐ Non-biologic DMARDs:

• Baricitinib	• Tofacitinib

   - ☐ Biologic agents **(yes/no – select all that apply)**

• Rituximab	• Etanercept	• Infliximab	• Adalimumab	• Certolizumab
• Golimumab	• Anakinra	• Canakinumab	• Rilonacept	• Abatacept
• Tocilizumab	• Other: specify	•	•	•

Form C: At calcinosis diagnosis

Instructions: Answer the following questions regarding the patient's initial appearance and diagnosis of calcinosis

****Form E: JDM Disease activity, must also be completed for this time point.****

1. Date of calcinosis diagnosis: _____ (MM/YY/YYYY, enter in spreadsheet)
2. Date of last follow-up visit without calcinosis, prior to diagnosis of calcinosis: _____ (MM/DD/YYYY, enter in spreadsheet)
3. Were any symptoms, attributed to calcinosis, present prior to its diagnosis? **(yes/no)**
   - ☐ Estimate of duration present ____ (days)
   - ☐ Select the major symptom(s)? **(select all that apply)**
     - Pain
     - Swelling
     - Drainage
     - Ulceration
     - Incidental finding
     - Other: _____
4. Was there any physical trauma or other repetitive motion or contact to the area where calcinosis developed?
   - ☐ Describe _____
5. How did the physician identify calcinosis? **(select all that apply)**
   - ☐ Patient reported
   - ☐ History
   - ☐ Physical exam
   - ☐ Imaging
     a. If imaging was used, which modality first identified calcinosis?
     - X-ray
     - Computed tomography
     - Magnetic resonance imaging
     - Ultrasound
     - Scintigraphy
     - Other: specify
5. What physical exam findings were present? **(select all that apply)**
   - ☐ None
   - ☐ Visible nodules or masses
   - ☐ Palpable nodules or masses
   - ☐ Palpable firmness along fascial plane
   - ☐ Skin ulceration
   - ☐ Warmth, and/or redness
   - ☐ Drainage
   - ☐ Soft tissue swelling
   - ☐ Abnormal texture:
     - Firm
     - Fluctuant
   - ☐ Other, specify

6. What other effects of calcinosis were present, if any? **(select all that apply)**
   - ☐ Contracture
   - ☐ Mass effect
   - ☐ Infection
   - ☐ Erosion or fistula
   - ☐ Lipodystrophy
   - ☐ Lipoatrophy
   - ☐ Cutaneous atrophy or sclerosis
   - ☐ Other, specify
   - ☐ None

7. What phenotype of calcinosis was present? **(select all that were present) [if >1 checked = mixed]**
   - ☐ Circumscripta: superficial plaques or nodules.
   - ☐ Tumoral: larger nodular deposits with or without extension to deeper tissue layers.
   - ☐ Universalis: collections along fascial planes of tendons or muscles.
   - ☐ Exoskeleton: extensive hard deposits over all surface areas.
   - ☐ Other: describe

8. How many lesions were present? (if lesions were each discrete)

○ 1-5	○ 6-10	○ 11-15
○ 16-20	○ 21-30	○ > 30

9. Approximate size of largest lesions (if lesions were each discrete)

○ ≤ 1 cm	○ > 1 to ≤ 5 cm	○ > 5 to ≤ 10 cm
○ >10 to ≤ 20 cm	○ > 20 cm	○

10. Location of lesion(s) **(select all that apply)**

☐ Head and neck	☐ Chest	☐ Back	☐ Abdomen	☐ Buttock and genital areas
☐ Upper extremities	☐ Hands and fingers	☐ Lower extremities	☐ Feet and toes	☐ Other: specify

11. How was the patient impacted? **(select all that apply)**
   - ☐ Pain
   - ☐ Decreased range of motion or reduced mobility of affected area
   - ☐ Poor cosmesis
   - ☐ Worsened QOL score [# not required]
   - ☐ Worsened CHAQ score [# not required]
   - ☐ Other: specify

12. Which laboratory evaluations were obtained at the time of calcinosis diagnosis? **(select all that apply)**

☐ Lipids	☐ Hemoglobin A1C	☐ Ionized or total calcium
☐ Vitamin D level	☐ Parathyroid hormone	☐ Disease activity labs**
☐ Urinary calcium	☐ None	☐

** if disease activity selected will prompt to select from muscle enzymes, inflammatory markers

13. If obtained, which of these laboratory evaluations were low, high, normal or abnormal? **(select all that apply)**

☐ Lipids	☐ Hemoglobin A1C	☐ Ionized or total calcium
☐ Vitamin D level	☐ Parathyroid hormone	☐ Disease activity labs**
☐ Urinary Calcium	☐	☐

** if disease activity selected will prompt to select from muscle enzymes, inflammatory markers

14. Which of the following medications was the patient receiving in the interval from the most recent visit to the diagnosis of calcinosis? *(i.e. what were the background medications at the time calcinosis was diagnosed)* **(select all that apply)**

☐ None (patient was not on any medications at the time of calcinosis diagnosis)
☐ Vitamin D and/or Calcium supplementation
☐ IV methylprednisolone greater than 2 mg/kg/d? **(yes/no)**
☐ What interval/duration?

○ ≤ 5 days	○ Weekly	○ Every 2 weeks	○ Monthly

☐ Oral prednisone **(select dose)**

○ ≤ 0.5 mg/kg/d	○ > 0.5 to ≤ 1 mg/kg/d	○ > 1 to < 2 mg/kg/d	○ ≥ 2 mg/kg/d

☐ Steroid-sparing immunosuppressant **(yes/no – select all that apply)**

• Methotrexate	• Leflunomide	• Azathioprine	• Mycophenolate (CellCept)	• MPA (Myfortic)
• Sirolimus	• Tacrolimus	• Thalidomide	• Lenalinomide	• Cyclosporine
• Hydroxychloroquine	• Sulfasalazine	• CYC (po)	• CYC (IV)	• Colchicine
• Other _____	•	•	•	•

*CYC = cyclophosphamide
*If cyclosporine was used, what was the titrated drug level? _____ ng/mL

☐ IVIG

☐ Non-biologic DMARDs:

• Baricitinib	• Tofacitinib

☐ Biologic agents **(yes/no – select all that apply)**

• Rituximab	• Etanercept	• Infliximab	• Adalimumab	• Certolizumab
• Golimumab	• Anakinra	• Canakinumab	• Rilonacept	• Abatacept
• Tocilizumab	• Other: specify	•	•	•

ANSWER THE FOLLOWING QUESTIONS REGARDING THE TREATMENT PRESCRIBED ONCE CALCINOSIS WAS IDENTIFIED

15. Was an increase in systemic immunosuppression prescribed following calcinosis diagnosis? **(yes/no)**

☐ IV methylprednisolone **(yes/no)**
If yes, was the maximal dose greater than 2 mg/kg/day? **(yes/no)**
If yes, did the patient receive IV methylprednisolone ≥ 7 days? **(yes/no)**

☐ Oral prednisone **(yes/no)**
If yes, what was the maximal oral prednisone dose (mg/kg/d)?

○ ≤ 0.5 mg/kg/d	○ > 0.5 to ≤ 1 mg/kg/d	○ > 1 to < 2 mg/kg/d	○ ≥ 2 mg/kg/d

☐ Steroid-sparing immunosuppressant **(yes/no – select all that apply)**

• Methotrexate	• Leflunomide	• Azathioprine	• Mycophenolate (CellCept)	• MPA (Myfortic)
• Sirolimus	• Tacrolimus	• Thalidomide	• Lenalinomide	• Cyclosporine
• Hydroxychloroquine	• Sulfasalazine	• CYC (po)	• CYC (IV)	• Colchicine
• Other _____	•	•	•	•

*CYC = cyclophosphamide
*If cyclosporine was used, what was the titrated drug level? _____ ng/mL

☐ IVIG

☐ Non-biologic DMARDs:

• Baricitinib	• Tofacitinib

☐ Biologic agents **(yes/no – select all that apply)**

• Rituximab	• Etanercept	• Infliximab	• Adalimumab	• Certolizumab
• Golimumab	• Anakinra	• Canakinumab	• Rilonacept	• Abatacept
• Tocilizumab	• Other: specify	•	•	•

16. Other therapy started following calcinosis diagnosis? **(select all that apply)**

☐ Bisphosphonates **(select all that apply)**

• Alendronate	• Etidronate	• Pamidronate	• Zoledronic Acid

** If pamidronate was used, select which treatment regimen:
- ☐ 1 mg/kg every month
- ☐ 1 mg/kg/d for 3 consecutive days, repeated every 3 months
- ☐ Other pamidronate regimen _____

☐ Other agents that effect calcium or phosphorous

• Vit D or Ca	• Calcium-channel blocker	• Sodium thiosulfate, topical	• Sodium thiosulfate, intravenous
• Sodium thiosulfate intra-lesional	• Aluminum hydroxide	• Warfarin	• Minocycline
• Probenecid	• Glucocorticoid, topical	• Glucocorticoid, intra-lesional	• Other

17. Was the patient referred for surgical excision **(yes/no)**
    a. If referred, did surgical excision occur? **(yes/no)**
        - ☐ Was the entire lesion(s) resected? (yes/no)
        - ☐ Were there complications with wound healing (yes/no)
        - ☐ Did the lesion recur? (yes/no)
            1. If recurred, how long after resection? ____ (months)

Form D: Treatment response of calcinosis

Instructions: Answer the following questions regarding the treatment regimen that was prescribed at calcinosis diagnosis once the maximal treatment response was achieved and/or when the treatment regimen was altered or abandoned.

****Form E: JDM Disease activity, must also be completed for this time point****

1. Based on above instructions, what was the duration of the prescribed treatment regimen against calcinosis?
   _____ (days)

2. What was the degree of response?
   - ☐ Worsened calcinosis.
   - ☐ Unchanged calcinosis, without new lesions
   - ☐ Mild/partial improvement
   - ☐ Moderate/significant improvement
   - ☐ Complete/total resolution

3. If there was a positive response, how was this response measured? **(select all that apply)**
   - ☐ Change **(Increase/Decrease/None)** in features on history or physical exam?

☐ Size or Number	☐ Warmth or redness	☐ Drainage	☐ Texture

   - ☐ Change **(increase/decrease/none)** of other effects **(select all that apply)**

☐ mass effect	☐ contracture	☐ infection	☐ erosion or fistula
☐ lipoatrophy	☐ lipodystrophy	☐ cutaneous atrophy or sclerosis	☐

   - ☐ Changes **(Increase/Decrease/None)** in patient factors **(select all that apply)**
     - ○ Change in level of pain
     - ○ Change in range of motion or mobility
     - ○ Change in cosmesis
     - ○ Change in QOL
     - ○ Change in CHAQ
     - ○ Change in DAS

   - ☐ Changes **(Increase/Decrease/None)** by imaging? **(select all that apply)**

☐ X-ray	☐ Magnetic resonance imaging	☐ Scintigraphy
☐ Computed tomography	☐ Ultrasound	

4. What was the fate of the treatment at the time of *maximal* response and/or change in therapy?
   - ☐ Continued, but with decreased dose or interval.
   - ☐ Continued, but with increased dose or interval.
   - ☐ Continued unchanged.
   - ☐ Continued, but also added another treatment: **[prompts treatment selection choices, below (i)]**
   - ☐ Changed to an alternative treatment: **[prompts treatment selection choices, below (i)]**
   - ☐ If changed or discontinued, what was the main reason for doing so?
     - ○ Complete response (no longer needed)
     - ○ Inadequate response
     - ○ Adverse effects (infection, organ toxicity, etc), specify which.
     - ○ Medication intolerance, specify which.
     - ○ Prohibitive cost / insufficient insurance coverage

[Based on selections in question #4]

i. Please select the corresponding treatment(s) added in place of or in addition to the previous therapy **(select all that apply)**

- ☐ IV methylprednisolone
- ☐ Oral prednisone
- ☐ Steroid-sparing immunosuppressant **(yes/no – select all that apply)**

• Methotrexate	• Leflunomide	• Azathioprine	• Mycophenolate (CellCept)	• MPA (Myfortic)
• Sirolimus	• Tacrolimus	• Thalidomide	• Lenalinomide	• Cyclosporine
• Hydroxychloroquine	• Sulfasalazine	• CYC (po)	• CYC (IV)	• Colchicine
• Other _____	•	•	•	•

*CYC = cyclophosphamide

*If cyclosporine was used, what was the titrated drug level? _____ ng/mL

- ☐ IVIG
- ☐ Non-biologic DMARDs:

• Baricitinib	• Tofacitinib

- ☐ Biologic agents **(yes/no – select all that apply)**

• Rituximab	• Etanercept	• Infliximab	• Adalimumab	• Certolizumab
• Golimumab	• Anakinra	• Canakinumab	• Rilonacept	• Abatacept
• Tocilizumab	• Other: specify	•	•	•

- ☐ Bisphosphonates **(select all that apply)**

○ Alendronate	○ Etidronate	○ Pamidronate	○ Zoledronic Acid

If pamidronate was used, select which treatment regimen:

- ☐ 1 mg/kg every month
- ☐ 1 mg/kg/d for 3 days every 3 months
- ☐ Other pamidronate regimen: _____

- ☐ Other agents that effect calcium or phosphorous

☐ Vit D or Ca	☐ Calcium-channel blocker	☐ Sodium thiosulfate, topical	☐ Sodium thiosulfate, intravenous
☐ Probenecid	☐ Aluminum hydroxide	☐ Warfarin	☐ Minocycline
☐ Topical glucocorticoid	☐ Intra-lesional glucocorticoid	☐	☐

- ☐ Was the patient referred for surgical excision **(yes/no)**
  a. If referred, did surgical excision occur? **(yes/no)**
     - ○ Was the entire lesion(s) resected? (yes/no)
     - ○ Were there complications with wound healing (yes/no)
     - ○ Did the lesion recur? (yes/no)
       - ▪ If recurred, how long after resection? ____ (months)

If the Form D, question #4 answer selection(s) prompts new treatment options, then Form D (and Form E) need to be completed again to capture the effect(s) of the additional treatment(s). This pattern continues as long as each successive entry of Form D, question #4 contains selections that prompt new treatment selection options.

Form E: JDM disease activity

Instructions: Answer the following questions related to overall JDM disease activity. This form should be completed for each patient entry to describe the overall JDM disease activity at different time points of the patient's course:

- At initial JDM diagnosis
- At initial diagnosis of calcinosis
- At time of maximal response to the therapy prescribed for calcinosis

1. Is the patient's JDM active?
   - ☐ No.
   - ☐ Yes

○ Mild	○ Moderate	○ Severe

2. Is there active muscle disease?
   - ☐ No.
   - ☐ Yes **(select all that apply)**

○ Symmetric proximal muscle weakness on exam	○ Abnormal CMAS: ____	○ Abnormal MMT8: ____
○ Abnormal muscle MRI	○ Abnormal EMG	○ Abnormal muscle biopsy

   a. Elevated muscle enzymes? **(yes/no - select all that apply)**
      a. Creatine Kinase

☐ ____ units/L	☐ ____ ULN

      b. Lactate dehydrogenase

☐ ____ units/L	☐ ____ ULN

      c. Aldolase

☐ ____ units/L	☐ ____ ULN

      d. Aspartate aminotransferase (AST)

☐ ____ units/L	☐ ____ ULN

      e. Alanine aminotransferase (ALT)

☐ ____ units/L	☐ ____ ULN

3. Presence of other abnormally elevated markers? **(select all that apply)**
   - ☐ Erythrocyte sedimentation rate
   - ☐ C-reactive protein
   - ☐ Von Willebrand Factor Antigen
   - ☐ Neopterin
4. Is there active skin disease?
   - ☐ No.
   - ☐ Yes **(select all that apply)**

○ Malar or facial rash	○ Gottron's papules or sign	○ Heliotrope
○ Cutaneous ulceration	○ Periungal capillary loop changes	○ Cuticular overgrowth
○ Non-sun exposed erythema	○ Extensive cutaneous erythema	○ Livedo reticularis
○ Linear extensor erythema	○ Mucus membrane lesions	○ Subcutaneous edema
○ Panniculitis	○ Alopecia (non-scarring)	○ Shawl sign
○ V sign	○	○

5. Are there other active disease features present? **(select all that apply)**
   - ☐ Fever
   - ☐ Fatigue
   - ☐ Weight loss

- ☐ Arthritis
- ☐ Organ involvement related to JDM

○ GI ulceration	○ Pericarditis and/or Myocarditis	○ Arrhythmia
○ Dysphonia	○ Respiratory muscle weakness	○ ILD
○ Dysphagia	○ Abdominal pain	○

6. Are there any disease damage indicators?

Global

○ Death due to myositis	○ Malignancy	○

Muscle

○ Muscle atrophy	○ Weakness not due to active myositis	○ Muscle dysfunction (dec aerobic capacity)

Skeletal

○ Contractures	○ Osteoporosis with fracture	○ Avascular necrosis
○ Deforming arthropathy	○	○

Cutaneous

○ Depressed scar/atrophy	○ Poikiloderma	○ Lipoatrophy/lipodystrophy
○ Scarring alopecia	○ Sclerodactyly	○

Gastrointestinal

○ Persistent dysphagia	○ Dysmotility, constipation, diarrhea or pain	○ Infarction or resection of bowel or other GI organs

Pulmonary

○ Impaired lung function	○ Dysphonia	○ Pulmonary fibrosis
○ Pulmonary hypertension	○	○

Cardiovascular

○ Ventricular dysfunction	○ Hypertension treated ≥ 6 months	○ Angina or CAB
○ Myocardial infarction	○	○

Vascular

○ Tissue or pulp loss	○ Digit or limb loss	○ Venous or arterial thrombosis
○ Swelling, ulceration, venous stasis	○	○

Endocrine

○ Growth failure	○ Diabetes mellitus	○ Delay of secondary characteristics
○ Hirsutism	○ Irregular menses	○ Amenorrhea
○ Infertility	○ Sexual dysfunction	○

Ocular

○ Cataract	○ Visual loss, not due to cataract	○

Infection

○ Chronic infection	○ Multiple infections	○