
FOR A CONSISTENT LOOK

Myositis Overlapping Diseases

Rossitza I. Chichkova, MD, MS

Associate Professor, Neurology and Internal Medicine
University of South Florida/James A. Haley Veterans
Hospital, Tampa, FL

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- Systemic sclerosis (scleroderma)
- Sjogren syndrome
- Rheumatoid arthritis
- Lupus erythematosus
- Mixed connective tissue disease
- Raynaud syndrome

SCLERODERMA

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- Scleroderma - rare disease that involves hardening and tightening of the skin and connective tissues
- The connective tissues support the body structures
- It may affect only the skin
- In many people, scleroderma also affects the blood vessels, internal organs and the digestive tract
- Women are more often affected than men
- Age of onset - 30-50 years of age
- There is no cure, but there are medications that help the symptoms and improve quality of life

Skin

- Involved in majority of the patients
- Skin gets thick and hard
- Patchy or large areas - variable number, location and
- Shiny skin
- Restricted movement
- Contractures
- Calcium deposits
- Tightening of facial skin – small and narrow mouth, hard to brush teeth or have dental exam and cleaning
- Dry mouth and risk of dental decay



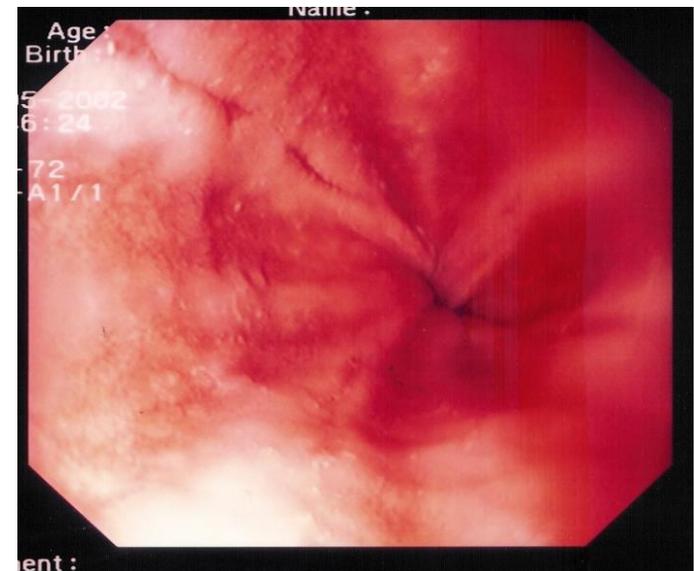
Raynaud phenomenon

- Early sign
- Exaggerated response to cold temperatures or emotional distress
- Fingers and toes get numb and painful
- Color changes in the fingers or toes
- Seen not only in scleroderma
- Can cause gangrene

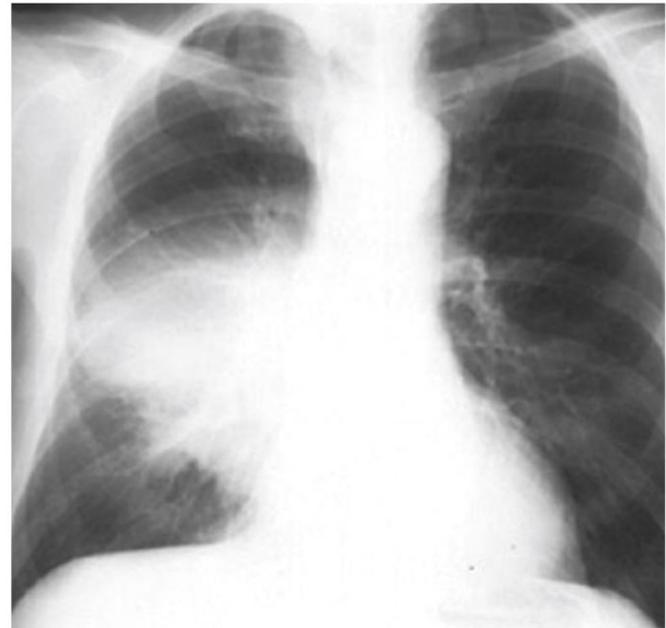
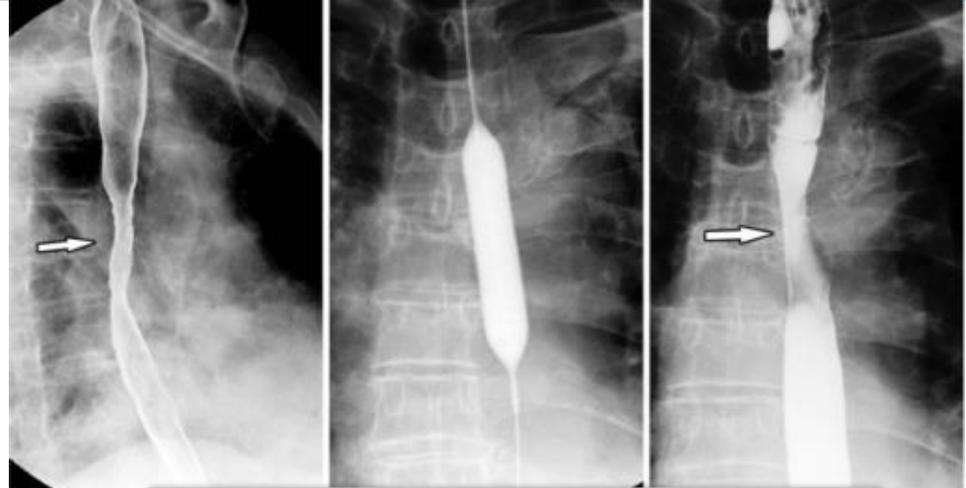


Digestive system

- Acid reflux
- Damage of the esophagus due to reflux
- Abnormal motility of the GI tract
- Difficulty swallowing
- Decreased absorption of nutrients
- Malabsorption
- Early recognition
- Swallowing /GI evaluation
- Recommendations may be different for the same person at different times
- Elevate bed at night



- Aspiration
- Aspiration pneumonia
- Asphyxia
- Enlargement of the esophagus
- Strictures of esophagus
- Esophagitis
- Ulcers



Heart

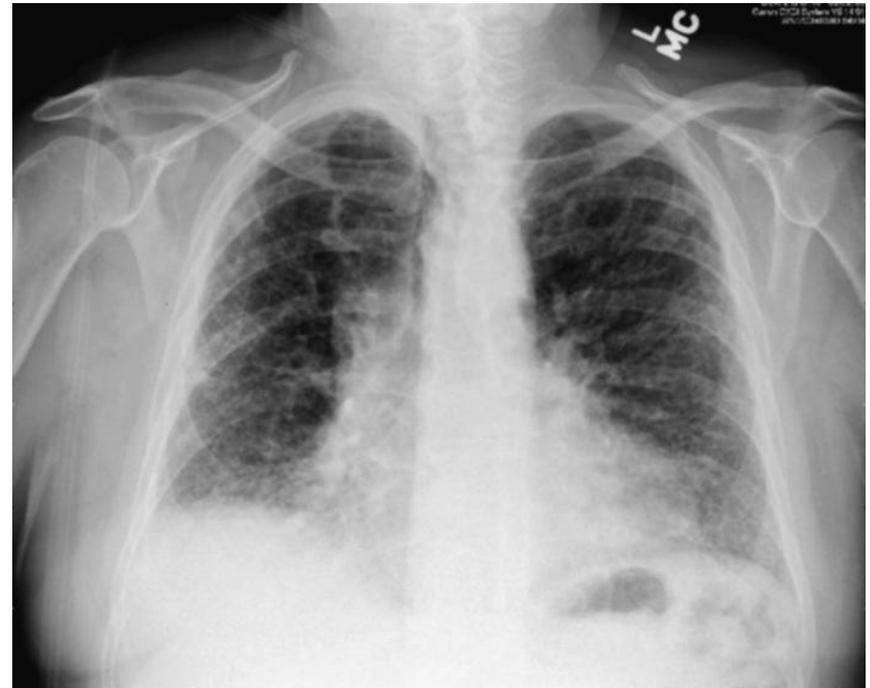
- The heart muscle becomes thickened from fibrous scar tissue accumulation
- Decreased the filling with blood and force of contractions
- Ultimately may result in heart failure
- Spasm of the coronary arteries (the main blood vessels to the heart), may cause chest pain and lead to heart attack
- Similar to the spasm of blood vessels causing Raynaud phenomenon of the fingers
- Inflammation of the outer heart lining (pericarditis) may cause pain and accumulation of fluid around the heart that restricts the filling and output
- Irregular heartbeats – arrhythmia
- Cardiology evaluation

Lungs

- Collagen build-up thickens lung tissue and causes fibrosis
- Impairs oxygen transport into the blood
- Pulmonary fibrosis - type of Interstitial Lung Disease (ILD)
- Pulmonary hypertension-narrowing and high pressure in the pulmonary arteries that burdens the right side of the heart to move blood into the lungs to pick up oxygen—may lead to right heart failure
- Shortness of breath
- Decreased exercise tolerance
- Persistent cough

Lungs

- Quit smoking
- Avoid air pollutants
- Breathing exercises and aerobic exercise
- Evaluation by cardiology/pulmonary specialists
- Chest X-ray
- Echocardiogram (ultrasound of the heart)
- Pulmonary function tests (breathing test)
- CT of the lungs



Kidneys/Genitals

- Kidney involvement may lead to high blood pressure
- Protein in the urine and abnormal kidney function tests
- Acute renal failure - kidneys can't eliminate waste products – decreased urine output, blood pressure rise, headache, vision problems, confusion
- Monitor blood pressure
- Urine and kidney function tests
- Monitor urine output – decreasing output is alarming sign – go to hospital
- Follow up with PCP or nephrologist
- Sexual dysfunction - erectile dysfunction in men, decreased sexual lubrication and constricting of the vagina in women

Muscle involvement

- Frequency: 14% to 17%
- Weakness – proximal, mild
- Usual mean onset - 6 years after scleroderma onset
- Often non-progressive
- Male more than female
- Higher prevalence of cardiac, lung, skin involvement, neuropathy
- Antibodies –ANA (95%), Scl-70, PM-Scl- 75, U3-RNP, Ku etc.
- EMG – Myopathy
- Serum CK - May be high
- Muscle biopsy is not well defined
- Treatment: Corticosteroids; Cyclosporine; Methotrexate

SJOGREN'S SYNDROME



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- Immune disorder affecting the mucous membranes and moisture-secreting glands
- Dry eyes –itchy and as if there is sand in them
- Dry mouth – hard to speak and swallow
- Dry nose – may have change in smell
- Joint pain, swelling and stiffness
- Swollen salivary glands- behind the jaw, in front of the ears
- Skin rashes, dry skin, vaginal dryness
- Persistent dry cough
- Prolonged fatigue
- May accompany other immune system disorders -rheumatoid arthritis, lupus, myasthenia gravis, myositis
- Develops at any age, most are older than 40, more common in women

Common complications

- Dental cavities - saliva protects from bacteria and decay
- Yeast infections – predisposed to oral thrush- mouth yeast infection
- Vision problems - dry eyes lead to light sensitivity, blurred vision and corneal damage

Less common complications:

- Lungs, kidneys, liver - Inflammation may cause pneumonia and bronchitis, kidney dysfunction, hepatitis or cirrhosis of the liver
- Lymph nodes - Small % develop lymphoma
- Muscle pain – 20-30%
- Myopathy – 10% - inflammatory 50-70%
- Peripheral neuropathy - numbness, tingling, burning in hands and feet

Sjogren's - Diagnosis

- Can be difficult to diagnose
- Medications can cause dry mouth, allergies can cause dry eyes
- Blood tests-ANA (70%), RF (65%), SS-A (70%) and SS-B (40%), sed rate, IgG, liver and kidney tests
- Eye tests for eye dryness - Schirmer test
- Eye exam-look for damage to the cornea
- Imaging - sialogram - shows amount of saliva; scintigraphy - radioactive isotope tracks flow of saliva
- Lip biopsy - detect the presence of inflammatory cells, which can indicate Sjogren's syndrome



- **Decrease eye inflammation** – OTC eye drops for mild to moderate dry eyes. Prescription eyedrops - cyclosporine (Restasis) or lifitegrast (Xiidra) for moderate to severe dry eyes
- **Increase production of saliva** - pilocarpine (Salagen) and cevimeline (Evoxac) can increase the production of saliva
- **Address specific complications.** Arthritis symptoms - nonsteroidal anti-inflammatory drugs (NSAIDs) or other arthritis medications.
Yeast infections - antifungal medications
- **Treat systemic symptoms.** Hydroxychloroquine (Plaquenil), a drug designed to treat malaria, is often helpful in treating Sjogren's syndrome. Other immune suppressants - methotrexate (Trexall).
- Surgery - minor procedure of the tear ducts.

RHEUMATOID ARTHRITIS

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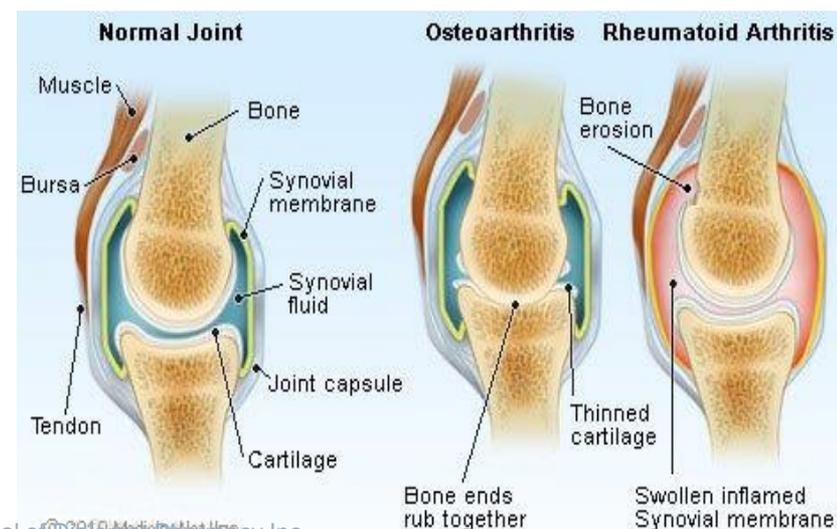
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- Chronic autoimmune inflammatory disorder that affects joints, skin, eyes, lungs, heart and blood vessels
- It is not osteoarthritis (OA) caused by wear-and-tear; RA affects the lining of your joints, causing a painful swelling, bone erosion and joint deformity
- Treatment dramatically improved
- RA can cause severe physical disabilities
- Women more affected than men
- Any age, most common between 40 and 60.
- Increased risk with family history of RA, smoking, environmental exposures? (emergency workers from 911 with higher risk for autoimmune disorders), obesity

RA - Symptoms

- Tender, warm, swollen joints of the fingers and toes
- Joint stiffness - worse in the mornings and after inactivity
- May progress to wrists, knees, ankles, elbows, hips and shoulders
- Fatigue, fever and weight loss
- RA can affect skin, eyes, lungs, heart, kidneys, salivary glands, nerve tissue, bone marrow, blood vessels
- Varies in severity, flairs and remission
- Over time, RA can cause joints to deform and shift out of place



- Osteoporosis - due to RA or with some medications
- Rheumatoid nodules - hard bumps of tissue that form around pressure points, such as the elbows or elsewhere
- Dry eyes and mouth
- Infections – due to RA or medications
- Compression of nerves - Carpal tunnel syndrome
- Muscle weakness – due to inflammation of the muscle arteries
- Neuropathies - due to inflammation of the nerve arteries
- Heart – increases risk of blockage of arteries (angina, heart attack) and inflammation of the heart sac (pericarditis)
- Lungs - risk of inflammation and scarring of the lungs - leads to progressive shortness of breath
- Lymphoma - increases the risk of lymphoma

- Can be difficult in early stages – mimics other diseases
- No one single blood test to confirm the diagnosis
- Blood tests - elevated sed rate or C-reactive protein (CRP), rheumatoid factor and anti-cyclic citrullinated peptide (anti-CCP) antibodies.
- Imaging tests
- X-rays of bones and joints - to diagnose or track progression of the disease or organ involvement
- MRI
- CT
- Ultrasound
- EMG/NCS

Medications - depend on the severity of the disease

- NSAIDs - relieve pain and reduce inflammation, OTC or prescription.
- Steroids - prednisone reduces inflammation and pain and slows joint damage. Have a lot of side effects
- **Disease-modifying antirheumatic drugs (DMARD)** - slow RA progression, joint and tissue damage. Examples: methotrexate, leflunomide (Arava), hydroxychloroquine (Plaquenil) and sulfasalazine (Azulfidine). Side effects may include liver damage, bone marrow suppression and severe lung infections
- **Biologic agents** - abatacept (Orencia), adalimumab (Humira), anakinra (Kineret), certolizumab (Cimzia), etanercept (Enbrel), golimumab (Simponi), infliximab (Remicade), rituximab (Rituxan), tocilizumab (Actemra) and tofacitinib (Xeljanz) - target parts of the immune system that trigger inflammation. Increase infection risk.
- Biologic are usually most effective when paired with a nonbiologic DMARD, such as methotrexate.

Therapy

- Physical or occupational therapy to keep joints flexible
- Techniques to deal with ADL when physical limitations are present
- Assistive devices to avoid stressing of painful joints.
- Adapted tools for kitchen, dressing, hygiene etc.
- Catalogs and medical supply stores

Surgery

- Repair damaged joints
- Restore ability to use your joint
- Reduce pain and correct deformities
- Synovectomy - remove inflamed joint lining -knees, elbows, wrists, fingers and hips
- Tendon repair
- Joint fusion - to stabilize or realign; for pain relief
- Total joint replacement

LUPUS ERYTHEMATOSUS

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- Systemic autoimmune disease that causes inflammation of many systems - joints, skin, kidneys, blood cells, brain, heart and lungs.
- Symptoms may mimic those of other diseases
- Butterfly-shaped rash on the face that covers the cheeks and bridge of the nose or rashes elsewhere on the body



- Symptoms may come on suddenly or develop slowly
- Severity varies; episodes of flares and remissions
- Most common signs and symptoms – fatigue, fever, joint pain, stiffness and swelling
- Other symptoms - Raynaud's phenomenon, shortness of breath, chest pain, dry eyes, headaches, weakness, confusion and memory loss
- Myopathy in 10 % - inflammation of muscle
- Some people are predisposed to LE

Potential triggers:

- Sunlight - brings on lupus skin lesions
- Infections - can initiate lupus or cause a relapse
- Medications that may induce LE – hydralazine, quinidine, isoniazid, procainamide
- Medications that may cause worsening - antibiotics, anticonvulsants, hormones, NSAIDs, dermatologic meds
- Usual improvement with stopping the medication. Rarely, they may persist.

Risk factors:

- More common - women, African-American, Hispanics and Asian-Americans
- Affects all ages, most often 15 and 45.

- **Kidneys-** can cause serious kidney damage. Kidney failure is one of the leading causes of death is LE
- **Nervous system** - headaches, dizziness, behavior changes, memory loss, vision problems, and even strokes or seizures.
- **Blood/blood vessels** – anemia, increased risk of bleeding or blood clotting, blood vessel inflammation (vasculitis)
- **Lungs** - inflammation of the chest cavity lining (pleurisy), which can cause pain with breathing. Bleeding into lungs and pneumonia also are possible
- **Heart** - inflammation of the heart muscle, arteries or heart membrane (pericarditis). The risk of cardiovascular disease and heart attacks increases greatly as well.

Other complications - Increases risk of:

- Infection – due to the disease and treatment
- Cancer - risk is small
- Avascular necrosis – with decreased blood supply to the bone leading to tiny breaks and collapse
- Pregnancy complications - increased risk of miscarriage, high blood pressure during pregnancy (preeclampsia) and preterm birth. It is better to delay pregnancy until disease is under control for at least 6 mo

- Diagnosing is difficult as signs and symptoms vary and may overlap with other disorders.
- No single test
- **Lab tests** – CBC (look for anemia or low white count), sed rate, kidney and liver assessment, urinalysis, ANA (autoimmune process), blood slotting labs
- **Imaging tests** - chest X-ray, heart echocardiogram, MRI brain
- **Biopsy**
 - Kidney – to determine type of damage and correct treatments
 - Skin biopsy of a lesion

- **No Cure**
- **NSAIDs**—OTC or prescription for pain, swelling and fever. Side effects - stomach bleed, increased risk of heart/kidney problems
- **Antimalarial drugs** -hydroxychloroquine (Plaquenil) decreases risk of flares. Side effects - stomach upset; very rarely, retina damage
- **Corticosteroids.** Prednisone controls inflammation. Methylprednisolone-used to control kidneys and brain involvement. Many side effects.
- **Immunosuppressants** - azathioprine (Imuran), mycophenolate mofetil (CellCept), methotrexate. Side effects- infection, liver damage, decreased fertility, increased risk of cancer.
- **Biologics** - belimumab (Benlysta) I.V. Side effects -nausea, diarrhea and infections
- Rituximab (Rituxan) can be beneficial in cases of resistant lupus. Side effects – allergic reaction

MIXED CONNECTIVE TISSUE DISEASE

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- Autoimmune disease
- Women, under 50 are more affected
- Signs and symptoms of a combination of disorders - primarily lupus, scleroderma and polymyositis
- Sometimes referred to as an overlap disease
- Symptoms of the separate diseases usually don't appear all at once. Instead, they occur in sequence over a number of years, which can make diagnosis more complicated
- Early signs and symptoms often involve the hands with swelling and color changes
- In later stages, some organs — such as the lungs, heart and kidneys — may be affected
- No cure
- Steroids used - prednisone. Side effects

- Malaise
- Increased fatigue
- Mild fever.
- Raynaud's disease – fingers turn white and then purplish blue with cold or stress, and red with warming
- Swelling of fingers or hands
- Muscle pain
- Joint pain, swelling and deformities, similar RA
- Rash - red or reddish brown patches over the knuckles.

- Pulmonary hypertension - main cause of death in people with MCTD
- Interstitial lung disease - causes scarring in lungs, which affects ability to breathe
- Heart disease – enlarged heart or inflammation; cause of death in about 20 %
- Kidney damage - in ~25%. Can lead to kidney failure
- Digestive tract damage - abdominal pain and problems with digesting food
- Anemia – 75% have iron deficiency anemia
- Tissue necrosis - Raynaud's can cause gangrene in fingers
- Hearing loss - almost 50% of patients

RAYNAUD'S DISEASE/ SYNDROME

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- During an attack, with cold or stress, affected skin areas usually first turn white. Then, they may turn blue and feel cold and numb
- With warming, blood circulation improves and the areas may become swollen, tingly and red
- Most commonly involves fingers and toes
- Can also affect nose, lips, ears and even nipples
- Smaller arteries that provide blood to the skin become narrow (spasm). This decreases the blood circulation
- Women affected more than men
- More common in colder climates
- Treatment of Raynaud's disease depends on its severity and associated conditions
- Raynaud's disease is not disabling for most, but can affect the quality of life

- **Primary Raynaud's** - Raynaud's disease; most common and not associated with other condition. It is often mild and people ignore it
- **Secondary Raynaud's** - Raynaud's phenomenon and is caused by an underlying condition. Therefore, it is more serious.
- Symptoms of primary Raynaud's usually appear earlier; around age 40 in the secondary

Causes of secondary Raynaud's:

- Connective tissue diseases- scleroderma, lupus, RA and Sjogren's
- Diseases of the arteries – buildup of plaques or inflammation of the blood vessels (Buerger's disease)
- Carpal tunnel syndrome –pressure on the median nerve at the wrist. Presents with numbness, pain, weakness and susceptibility to cold
- Repetitive action or vibration – usually occupational
- Smoking - constricts blood vessels
- Injuries to the hands or feet - fracture, surgery, frostbite
- Meds - beta blockers (for high blood pressure); ergotamine and sumatriptan (for migraine), chemotherapy, OTC cold medications.

Complications

- Tissue damage in severe cases
- With complete blockage, sores (skin ulcers) or dead tissue (gangrene) can develop and require debridement or even amputation

Prevention

- Warm clothes, including socks, shoes and gloves
- Chemical hand warmers
- Earmuffs, face mask
- Warm car before use in cold weather
- Take precautions indoors. Wear socks. When taking food out of the refrigerator or freezer, wear gloves, mittens or oven mitts. winter.
- Air conditioning can trigger attacks!!!
- Use insulated drinking glasses

Diagnosis - Primary vs. secondary Raynaud's

- Nail fold capillaroscopy – to find under magnification capillary deformities at the base of the fingernail skin
- Blood tests, if suspected autoimmune/connective tissue disease - ANA, sed rate, scleroderma, Sjogren's antibodies. No single test
- Studies of arteries – ultrasound, angiogram

Treatment – Prevention + Treat the underlying disease or condition

Medication to dilate blood vessels and promote circulation

- Ca²⁺-channel blockers – decrease frequency and severity of attacks, help heal skin ulcers - nifedipine (Procardia), amlodipine (Norvasc), felodipine and others
- Vasodilators - nitroglycerin cream, BP med losartan (Cozaar), erectile dysfunction med sildenafil (Viagra), antidepressant fluoxetine (Prozac)

Surgeries and medical procedures

- Nerve surgery – cutting of sympathetic nerves to the hands and feet can block the exaggerated vasospasm responses
- Botox injections - blocks sympathetic nerves in affected limbs

- Systemic sclerosis (scleroderma)
- Sjogren syndrome
- Rheumatoid arthritis
- Lupus erythematosus
- Mixed connective tissue disease
- Raynaud syndrome

Thank you!
Questions?

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