

*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 in this booklet. 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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National Institute of Arthritis and Musculoskeletal
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1 AMS Circle
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You can also find this booklet on the NIAMS Web site at www.niams.nih.gov/hi/topics/raynaud/ar125fs.htm.

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What Is Raynaud's Phenomenon?

Raynaud's phenomenon is a disorder that affects the blood vessels in the fingers, toes, ears, and nose. This disorder is characterized by episodic attacks, called vasospastic attacks, that cause the blood vessels in the digits (fingers and toes) to constrict (narrow). Raynaud's phenomenon can occur on its own, or it can be secondary to another condition such as scleroderma or lupus.

Although estimates vary, recent surveys show that Raynaud's phenomenon may affect 5 to 10 percent of the general population in the United States. Women are more likely than men to have the disorder. Raynaud's phenomenon appears to be more common in people who live in colder climates. However, people with the disorder who live in milder climates may have more attacks during periods of colder weather.

What Happens During an Attack?

For most people, an attack is usually triggered by exposure to cold or emotional stress. In general, attacks affect the fingers or toes but may affect the nose, lips, or ear lobes.

Reduced Blood Supply to the Extremities

When a person is exposed to cold, the body's normal response is to slow the loss of heat and preserve its core temperature. To maintain this temperature, the blood vessels that control blood flow to the skin surface move blood from arteries near the surface to veins deeper in the body. For people who have Raynaud's phenomenon, this normal body response is intensified by the sudden spasmodic contractions of the small blood vessels (arterioles) that supply blood to the fingers and toes. The arteries of the fingers and toes may also collapse. As a result, the blood supply to the extremities is greatly decreased, causing a reaction that includes skin discoloration and other changes.

Changes in Skin Color and Sensation

Once the attack begins, a person may experience three phases of skin color changes (white, blue, and red) in the fingers or toes. The order of the changes of color is not the same for all people, and not everyone has all three colors. Pallor (whiteness) may occur in response to spasm of the arterioles and the resulting collapse of the digital arteries. Cyanosis (blueness) may appear because the fingers or toes are not getting enough oxygen-rich blood. The fingers or toes may also feel cold and numb. Finally, as the arterioles dilate (relax) and blood returns to the digits, rubor (redness) may occur. As the attack ends, throbbing and tingling may occur in the fingers and toes. An attack can last from less than a minute to several hours.

How Is Raynaud's Phenomenon Classified?

Doctors classify Raynaud's phenomenon as either the primary or the secondary form. In medical literature, "primary Raynaud's phenomenon" may also be called Raynaud's disease, idiopathic Raynaud's phenomenon, or primary Raynaud's syndrome. The terms idiopathic and primary both mean that the cause is unknown.

Primary Raynaud's Phenomenon

Most people who have Raynaud's phenomenon have the primary form (the milder version). A person who has primary Raynaud's phenomenon has no underlying disease or associated medical problems. More women than men are affected, and approximately 75 percent of all cases are diagnosed in women who are between 15 and 40 years old.

People who have only vasospastic attacks for several years, without involvement of other body systems or organs, rarely have or will develop a secondary disease (that is, a connective tissue disorder such as scleroderma) later. Several researchers who studied people who appeared to have primary Raynaud's phenomenon over long periods of time found that less than 9 percent of these people developed a secondary disease.

Secondary Raynaud's Phenomenon

Although secondary Raynaud's phenomenon is less common than the primary form, it is often a more complex and serious disorder. Secondary means that patients have an underlying disease or condition that causes Raynaud's phenomenon. Connective tissue diseases are the most common cause of secondary Raynaud's phenomenon. Some of these diseases reduce blood flow to the digits by causing blood vessel walls to thicken and the vessels to constrict too easily. Raynaud's phenomenon is seen in approximately 85 to 95 percent of patients with scleroderma and mixed connective tissue disease, and it is present in about one-third of patients with systemic lupus erythematosus. Raynaud's phenomenon also can occur in patients who have other connective tissue diseases, including Sjögren's syndrome, dermatomyositis, and polymyositis.

Possible causes of secondary Raynaud's phenomenon, other than connective tissue diseases, are carpal tunnel syndrome and obstructive arterial disease (blood vessel disease). Some drugs, including beta-blockers (used to treat high blood pressure), ergotamine preparations (used for migraine headaches), certain agents used in cancer chemotherapy, and drugs that cause vasoconstriction (such as some over-the-counter cold medications and narcotics), are linked to Raynaud's phenomenon.

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) develop a scleroderma-like illness, of which 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.

People with secondary Raynaud's phenomenon often experience associated medical problems. The more serious problems are skin ulcers (sores) or gangrene (tissue death) in the fingers or toes. Painful ulcers and gangrene are fairly common and can be difficult to treat. In addition, a person may experience heartburn or difficulty in swallowing. These two problems are caused by weakness in the muscle of the esophagus (the tube that takes food and liquids from the mouth to the stomach) that can occur in people with connective tissue diseases.

How Does a Doctor Diagnose Raynaud's Phenomenon?

If a doctor suspects Raynaud's phenomenon, he or she will ask the patient for a detailed medical history. The doctor will then examine the patient to rule out other medical problems. The patient might have a vasospastic attack during the office visit, which makes it easier for the doctor to diagnose Raynaud's phenomenon. Most doctors find it fairly easy to diagnose Raynaud's phenomenon but more difficult to identify the form of the disorder. (See the box on page 7 for the criteria doctors use to diagnose primary or secondary Raynaud's phenomenon.)

Nailfold capillaroscopy (study of capillaries under a microscope) can help the doctor distinguish between primary and secondary Raynaud's phenomenon. 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 abnormalities of the tiny blood vessels called capillaries. If the capillaries are enlarged or deformed, the patient may have a connective tissue disease.

The doctor may also order two particular blood tests, an antinuclear antibody test (ANA) and an erythrocyte sedimentation rate (ESR). The ANA test determines whether the body is producing special proteins (antibodies) often found in people who have connective tissue diseases or other autoimmune disorders. The ESR test is a measure of inflammation in the body and tests how fast red blood cells settle out of unclotted blood. Inflammation in the body causes an elevated ESR.

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 in the fingers and toes. Most doctors are conservative in treating patients with primary and secondary Raynaud's phenomenon; that is, they recommend nondrug treatments and self-help measures first. Doctors may prescribe medications for some patients, usually those with secondary Raynaud's phe-

Diagnostic Criteria for Raynaud's Phenomenon

Primary Raynaud's Phenomenon

- Periodic vasospastic attacks of pallor or cyanosis (some doctors include the additional criterion of the presence of these attacks for at least 2 years)
- Normal nailfold capillary pattern
- Negative antinuclear antibody test
- Normal erythrocyte sedimentation rate
- Absence of pitting scars or ulcers of the skin, or gangrene (tissue death) in the fingers or toes

Secondary Raynaud's Phenomenon

- Periodic vasospastic attacks of pallor and cyanosis
- Abnormal nailfold capillary pattern
- Positive antinuclear antibody test
- Abnormal erythrocyte sedimentation rate
- Presence of pitting scars or ulcers of the skin, or gangrene in the fingers or toes

nomenon. In addition, patients are treated for any underlying disease or condition that causes secondary Raynaud's phenomenon.

Nondrug Treatments and Self-Help Measures

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

- **Take action during an attack**—An attack should not be ignored. Its length and severity can be lessened by a few simple actions. The first and most important action is to warm the hands or feet. In cold weather, people should go indoors. Running warm water over the fingers or toes or soaking them in a bowl of warm water will warm them. Taking time to relax will further help to end the attack. If a stressful situation triggers the attack, a person can help stop the attack by getting out of the stressful situation and relaxing. People who are trained in biofeedback can use this technique along with warming the hands or feet in water to help lessen the attack.
- **Keep warm**—It is important not only to keep the extremities warm but also to avoid chilling any part of the body. In cold weather, people with Raynaud's phenomenon must 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. Feet should be kept dry and warm. Some people find it helpful to wear mittens and socks to bed during 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.

People with Raynaud's phenomenon should also be aware that air conditioning can trigger attacks. Turning down the air conditioning or wearing a sweater may help prevent attacks. Some people find it helpful to use insulated drinking glasses and to put on gloves before handling frozen or refrigerated foods.

- **Quit smoking**—The nicotine in cigarettes causes the skin temperature to drop, which may lead to an attack.
- **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 or biofeedback training can help decrease the number and severity of attacks. Biofeedback training teaches people to bring the temperature of their fingers under voluntary control. Local hospitals and other community organizations, such as schools, often offer programs in stress management.

- **Exercise**—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 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 attacks occur only on one side of the body (one hand or one foot) and any time an attack 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 are calcium-channel blockers, which relax smooth muscle and dilate the small blood vessels. These drugs decrease the frequency and severity of attacks in about two-thirds of patients who have primary and secondary Raynaud’s phenomenon. These drugs also can help heal skin ulcers on the fingers or toes.

Other patients have found relief with drugs called alpha blockers that counteract the actions of norepinephrine, a hormone that constricts blood vessels. Some doctors prescribe a nonspecific vasodilator (drug that relaxes blood vessels), such as nitroglycerine paste, which is applied to the fingers, to help heal skin ulcers. 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.

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

Self-Help Reminders

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

know that the medications used to treat Raynaud's phenomenon may affect the growing fetus. Therefore, women who are pregnant or are trying to become pregnant should avoid taking these medications if possible.

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

Researchers are studying ways to better diagnose Raynaud's phenomenon and predict and monitor its course and association with other diseases. They are also evaluating the use of new drugs to improve blood flow in Raynaud's phenomenon; for example, the prostaglandins iloprost and alprostadil, the high blood pressure drug losartan, and a substance applied to the skin that generates the blood vessel dilating gas nitric oxide. Basic investigators are studying the molecular mechanisms behind Raynaud's phenomenon, the anatomy of blood vessels, and possible genetic associations. Some researchers are studying the use of biofeedback to control attacks. Researchers in scleroderma and other connective tissue diseases are also investigating Raynaud's phenomenon in relation to these diseases.

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

- **National Institute of Arthritis and Musculoskeletal and Skin Diseases (NIAMS)**

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

E-mail: NIAMSInfo@mail.nih.gov

www.niams.nih.gov

The clearinghouse provides information about various forms of arthritis and rheumatic disease 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
Information Center**

National Institutes of Health

P.O. Box 30105

Bethesda, MD 20824-0105

Phone: 301-592-8573

Fax: 301-592-8563

E-mail: NHLBIinfo@rover.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**

1330 West Peachtree Street, Suite 100

Atlanta, GA 30309

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 publishes a free pamphlet on Raynaud's phenomenon and also provides physician referrals.

- **Lupus Foundation of America**

2000 L Street, N.W., Suite 710

Washington, DC 20036

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

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)

E-mail: sfinfo@scleroderma.org

www.scleroderma.org

This voluntary organization publishes information on scleroderma, including a booklet for scleroderma patients with Raynaud's phenomenon. It also offers patient education seminars, support groups, physician referrals, and information hotlines.

- **Sjögren's Syndrome Foundation**

8120 Woodmont Avenue, Suite 530

Bethesda, MD 20814-2771

Phone: 301-718-0300 or 800-475-6473 (free of charge)

Fax: 301-718-0322

E-mail: ssf@sjogrens.org

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 provides clinic and physician referrals.

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