RAYNAUD'S DISEASE

What Is Raynaud's Disease?

Raynaud's disease and Raynaud's phenomenon are rare disorders that affect blood vessels. These disorders are marked by brief episodes of vasospasm (narrowing of the blood vessels). Vasospasm causes decreased blood flow to the fingers and toes, and rarely to the nose, ears, nipples, and lips. The fingers are the most commonly affected area, but the toes also are affected in 40 percent of people with Raynaud's.

When this disorder occurs without any known cause, it is called Raynaud's disease, or primary Raynaud's. When the condition occurs along with a likely cause, it is known as Raynaud's phenomenon, or secondary Raynaud's. Primary Raynaud's is more common and tends to be less severe than secondary Raynaud's.

When you have primary or secondary Raynaud's, cold temperatures or stressful emotions can trigger attacks. During these attacks, there is a brief lack of blood flow to the affected body part(s), and the skin can temporarily become white then bluish. As blood flow returns to the area, the skin turns red. The affected areas can throb or feel numb and tingly. With severe Raynaud's, prolonged or repeated episodes can cause sores or tissue death (gangrene).

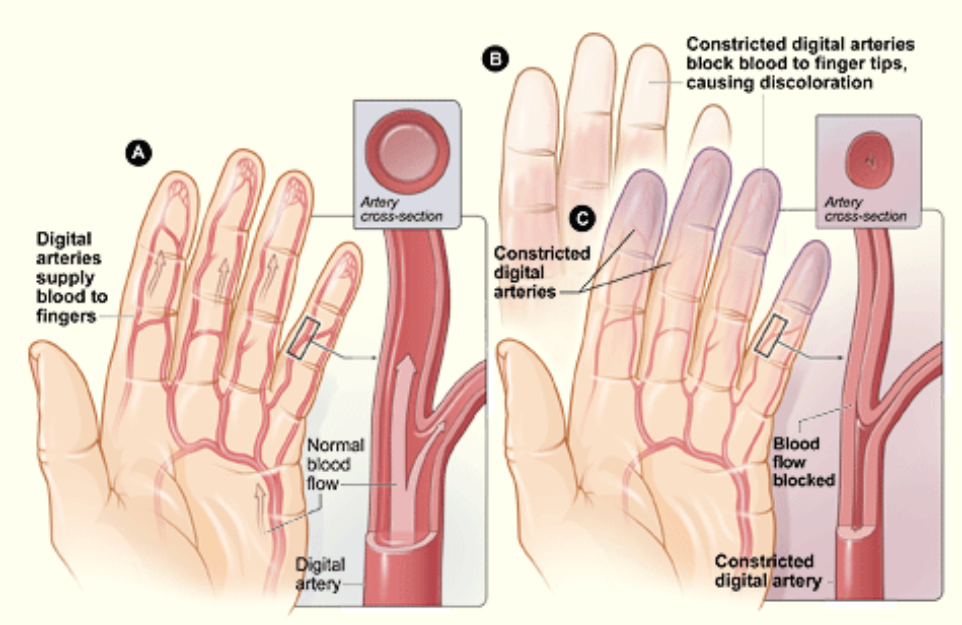

Figure A shows the normal digital arteries with normal blood flow to the fingers. The inset images show cross-sections of a normal artery. Figure B shows white discoloration of the fingertips caused by blocked blood flow. Figure C shows narrowed digital arteries, causing blocked blood flow and purple discoloration of the fingertips. The inset images show cross-sections of a narrowed artery blocking the flow of blood.

It is normal for the body to keep its vital inner organs warm by limiting blood flow to the arms, legs, fingers, and toes. The body naturally does this in response to a long period of cold. This response can cause frostbite. In people with Raynaud's, the response to cold is quicker and stronger. The response can be triggered by mild or short-lived changes in temperature, such as:

- Taking something out of the freezer
- Temperatures that dip below 60 degrees Fahrenheit

In people with Raynaud's, blood flow is more strongly reduced in response to cold temperatures than in people without the disorder. When Raynaud's is severe (which is uncommon), exposure to cold for as little as 20 minutes can cause major tissue damage.

The blood vessels of people with Raynaud's also physically overreact to stressful emotions. It is normal during times of psychological stress for the body to release hormones that narrow its blood vessels. But for people with Raynaud's, this squeezing of blood vessels is stronger. This results in less blood reaching fingers, toes, and sometimes other extremities.

Outlook

For most people, primary Raynaud's is more of a bother than a serious illness and it can usually be managed with minor lifestyle changes. Secondary Raynaud's can be more difficult to manage, but several treatments may help prevent or relieve symptoms. Among the most important treatments for secondary Raynaud's is treating of the underlying condition.

What Causes Raynaud's Disease?

In most cases of Raynaud's, no cause can be found. When this happens, the disorder is called primary Raynaud's. When a cause can be found, the

disorder is called secondary Raynaud's. Secondary Raynaud's can be linked to many different medical or workplace conditions, such as:

- Diseases that damage blood vessels or the nerves that control the function of blood vessels in the hands and feet
- Repetitive actions that damage the nerves that control blood vessels in the hands and feet
- Exposure to certain chemicals
- Use of medicines that narrow blood vessels or affect blood pressure

Diseases

Secondary Raynaud's is especially common in people who have scleroderma or systemic lupus erythematosus. About 9 out of 10 people with scleroderma have Raynaud's. About 1 out of 3 people with lupus have Raynaud's. The disease also is linked to other diseases that damage blood vessels or nerves, including:

- Rheumatoid arthritis
- Atherosclerosis
- Sjogren's syndrome, dermatomyositis, and polymyositis
- Buerger's disease

Raynaud's also can be associated with thyroid problems and pulmonary hypertension (high blood pressure in the arteries of the lungs).

Repetitive Actions

Typing, playing the piano, or another repetitive action done for long periods of time is often linked to secondary Raynaud's. Using vibrating tools at work also can make you more likely to develop the disorder.

Chemicals

Exposure to certain chemicals at work can cause a scleroderma-like illness that is linked to Raynaud's. Nicotine in cigarette smoke also can make you more prone to developing Raynaud's.

Medicines
Several medicines are linked to secondary Raynaud's, including:

- Migraine headache medicines that contain ergotamine
- Certain cancer medicines
- Some over-the-counter cold or allergy remedies
- Some blood pressure medicines (beta blockers)

Injuries to the hands or feet from surgery, frostbite, or other causes also can lead to secondary Raynaud's.

Who Is At Risk for Raynaud's Disease?
About 3 out of 4 cases of primary Raynaud's occur in women between the ages of 15 and 40 years, although the reason is unknown. People in colder climates are also more likely to develop Raynaud's than people in warmer areas.

What Are the Signs and Symptoms of Raynaud's Disease?
People with Raynaud's (primary or secondary) have attacks in response to cold or emotional stress. The attacks can affect the fingers and toes, and rarely the nose, ears, nipples, or lips. The affected body parts will usually have two or more of the following changes:

- Look pale due to lack of blood flow
- Look bluish due to a lack of oxygen
- Feel numb, cold, or painful
- Redden and throb or tingle as blood returns to the affected area

Attacks usually last about 15 minutes. They can last less than a minute or as long as several hours. Attacks can occur daily or weekly.

Sometimes attacks affect only one or two fingers or toes. Different areas may be affected at different times. Attacks can cause sores or tissue death (gangrene) in people with severe secondary Raynaud's. However, severe Raynaud's is very uncommon.

How Is Raynaud's Disease Diagnosed?

Raynaud's is usually diagnosed based on a patient's history of experiencing color changes in their extremities in response to exposure to cold or emotional stress. A cold simulation test also may be used to provoke symptoms for the doctor to see. Doctors use the patient's history of symptoms, a physical exam, and diagnostic tests to rule out other conditions that might act like Raynaud's.

Specialists Involved

A rheumatologist (roo-ma-TOL-o-jist) is a doctor who specializes in treating disorders of the joints, bones, and muscles. Often, a rheumatologist diagnoses and treats patients with Raynaud's, but internists and family practice doctors also are able to diagnose and treat Raynaud's.

Diagnostic Tests and Procedures

To help diagnose Raynaud's, doctors may do a cold simulation test. This test can trigger an attack that is typical of the disorder. They also may do tests for inflammatory disorders that damage blood vessels or nerves. Having these conditions along with Raynaud's symptoms makes it more likely that a patient has secondary Raynaud's disease.

In the cold simulation test, temperature sensors are taped to the fingers of the hand. The hand is then briefly exposed to the cold, usually by dunking it in ice water. If the patient likely has Raynaud's, it will take more than the normal time for the finger temperature to return to what it was at the start of the test.

Doctors also may do a test called a nailfold capillaroscopy (KAP-i-lar-OS-ko-pe). For this test, the doctor puts a drop of oil on the skin at the base of the fingernail and then looks at it under a microscope. If the doctor sees abnormal looking blood vessels, this suggests an inflammatory disorder such as scleroderma.

Doctors also can use two specific blood tests to look for inflammatory conditions: the antinuclear antibody test and the erythrocyte sedimentation rate.

How Is Raynaud's Disease Treated?

There is no cure for primary or secondary Raynaud's, but many measures can reduce the number or intensity of attacks, including:

- Lifestyle changes
- Medicines
- Treatments for the disease or condition that may help cause secondary Raynaud's
- Surgery for the tissue damage that some people with secondary Raynaud's develop

In most people with primary Raynaud's, the disorder is successfully managed with lifestyle adjustments. Patients with secondary Raynaud's may need medicines in addition to lifestyle changes, and in rare cases, they may need surgery. Anyone with Raynaud's who develops sores on their fingers or toes or elsewhere on their body should see a doctor right away to prevent tissue loss.

Lifestyle Changes

Most of the lifestyle changes that help people with Raynaud's aim to avoid the triggers of attacks. These triggers include cold, emotional stress, and certain medicines, chemicals, or actions. To protect the body from cold, people can:

- Wear a hat, gloves, scarf, and a coat with snug cuffs during cold weather.
- Wear gloves or mittens when taking food out of the refrigerator or freezer.
- Turn down air conditioning, or dress warmly while in an air conditioned space.
- Warm up the car before driving in cold weather.
- To avoid emotional triggers, people can steer clear of stressful situations if possible. Relaxation techniques also can be helpful under stress.

To avoid workplace or recreational triggers, people can:

- Limit use of vibrating tools.
- Wear proper protective gear if they work with industrial chemicals.

- Limit frequent and repeated actions of the hands, such as typing or playing the piano.

A number of medicines can trigger attacks. People with Raynaud's should avoid:

- Beta blockers
- Over-the-counter cold or allergy remedies or diet aids that narrow blood vessels
- Birth control pills, which affect blood flow
- Headache medicines that contain ergotamine

Other helpful lifestyle changes for people with Raynaud's are those that boost blood flow in the body. These include exercising regularly and quitting smoking.

When attacks do occur, people with Raynaud's can take several steps to limit the length and strength of the attacks. These steps include:

- Moving to a warmer spot, such as indoors during cold weather.
- Warming the hands or feet. Hands can be placed under the armpits, and feet or hands can be soaked in warm water.
- Wiggling or massaging the fingers and toes.
- Moving the arms in circles or shaking arms or feet.
- Relaxing and getting out of stressful situations that trigger the attacks.

Medicines and Surgery

Most of the medicines used to treat people with Raynaud's are given to improve blood flow to the extremities. These medicines include calcium channel-blockers, such as:

- Nifedipine
- Amlodipine
- Diltiazem
- Felodipine
- Isradipine

Calcium channel-blockers help limit the number and severity of attacks in about 2 out of 3 patients with Raynaud's.

Also helpful are alpha-blockers, such as prazosin and doxazosin. In addition, skin creams that dilate blood vessels, such as nitroglycerine paste, can help heal skin sores.

The rare patient who develops sores or tissue death (gangrene) needs more aggressive treatment. Such treatment includes antibiotics and surgery to cut out damaged tissue. People with severe, worsening Raynaud's may have surgery or shots to block the action of nerves in the hands and feet that control blood flow in the skin. This surgery often gets rid of symptoms for 1–2 years. Patients may need shots more than once.

Living With Raynaud's Disease

Primary and secondary Raynaud's are conditions that may be lifelong. Most people with primary Raynaud's respond to simple lifestyle changes or medicines. Such treatment is not always as successful in secondary Raynaud's. If you have secondary Raynaud's, you may find that in time your medicines are less effective and your attacks become more frequent and/or more severe. Switching to a new treatment may help relieve or prevent your symptoms. Be sure to seek a doctor's care if you develop sores on your fingers or toes or elsewhere on the body.

Key Points

- Raynaud's is a rare disorder that affects blood flow to the fingers and toes, and rarely affects other areas such as the nose, ears, nipples, and lips.
- When Raynaud's occurs without any known cause, it is called primary Raynaud's. When a cause can be identified, the disease is called secondary Raynaud's.
- People with Raynaud's have attacks in response to cold or emotional stress. During these attacks, their fingers, toes, or other extremities temporarily pale and/or become bluish due to a lack of blood flow. As blood flow returns, the area turns red. The affected areas also may throb or feel cold, numb, or tingly.
- Raynaud's is usually diagnosed based on a history of symptoms, the exclusion of other conditions, and certain diagnostic tests.
- Common causes of secondary Raynaud's include other disorders, medicines, chemicals, or work conditions that affect blood flow to the extremities.

- Primary Raynaud's often can be managed with minor lifestyle changes. Secondary Raynaud's may require changing or stopping certain medicines, treating any underlying conditions, and having surgery (in severe cases).
- People with severe secondary Raynaud's may develop sores or tissue death (gangrene) in the extremities. This is uncommon.

Additional Information

- Vascular Disease Foundation - http://www.vdf.org/diseaseinfo/raynaud/
- Raynaud's Phenomenon - http://www.cedars-sinai.edu/5623.html

Source: NIH
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