

Recognizing
The National
Bone and Joint Decade
2002–2011

*Questions
& Answers*
about . . .

Raynaud's
Phenomenon

*National Institute of Arthritis and Musculoskeletal and Skin Diseases (NIAMS)
National Institutes of Health
Public Health Service • U.S. Department of Health and Human Services*

For Your Information

This publication contains information about medications used to treat the health condition discussed here. When this booklet was printed, we included the most up-to-date (accurate) information available. Occasionally, new information on medication is released.

For updates and for any questions about any medications you are taking, please contact the U.S. Food and Drug Administration at 1-888-INFO-FDA (1-888-463-6332, a toll-free call) or visit their Web site at www.fda.gov.

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NIAMS/National Institutes of Health
1 AMS Circle
Bethesda, MD 20892-3675

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What Is Raynaud's Phenomenon?

Raynaud's phenomenon is a condition that affects the blood vessels in the extremities – generally, the fingers and toes. It is characterized by episodic attacks, called vasospastic attacks, in which the blood vessels in the digits (fingers and toes) constrict (narrow), usually in response to cold temperatures and/or emotional stress. When this condition occurs on its own, it is called primary Raynaud's phenomenon. When it occurs with another condition such as scleroderma or lupus, it is called secondary Raynaud's phenomenon.

Who Gets Raynaud's Phenomenon?

Although people of any age can have Raynaud's phenomenon, the primary form typically begins between the ages of 15 and 25. Women are more likely than men to have Raynaud's phenomenon. It appears to be more common in people who live in colder climates. This is likely true because people with the disorder have more Raynaud's attacks during periods of colder weather.

Although estimates vary, most studies show that Raynaud's phenomenon affects about 3 percent of the general population. For most, the symptoms are mild and not associated with any blood vessel or tissue damage.

Most people with Raynaud's phenomenon have the primary form, which is not associated with any underlying disease. In fact, in these individuals it is thought to be an exaggeration of normal responses to cold temperature and/or stress.

When Raynaud's phenomenon is caused by or associated with an underlying disease, it is referred to as secondary Raynaud's phenomenon. Secondary Raynaud's phenomenon tends to begin later in life than the primary form, typically after 35 to 40 years of age.

It is common for patients with a connective tissue disease to have Raynaud's phenomenon. It occurs in more than 90 percent of patients with scleroderma, and in about 30 percent of patients with systemic lupus erythematosus and with Sjögren's syndrome. Secondary Raynaud's phenomenon may also be associated with exposure to vibrating tools such as jackhammers, which cause trauma to the hands and wrists. And it may be linked to certain drugs, such as chemotherapy agents, or to chemicals such as vinyl chloride.

What Happens During an Attack?

Attacks of Raynaud's phenomenon are caused by an intensification of the body's natural response to cold. When a person is exposed to cold, the body's normal response is to slow the loss of heat and preserve its core temperature. Blood vessels in the surface of the skin are called thermoregulatory vessels because they react to changes in the ambient temperature. To maintain normal core temperature, these specialized blood vessels in the skin surface constrict and move blood from arteries near the surface to veins deeper in the body. But for people who have Raynaud's phenomenon, the thermoregulatory vessels overreact to cold exposure with sudden

and intense spasmodic contractions of these small blood vessels that supply blood to the skin of the fingers, toes, ears, face, and other body areas.

Once an attack begins, a person may experience three phases (though not all people have all three) of skin color changes – typically from white to blue to red – in the fingers or toes. Whiteness (called pallor) may occur in response to spasms of the arterioles (small branches of an artery) and the resulting collapse of the arteries supplying the fingers and toes. Blueness (cyanosis) may appear because the fingers or toes are not getting enough oxygen-rich blood. Finally, as the arterioles dilate (relax) and blood returns to the digits, redness (rubor) may occur.

During the attack, the fingers or toes may feel cold and numb as blood flow to them is interrupted. As the attack ends and blood flow returns, fingers or toes may throb and tingle. Typically, the blood flow to the skin will remain low until the skin is rewarmed. After warming, it usually takes 15 minutes to recover normal blood flow to the skin.

What Is the Difference Between Primary and Secondary Raynaud's Phenomenon?

In medical literature, primary Raynaud's phenomenon may also be called idiopathic Raynaud's phenomenon, primary Raynaud's syndrome, or Raynaud's disease. There is no known cause for primary Raynaud's phenomenon. It is more common than the secondary form and often is so

mild the patient never seeks medical attention. It generally is an annoyance that causes little disability. Secondary Raynaud's phenomenon is a more complex and serious disorder.

The most common cause of secondary Raynaud's phenomenon is connective tissue disease. The condition most commonly occurs with scleroderma or lupus, but is also associated with Sjögren's syndrome, dermatomyositis, and polymyositis. Some of these diseases reduce blood flow to the fingers and toes by causing blood vessel walls to thicken and the vessels to constrict too easily.

Other possible causes of secondary Raynaud's phenomenon are carpal tunnel syndrome and obstructive arterial disease (blood vessel disease). Some drugs are also linked to Raynaud's phenomenon. They include beta-blockers, such as Lopressor* or Cartrol, used to treat high blood pressure; ergotamine preparations, such as Cafergot or Wigrane, used for migraine headaches; certain agents used in cancer chemotherapy; and drugs, such as over-the-counter cold medication and narcotics, that cause vasoconstriction.

People in certain occupations may be more vulnerable to secondary Raynaud's phenomenon. Some workers in the plastics industry who are exposed to vinyl chloride, for example, develop a scleroderma-like illness, of which

* Brand names included in this fact sheet are provided as examples only, and their inclusion does not mean that these products are endorsed by the National Institutes of Health or any other Government agency. Also, if a particular brand name is not mentioned, this does not mean or imply that the product is unsatisfactory.

Raynaud's phenomenon can be a part. Workers who operate vibrating tools can develop a type of Raynaud's phenomenon called vibration-induced white finger.

Severe cases of Raynaud's phenomenon – usually of the secondary form – can lead to problems such as skin ulcers (sores) or gangrene (tissue death) in the fingers and toes, which can be painful and difficult to treat.

How Does a Doctor Diagnose Raynaud's Phenomenon?

Most doctors find it fairly easy to diagnose Raynaud's phenomenon but find it more difficult to identify the form of the disorder.

Physicians can now distinguish primary from secondary Raynaud's phenomenon with a complete history and physical examination. Sometimes, special blood testing is needed. If the evaluation and special testing studies are normal, then the diagnosis of primary Raynaud's phenomenon can be made and it is unlikely to change into a secondary form. Interestingly, about 30 percent of first-degree relatives of patients with primary Raynaud's phenomenon also have the condition. This finding suggests that primary Raynaud's phenomenon is determined by some yet-to-be discovered genetic trait.

A few tests can help the doctor distinguish between primary and secondary Raynaud's phenomenon. They are:

Nailfold capillaroscopy – During this test, the doctor puts a drop of oil on the patient's nailfolds, the skin at the base of the fingernail. The doctor then examines the nailfolds under a microscope to look for problems in the tiny blood vessels called capillaries. If the capillaries are enlarged or malformed, the patient may have a connective tissue disease.

Antinuclear antibody (ANA) test – In this blood test, the doctor determines whether the body is producing special proteins called antibodies that are directed against the nuclei of the body's cells. These abnormal antibodies are often found in people who have connective tissue diseases or other autoimmune disorders.

Erythrocyte sedimentation rate (ESR or sed rate) – This is a laboratory test for inflammation that measures how quickly red blood cells fall to the bottom of a test tube of unclotted blood. Rapidly descending cells (an elevated sed rate) indicate inflammation in the body.

What Is the Treatment for Raynaud's Phenomenon?

The aims of treatment are to reduce the number and severity of attacks and to prevent tissue damage and loss of tissue in the fingers and toes. Most doctors are conservative in treating patients with primary Raynaud's phenomenon because they do not get tissue damage. For these patients, doctors tend to

recommend nondrug treatments before moving onto medications. For patients with secondary Raynaud's phenomenon, medications are more often prescribed, because severe attacks with ulcers or tissue damage are more likely.

In the most severe cases, Raynaud's causes ulcers and serious tissue damage that does not respond to medications. Doctors may use a surgical procedure called a digital sympathectomy with adventitial stripping (which involves removing the tissue and nerves around the blood vessels supplying the affected digits). While this procedure may result in reducing symptoms and healing tissue, it only helps temporarily and therefore is reserved for difficult cases.

The most common treatments and self-help measures are described below.

Nondrug Treatments and Self-Help Measures

The following nondrug treatments and self-help measures can decrease the severity of Raynaud's attacks and promote overall well-being.

- **Take action during an attack.** You can decrease both its length and severity by a few simple actions. The first and most important one is avoid the cold. Warming the body and the hands or feet is also helpful. If you're outside and the weather is cold, go indoors. Run warm water over your fingers or toes or soak them in a bowl of warm water to warm them. If a

stressful situation triggers the attack, get out of the stressful situation, if possible, and relax. While biofeedback and similar nondrug methods are used, formal studies have suggested they are not helpful.

- **Keep warm.** It is important not only to keep the extremities warm but also to avoid chilling any part of the body. Remember, a drop in the body's core temperature triggers the attack. Shifting temperature (for example, rapidly moving from 90 degrees outside to a 70 degree air-conditioned room) and damp rainy weather are to be avoided. In cold weather, pay particular attention to dressing. Several layers of loose clothing, socks, hats, and gloves or mittens are recommended. A hat is important because a great deal of body heat is lost through the scalp. Keep feet warm and dry. Some people find it helpful to wear mittens and socks to bed during the winter. Chemical warmers, such as small heating pouches that can be placed in pockets, mittens, boots, or shoes, can give added protection during long periods outdoors.

People who have secondary Raynaud's phenomenon should talk to their doctors before exercising outdoors in cold weather.

In warm weather, be aware that air conditioning also can trigger attacks. Setting the thermostat for a higher temperature or wearing a sweater indoors can help prevent an attack. Some people find it helpful to use insulated drinking glasses and to put on gloves before handling frozen or refrigerated foods.

- **Do not smoke.** The nicotine in cigarettes causes the skin temperature to drop, which may lead to an attack.
- **Avoid aggravating medications** such as vasoconstrictors, which cause the blood vessels to narrow. Vasoconstrictors include beta-blockers, many cold preparations, caffeine, narcotics, some migraine headache medications, some chemotherapeutic drugs, and clonidine, a blood pressure medication. Some studies also associate the use of estrogen with Raynaud's phenomenon.
- **Control stress.** Because stress and emotional upsets may trigger an attack, particularly for people who have primary Raynaud's phenomenon, learning to recognize and avoid stressful situations may help control the number of attacks. Many people have found that relaxation can help decrease the number and severity of attacks. Local hospitals and other community organizations, such as schools, often offer programs in stress management.

- **Exercise regularly.** Many doctors encourage patients who have Raynaud’s phenomenon – particularly the primary form – to exercise regularly. Most people find that exercise promotes overall well-being, increases energy level, helps control weight, and promotes cardiovascular fitness and restful sleep. Patients with Raynaud’s phenomenon should talk to their doctors before starting an exercise program.
- **See a doctor.** People with Raynaud’s phenomenon should see their doctors if they are worried or frightened about attacks or if they have questions about caring for themselves. They should always see their doctors if episodes occur only on one side of the body (one hand or one foot) and any time one results in sores or ulcers on the fingers or toes.

Treatment with Medications

People with secondary Raynaud’s phenomenon are more likely than those with the primary form to be treated with medications. Many doctors believe that the most effective and safest drugs for Raynaud’s phenomenon are calcium-channel blockers such as nifedipine (Procardia) or amlodipine (Norvasc). These drugs, which are used to treat high blood pressure, work by relaxing the smooth muscle and dilating the small blood vessels. This decreases the frequency and severity of Raynaud’s attacks. These drugs can also help heal skin ulcers on the fingers or toes.

Some patients have found relief with alpha receptor blockers, which are high blood pressure medications such as prazosin (Minipres) or doxazosin (Cardura). These medications counteract the actions of norepinephrine, a hormone that constricts blood vessels. Effects are reported to be modest and side effects are associated with long-term use. However, preliminary research has found that a more highly targeted blocker for a specific alpha receptor shows promise. This receptor blocker is under investigation.

To help heal skin ulcers, some doctors prescribe a nonspecific vasodilator (drug that relaxes blood vessels) such as nitroglycerine paste, which is applied to the fingers. Many new agents that vasodilate are being used in cases that do not respond. These include the antidepressant fluoxetine (Prozac); phosphodiesterase inhibitors such as cilostazol (Pletal), pentoxifylline (Trental), and sildenafil (Viagra); and an angiotensin II receptor antagonist (used for blood pressure control), losartan (Cozaar). Patients should keep in mind that the treatment for Raynaud's phenomenon is not always successful. Often, patients with the secondary form will not respond as well to treatment as those with the primary form of the disorder. In cases of critical digital ischemia (where the blood flow will not return and finger loss may result), intravenous vasodilator therapy is used with prostaglandins such as epoprostenol (Flolan).

Patients may find that one drug works better than another. Some people may experience side effects that require stopping the medication. For other people, a drug may become less effective over time. Women of childbearing age should know that the medications used to treat Raynaud's phenomenon may affect the growing fetus. Therefore, women who are pregnant or who might become pregnant should avoid taking these medications if possible. Interestingly, Raynaud's phenomenon gets better or goes away during pregnancy.

Self-Help Reminders

- Take action during an attack.
- Keep warm.
- Don't smoke.
- Avoid aggravating medications.
- Control stress.
- Exercise regularly.
- See a doctor if questions or concerns develop.

What Research Is Being Conducted to Help People Who Have Raynaud's Phenomenon?

Researchers are studying techniques such as laser Doppler imaging to better diagnose Raynaud's phenomenon and to predict and monitor its course and responsiveness to treatment.

They are also evaluating the use of new treatments to improve blood flow for those who have Raynaud's phenomenon. These include the high blood pressure drug losartan (Cozaar); prostaglandins such as iloprost and alprostadil (Caverject, Edex); the male erectile dysfunction drug sildenafil (Viagra); the blood-clot-preventing drug ticlopidine (Ticlid); and the herbal remedy ginkgo biloba.

Treatments such as L-arginine, taken orally, have been studied as a way to reverse Raynaud's-related damage to tissue in the toes and fingers, but they have been found ineffective in most studies.

Basic investigators are studying the molecular mechanisms behind Raynaud's phenomenon, the anatomy of blood vessels, and possible genetic associations. Researchers in scleroderma and other connective tissue diseases are also investigating Raynaud's phenomenon in relation to these diseases.

Where Can People Find More Information About Raynaud's Phenomenon?

- **National Institute of Arthritis and Musculoskeletal and Skin Diseases Information Clearinghouse**
National Institutes of Health
1 AMS Circle
Bethesda, MD 20892-3675
Phone: 301-495-4484 or
877-22-NIAMS (226-4267) (free of charge)
TTY: 301-565-2966
Fax: 301-718-6366
www.niams.nih.gov

The clearinghouse provides information about various forms of arthritis and rheumatic diseases and bone, muscle, and skin diseases. It distributes patient and professional education materials and refers people to other sources of information. Additional information and updates can also be found on the NIAMS Web site.

- **National Heart, Lung, and Blood Institute**

National Institutes of Health

P.O. Box 30105

Bethesda, MD 20824–0105

Phone: 301–592–8573

Fax: 301–592–8563

E-mail: nhlbiinfo@nhlbi.nih.gov

www.nhlbi.nih.gov

The National Heart, Lung, and Blood Institute (NHLBI) provides leadership for a national program in diseases of the heart, blood vessels, lung, and blood; blood resources; and sleep disorders. The NHLBI Information Center develops and maintains information on numerous topics to respond to inquiries on specific diseases related to the heart, lungs, and blood.

- **American College of Rheumatology**

1800 Century Place, Suite 250

Atlanta, GA 30345

Phone: 404–633–3777

E-mail: acr@rheumatology.org

www.rheumatology.org

This professional organization of rheumatologists (doctors specializing in arthritis and related conditions such as Raynaud's phenomenon) and associated health professionals is involved in research, education, and patient care. It provides referrals to rheumatologists and other health professionals.

- **Arthritis Foundation**

P.O. Box 7669

Atlanta, GA 30357-0669

Phone: 404-872-7100 or 800-568-4045 (free of charge)
or call your local chapter (listed in the telephone directory)

www.arthritis.org

This is the main voluntary organization devoted to all forms of arthritis. The foundation offers free information about Raynaud's on its Web site and also provides physician referrals.

- **Lupus Foundation of America, Inc.**

2000 L Street, N.W., Suite 710

Washington, DC 20036

Phone: 202-349-1155 or 800-558-0121 (free of charge)

Fax: 202-349-1156

E-mail: lupusinfo@lupus.org

www.lupus.org

This is the main voluntary organization devoted to lupus. The LFA offers information and referral services, health fairs, newsletters, publications, seminars, support group meetings, hospital visits, and telephone help lines.

- **Scleroderma Foundation**

300 Rosewood Drive, Suite 105

Danvers, MA 01923

Phone: 800-722-HOPE (4673) (free of charge)

Fax: 978-463-5809

E-mail: sfinfo@scleroderma.org

www.scleroderma.org

This voluntary organization publishes information on scleroderma and funds research. It also offers patient education seminars, support groups, physician referrals, and information hotlines.

- **Scleroderma Research Foundation**

220 Montgomery Street, Suite 1411

San Francisco, CA 94104

Phone: 415-834-9444 or

800-441-CURE (2873) (free of charge)

Fax: 415-834-9177

E-mail: info@sclerodermaresearch.org

www.sclerodermaresearch.org

The foundation's goal is to find a cure for scleroderma by funding and facilitating the most promising, highest quality research and by placing the disease and its need for a cure in the public eye. The foundation distributes patient handbooks and a twice yearly, research-related newsletter.

- **Sjögren's Syndrome Foundation**
6707 Democracy Boulevard, Suite 325
Bethesda, MD 20817
Phone: 301-530-4420 or
800-475-6473 (free of charge)
Fax: 301-530-4415
www.sjogrens.org

This organization is devoted to Sjögren's syndrome. It publishes free pamphlets and a newsletter for members that provides up-to-date information. It also provides clinic and physician referrals.

Key Words

Adventitial stripping – a surgical treatment for severe Raynaud's phenomenon that involves removing the outer layer of the blood vessels supplying the affected extremities.

Antinuclear antibody (ANA) – an abnormal protein called an autoantibody that is directed against the nuclei in the body's cells. The presence of these autoantibodies in the body, particularly at high levels, often indicates a connective tissue disease.

Arteriole – a small branch of an artery (blood vessel that carries blood away from the heart) that leads into many smaller blood vessels near the skin's surface.

Beta-blocker – a type of medication used to treat high blood pressure and other cardiovascular problems. The medications work by blocking the effects of stress hormones on the body's beta receptors. This slows the nerve impulses that travel through the heart. As a result, your heart does not have to work as hard because it needs less blood and oxygen.

Biofeedback – a technique that uses electronic instruments to measure body functions and feed that information back to you, teaching you to bring involuntary body processes, such as blood pressure or skin temperature, under voluntary control.

Calcium-channel blocker – a type of medication that affects the movement of calcium within the cells of the heart and blood vessels, thus relaxing the blood vessels and increasing the supply of blood and oxygen to the heart.

Carpal tunnel syndrome – a condition in which the median nerve, which supplies the thumb side of the palm, becomes compressed in the space between the bones of the wrist through which the nerves and tendons run. It can cause tingling of the middle and index finger and weakness of the thumb.

Connective tissue disease – any of a group of diseases characterized by degeneration of collagen, a key component of connective tissues such as skin, muscles, tendons, and ligaments. Connective tissue diseases include scleroderma, lupus, polymyositis, and dermatomyositis.

Cyanosis – a bluish discoloration of the skin and mucous membranes caused by a lack of oxygen in the blood.

Dermatomyositis – an inflammatory disorder of the skin and underlying tissue, including the muscles.

Erythrocyte sedimentation rate (ESR or sed rate) – a laboratory test for inflammation that measures how quickly red blood cells fall to the bottom of a test tube of unclotted blood. Rapidly descending cells (an elevated sed rate) indicate inflammation in the body.

Gangrene – death and decay of part of the body caused by lack of blood supply.

Idiopathic – a word used to describe a disease for which the cause is not known.

Lupus – short for systemic lupus erythematosus, an inflammatory disease of the connective tissue, which can affect the skin, joints, and internal organs.

Nailfold capillaroscopy – a test used to look for abnormalities of the tiny blood vessels called capillaries in the skin at the base of the fingernails. Enlarged or malformed capillaries may indicate a connective tissue disease.

Vasodilator – a drug that relaxes blood vessels, making the cavities within the vessels larger and allowing more blood to pass through them.

Pallor – paleness of the skin due to reduced blood flow.

Polymyositis – a disease characterized by inflammation of the muscles, particularly those of the shoulders and hip girdles, which may become weak and tender to the touch.

Rubor – redness of tissue. In Raynaud's phenomenon, redness is caused by an increase in size of the small blood vessels of the affected extremities.

Sed rate – (see erythrocyte sedimentation rate).

Sjögren's syndrome – a condition in which the body's immune system attacks the moisture-producing glands, resulting in uncomfortable and sometimes damaging dryness of tissues, particularly those of the eyes and mouth.

Vasospastic attack – an episode of the small blood vessels going into spasms, temporarily reducing blood flow to the extremities.

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The mission of the National Institute of Arthritis and Musculoskeletal and Skin Diseases (NIAMS), a part of the Department of Health and Human Services' National Institutes of Health (NIH), is to support research into the causes, treatment, and prevention of arthritis and musculoskeletal and skin diseases; the training of basic and clinical scientists to carry out this research; and the dissemination of information on research progress in these diseases. The National Institute of Arthritis and Musculoskeletal and Skin Diseases Information Clearinghouse is a public service sponsored by the NIAMS that provides health information and information sources. Additional information can be found on the NIAMS Web site at www.niams.nih.gov.



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