Understanding CREST Syndrome



CREST syndrome is a type of scleroderma. It's sometimes called limited cutaneous scleroderma.

Scleroderma refers to a number of conditions that cause an abnormal buildup of collagen in the body. Too much of this protein makes your skin and other connective tissues thicken and harden. In some cases, it can also affect your organs, such as the lungs and heart.

CREST syndrome is an autoimmune disease. That means the immune system is not working the right way. It attacks itself. This rare health problem is more likely to happen in women. It often strikes people in their 30s to 50s.

What causes CREST syndrome?

Healthcare providers don't know the exact cause of CREST syndrome. It may be caused by genes or hormones. Some research suggests that a virus may trigger it. People exposed to radiation and certain chemicals, such as pesticides or solvents, have a higher risk for it.

Symptoms of CREST syndrome

CREST stands for its group of symptoms. These are calcinosis, Raynaud syndrome, esophageal dysmotility, sclerodactyly, and telangiectasia.

These symptoms tend to develop over time. You may first notice changes to your skin on your hands, legs, and face. You may also have problems swallowing. Symptoms may include:

- Calcinosis. Abnormal collection of calcium salts in or under the skin that form hard irregular lumps. These lumps can occur in any part of the body.
- Raynaud syndrome. This is a condition when arteries go into a spasm and limit blood flow. You may lose blood flow to your hands and feet when you are cold or under stress. This may cause your hands and feet to turn white and then blue. Color returns when they warm back up. As blood flow returns, skin turns red, tingles, burns, or may feel numb.
- Esophageal dysmotility. The extra collagen in your body may break down the muscles in your
 esophagus, the tube that links your mouth to your stomach. As a result, you may have acid reflux or
 problems swallowing.
- Sclerodactyly. As your skin thickens, it becomes stiff and shiny. You may find it hard to move your fingers. They can lock in a claw position. If the syndrome affects your face, you may have trouble opening your mouth.
- **Telangiectasia.** The blood vessels under your skin may swell and become expanded (dilated). They can cause small, red spots to form. These often show up on the face and lips.

You may also feel very tired. Some people describe this tiredness as similar to that in people getting cancer treatment. Or it's similar to the fatigue caused by lupus or rheumatoid arthritis.

Over time, CREST syndrome can cause problems with your internal organs, such as your heart and lungs. They may need to work harder. For instance, you may develop pulmonary hypertension. That's high blood pressure in the arteries that carry blood to your lungs.

Treatment for CREST syndrome

Your healthcare provider will talk about treatments with you. They'll need to find out how severe the diseases are. Then your provider will come up with treatment plan that's right for you.

CREST syndrome has no known cure. Treatments address easing symptoms and preventing complications. They may include:

- Moisturizing lotions. These may help with dry, stiff skin.
- · Exercise and physical therapy. Moving more may help ease the pain and stiffness in your joints.
- Antacids. These medicines can help ease acid reflux.
- Blood pressure pills. These can treat pulmonary hypertension.
- Calcium-channel blocking agents. These medicines can raise blood flow to your hands and feet to
 prevent Raynaud syndrome. Staying warm can also limit this symptom.
- Laser treatment. It may reduce the red spots caused by swollen blood vessels.

Possible complications of CREST syndrome

- Lung problems
- Liver disease
- Heart failure
- Kidney failure
- Dental problems

When to call your healthcare provider

Call your healthcare provider right away if you have any of these:

- Fever of 100.4°F (38°C) or higher, or as advised by your provider
- · Pain that gets worse
- Symptoms that don't get better, or get worse
- New symptoms

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