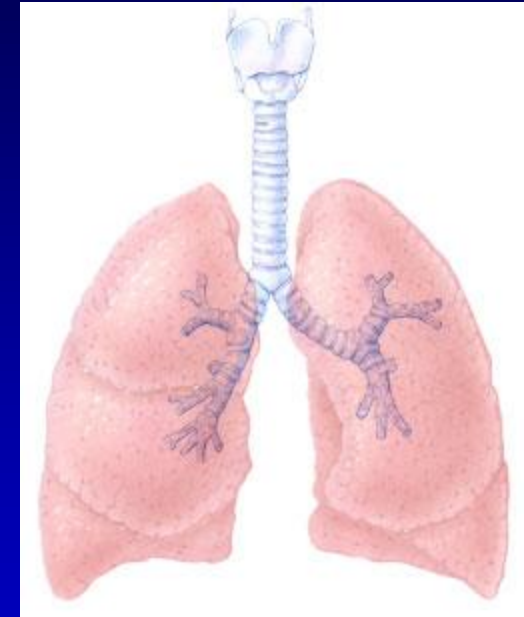
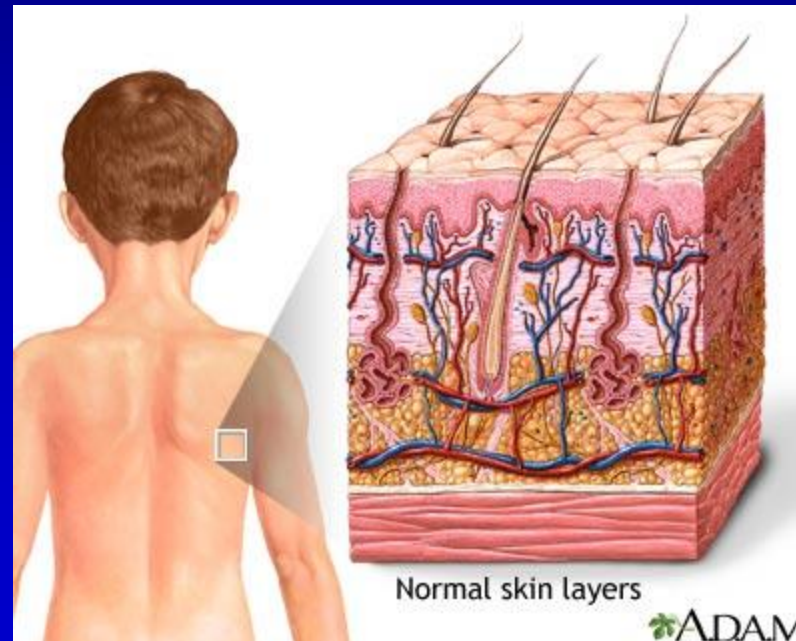
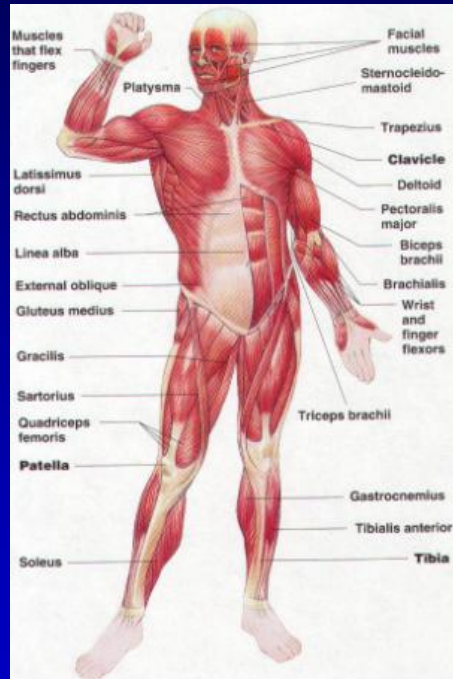


Treating dermatomyositis

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September 25, 2010



DM affects many organs



Approaching DM therapy

- **Avoid things that make disease worse**

Cardinal Rule:

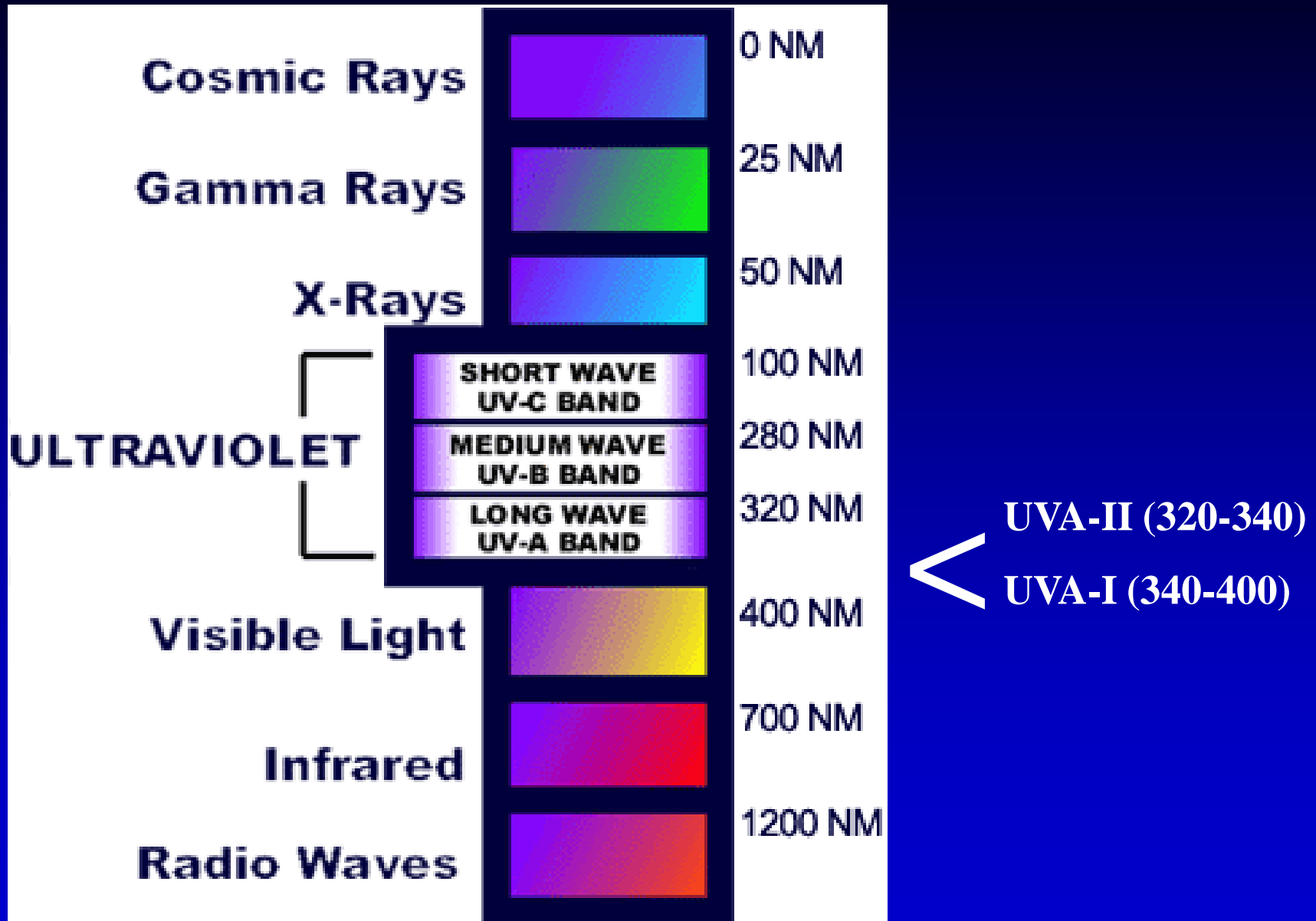
Avoid things that worsen skin disease

- **Dry skin**
- **Smoking**
- **Sunlight**

Sunprotection



Components of Radiation from the Sun



SPF factor refers to protection from what UV wavelengths?

- **UVA**
- **UVB**
- **UVA and UVB**
- **Neither**

SPF factor refers to protection from what UV wavelengths?

- **UVB**

Ultraviolet Light: Physical Protection

- Wear tightly-woven protective *broad-brim* hats and clothing, e.g. long-sleeves and pants





SPF 3-7



SPF 6.5

Ultraviolet Light: Physical Protection

- Wear tightly-woven protective *broad-brim* hats and clothing, e.g. long-sleeves and pants
- Use laundry product that can be added to detergent to increase UV protection in clothing, e.g. Rit Sun Guard (ritdye.com)
- Apply UV blocking films to windows in cars and home

Sunscreen: which one??

- **SPF >30**
- **Look for**
 - **Titanium dioxide; OR**
 - **Zinc oxide**





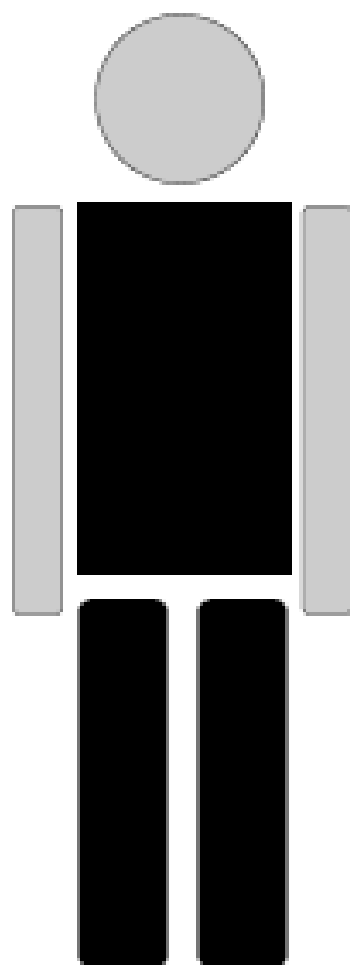
Sunscreen: which one??

- **SPF >30**
- **Look for**
 - Titanium dioxide; OR
 - Zinc oxide
- **Need multiple chemical blockers**

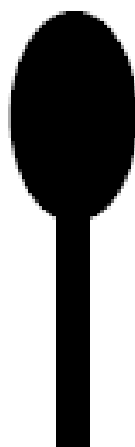


Chemical UVA blockers

- **Avabenzone (Parsol 1789)**
- **Oxybenzone**
- **Mexoryl SX**



Use More Than Half a
Teaspoon Each on
Head and Neck Area
Right Arm
Left Arm



Use More Than a
Teaspoon Each on
Anterior Torso
Posterior Torso
Right Leg
Left Leg

Approaching DM therapy

- **Avoid things that make disease worse**
- **Determine if medical therapy is indicated**

Activity



Damage

Determining disease activity

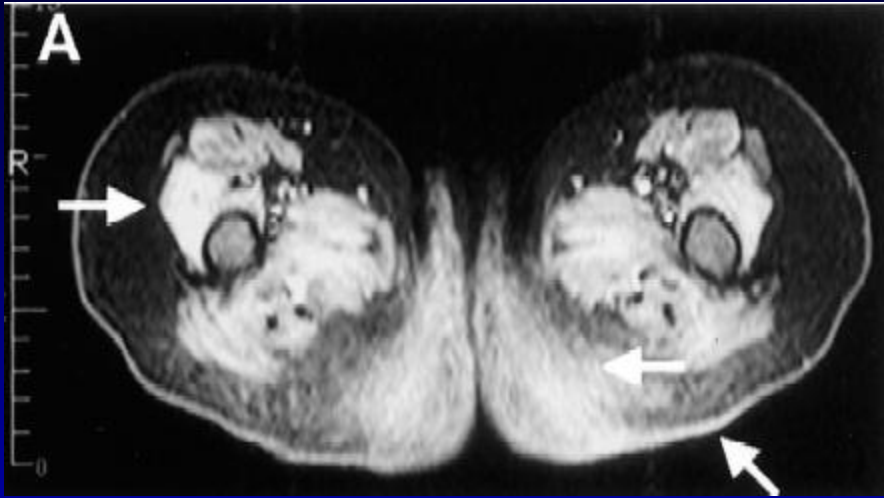
- **Muscle**
 - **Weakness not necessarily helpful**
 - **Muscle enzymes in blood**

The role of EMG

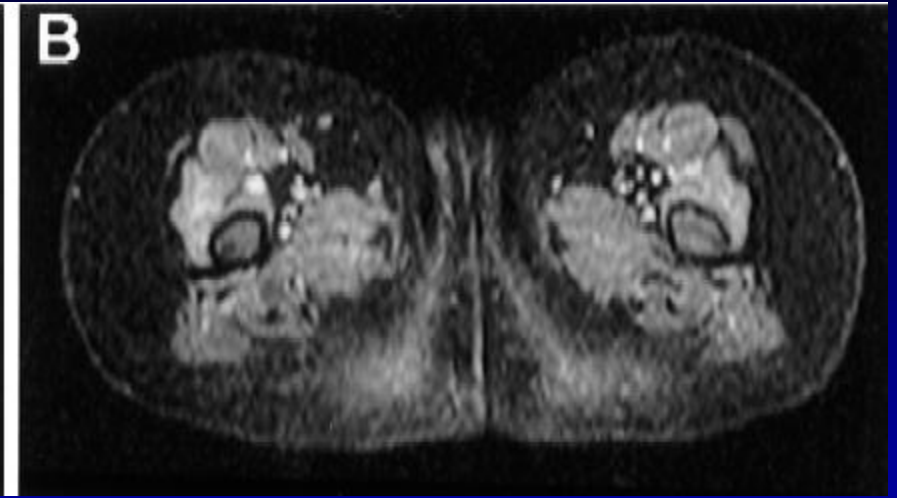
- **Can diagnose a primary myopathy**
- **Cannot differentiate between myopathies**
- **Helpful if weak on exam but muscle enzymes normal**



MRI



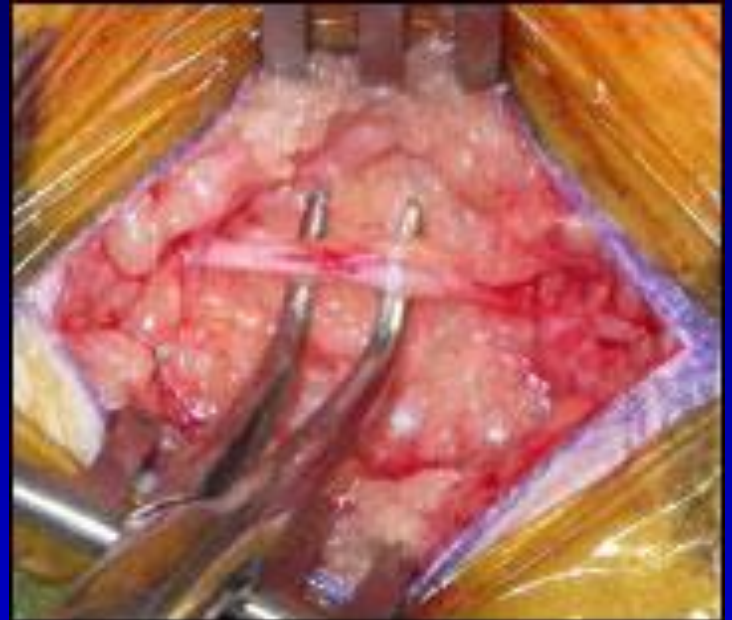
Before therapy



After therapy

The role of muscle biopsy

- Can be diagnostic for dermatomyositis
- Not 100% sensitive



Determining disease activity

- **Skin**
 - **Redness not always helpful**







Determining disease activity

- **Skin**
 - Redness not always helpful
 - ITCH

Approaching DM therapy

- **Avoid things that make disease worse**
- **Determine if medical therapy is indicated**
- **Select optimal medical therapy**

General concepts

- **Not every therapy works for everybody**
- **Therapy is empiric—trial and error**
- **Don't give up!**

Corticosteroids

- **Generally effective for most organ systems**
- **Act quickly**
- **Cheap**
- **Limited by long term side effects**

IVIG

- **Pooled preparation of blood proteins from multiple donors**
- **Generally effective for skin and muscle**
- **Acts quickly**
- **Side effects: allergic response, kidney, rare chance of transmitting infection**
- **EXPENSIVE**
- **Tends to lose effectiveness over time**

Methotrexate

- **Mechanism of action actually not clear**
- **Often effective for muscle and/or skin**
- **Slower to act**
- **Cheap**
- **Side effects: liver, GI, low blood counts, rare lung inflammation**
- **Risks of infection, ?cancer (lymphoma)**

Azathioprine

(Imuran)

- **Inhibits ability of lymphocytes to multiply**
- **Can be effective for muscle or skin (lung)**
- **Cheap**
- **Side effects: allergy, low white blood cells, liver irritation**
- **Risks of infection, ?cancer**

Mycophenolate mofetil

(Cell Cept)

- **Inhibits ability of lymphocytes to multiply**
- **Can be very effective for muscle/skin**
- **Lung data look promising**
- **Side effects: GI upset, low blood counts,**
- **Risks of infection (?herpes, shingles), cancer?**
- **EXPENSIVE**

Cyclophosphamide (Cytosxan)

- **Inhibits ability of lymphocytes to multiply**
- **Generally reserved for severe disease**
- **Can be useful for ulcerations/gangrene of skin**
- **Side effects: low blood counts, ovarian failure, bladder irritation/cancer**

Skin-specific therapies

ITCH



Itch: Basic Treatments

- **Dry skin care**
 - **Moisturizing soaps**
 - **Apply moisturizing cream within 3 minutes after bathing and again during day**
 - **Avoid hot showers; keep less than 10 min**
 - **Consider use of humidifiers**
 - **Minimize use of central heating**

Itch: Topical Therapy

- **Immediately soothing**
 - **Menthol-based (i.e. Sarna lotion)**
 - **Pramoxine-based (i.e. Prax lotion, pramasone)**
 - **Avoid topical anti-histamines**
 - **May cause allergies over time**
 - **Cool compresses or ice**

Itch: Prescription Therapy

- **Systemic**
 - **Anti-histamines (hydroxyzine, doxepin)**
 - **Naltrexone**
 - **Mirtazapine**
 - **Gabapentin**
 - **Thalidomide**
 - **Butorphanol**

Best therapy for itch.....

Treat underlying inflammation!

Topical therapies for DM

- Corticosteroids

Topical Corticosteroids

- **Group I steroids (e.g., Temovate)** - use cyclically in others
- **Group II steroids (e.g., Lidex)**
- **Group III-IV steroids (e.g., triamcinolone acetonide 0.025-0.1%)** for general widespread use on trunk and extremities
- **Group VI-VII steroids (e.g., hydrocorticone cream 0.1%)** for face and flexural areas/body folds

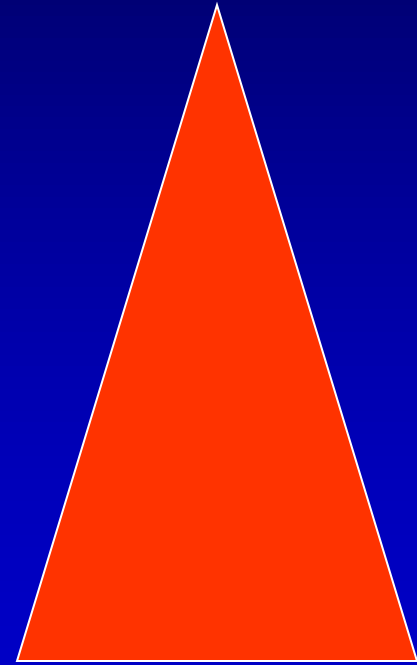
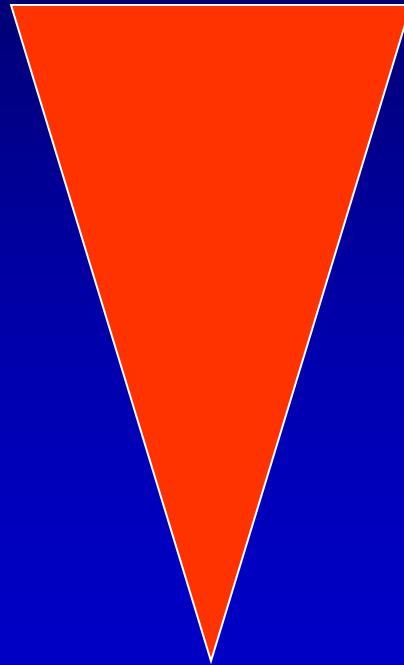
Topical Corticosteroids

Patient acceptance
& compliance

Moisturization

- Vehicles

- sprays
- solutions
- foams
- gels
- creams
- ointments



Topical therapies for DM

- **Corticosteroids**
- **Immunomodulators**
 - **Tacrolimus (Protopic)**
 - **Pimecrolimus (Elidel)**

Hydroxychloroquine (Plaquenil)

- **Mechanism of action unknown—generally NOT immunosuppressive**
- **Can be effective for skin, not muscle**
- **Side effects: GI, rare liver/muscle inflammation, rare eye toxicity**
- **SKIN RASH (up to 30% DM patients)**
- **Consider switch to chloroquine**

Dapsone

- **Sulfa type drug**
- **Blocks lymphocyte function**
- **Can be effective for mild skin disease**
- **Side effects: allergy, anemia, liver irritation, nerve damage**

Leflunomide

(Arava)

- **Blocks ability of lymphocytes to multiply**
- **Approved for Rheumatoid Arthritis**
- **2 Case Reports of effectiveness for skin and muscle disease**
- **Side effects: GI, low blood counts, liver**

Approaching DM therapy

- **Avoid things that make disease worse**
- **Determine if medical therapy is indicated**
- **Select optimal medical therapy**
- **Damage reversal**

Muscle damage

- **Non inflammatory weakness**
- **Determine cause**
 - **Atrophy from previous DM inflammation**
 - **Corticosteroids**
 - **Lack of use**
- **Physical therapy**
- **Low weight bearing exercise is helpful**

Skin damage

- **Enlarged blood vessels**



Skin damage

- **Enlarged blood vessels**
 - **Laser (intense pulsed light) therapy**

Skin damage

- **Enlarged blood vessels**
 - **Laser (intense pulsed light) therapy**
- **Calcium deposits**



Managing calcinosis cutis

- **Aggressive prevention is the key**
- **Treatments**
 - **Treat superinfection and pain**
 - **Aluminum hydroxide, warfarin, Ca⁺⁺ channel blockers, bisphosphonates**
 - **Local surgical removal**

New therapies?

- **Rituximab**
 - **Kills B lymphocytes**
 - **Preliminary data suggest it can help muscle disease; we have not found it effective for skin disease**
 - **Large, controlled trial (RIM) completed and await data**

New therapies?

- **Blocking interferon signaling**
 - Evidence that interferon is overactive in skin, muscle and blood of DM patients
 - **MEDI 545 (Medimmune)**—early phase I trial completed; no data released.

What will improve our therapy?

- **Need “biomarkers” to determine how much of symptoms will be reversible with medication**
- **Need better classification of disease so we can individualize therapy**
- **Better understanding of what actually causes the disease**