



PXE Guide

Vascular

What Is PXE?

Pseudoxanthoma elasticum (PXE) is an inherited condition that causes abnormal mineralization in some elastic tissues of the body. Mineralization means that tiny deposits of calcium and other minerals build up where they do not usually belong.

PXE most often affects the skin, eyes, and mid-sized arteries. Less commonly, it may affect the gastrointestinal system. PXE was first recognized more than 100 years ago, and much has been learned about it in recent decades.

How Can PXE Affect the Arteries?

PXE can affect some of the body's arteries, especially peripheral medium-sized arteries. Arteries are the blood vessels that carry blood from the heart to the rest of the body. Peripheral arteries are the arteries outside the heart and brain, especially those that carry blood to the arms and legs.

In PXE, tiny mineral deposits can build up in the elastic part of these artery walls. Over time, this may make some arteries less flexible or narrower. When blood flow to the legs is reduced, a person may have cramping, aching, or pain with walking. This is called intermittent claudication.

This process is different from ordinary cholesterol plaque, although both can affect blood flow. For that reason, it is important for people with PXE to protect their overall vascular health.

What Symptoms Can Occur?

Many people with PXE do not have serious artery symptoms, especially in childhood or young adulthood. When vascular symptoms do occur, they may include:

- Weaker pulses in the wrists, ankles, or feet after walking or moving for a short time

- Cramping, aching, or pain in the legs during walking or exercise
- Symptoms that improve with rest and return with activity
- Coldness, numbness, or fatigue in the legs or feet
- Pain is not present when resting or in bed (this is not restless leg syndrome)

Pain or cramping in the legs during walking is called intermittent claudication. It happens when muscles need more blood during activity, but narrowed arteries cannot deliver enough blood. Typically, the pain decreases if a person perseveres and keeps walking despite the discomfort.

Blood Pressure, Cholesterol, and Smoking

People with PXE should pay close attention to common risk factors for artery disease. This isn't because PXE directly causes these factors, but because individuals should maintain the healthiest possible baseline health. These include:

- High blood pressure
- High cholesterol
- Diabetes
- Smoking or vaping
- Lack of physical activity
- Excess weight
- Family history of early heart or vascular disease

These risk factors do not cause PXE, but they may place additional stress on arteries. Any effect of PXE on top of these factors makes everything more difficult. Blood pressure and cholesterol should be monitored and treated according to standard medical guidance.

Smoking is especially important to avoid. Smoking damages blood vessels and increases the risk of vascular disease. Individuals who smoke experience far worse PXE symptoms.

Maintaining a weight that supports health can help reduce strain on the heart and arteries. This does not mean aiming for a particular appearance or relying only on BMI. A healthcare professional can help assess vascular risk using blood pressure, cholesterol, blood sugar, activity level, family history, and overall health.

What Kind of Medical Care Is Useful?

A primary care physician can help monitor vascular health over time. This may include:

- Checking blood pressure
- Checking cholesterol and other cardiovascular risk factors

- Tell them to check leg pain, walking distance, chest pain, or abdominal pain with meals
- Tell them to check pulses in the arms and legs
- Ask for a referral to a vascular specialist IF symptoms develop

Not every person with PXE needs extensive testing. Testing should be guided by symptoms, examination findings, age, and individual medical history.

A vascular specialist may recommend studies such as ankle-brachial index testing, Doppler ultrasound, or other imaging if symptoms suggest reduced blood flow.

When to Seek Prompt Medical Care

Most of these symptoms are not related to PXE, but you seek medical care promptly if you have:

- New chest pain, pressure, or shortness of breath
- Symptoms of stroke, such as face drooping, arm weakness, trouble speaking, or sudden confusion
- Sudden severe pain, coldness, or color change in an arm or leg
- New severe leg pain at rest
- Wounds on the feet or legs that do not heal
- Severe abdominal pain after eating, especially with weight loss or fear of eating

These symptoms can occur for many reasons and are not always related to PXE, but they should be evaluated.

Exercise and Activity

Regular physical activity is useful for general health and vascular health. Walking, cycling, swimming, strength training with proper technique, and other forms of movement may be appropriate for the individual.

If a person has leg pain with walking, supervised exercise or a structured walking plan may help, but this should be discussed with a healthcare professional.

People with PXE do not need to avoid weight lifting or resistance training simply because they have PXE. Strength training can be an important part of overall health, including vascular health, muscle strength, balance, and mobility.

The main caution is to avoid breath-holding and extreme straining, sometimes called the Valsalva maneuver. This can happen when someone holds their breath while lifting a very

heavy weight. Instead, people with PXE should learn good technique: breathe steadily, use appropriate weights, increase gradually, and stop if they develop pain, dizziness, chest symptoms, or vision changes.

For most people with PXE, the goal is not to avoid exercise. The goal is to exercise intelligently, protect the eyes from direct trauma, and discuss individual risks with a healthcare professional when there is active eye disease, recent retinal bleeding, significant vascular symptoms, or other medical concerns.

Medications

No approved medication reverses PXE-related artery mineralization or is specific to PXE. There are medications for intermittent claudication that can be effective in PXE.

Some people with PXE may need standard treatments for high blood pressure, high cholesterol, diabetes, peripheral artery disease, or heart disease. These decisions should be made with a healthcare professional.

People with PXE should discuss aspirin and non-steroidal anti-inflammatory drugs, such as ibuprofen and naproxen, with their physician before regular use. These medicines can increase bleeding risk, especially in the stomach or intestines. Sometimes these medicines are medically necessary, but the risks and benefits should be considered.

Gastrointestinal Bleeding

PXE is rarely be associated with bleeding in the stomach or intestines. This is uncommon, but it can be serious.

Seek urgent medical care if you vomit blood, pass black tarry stools, or have unexplained severe weakness, dizziness, or anemia. See our gastrointestinal guide.

PXE and Children

Circulation problems are uncommon in children with PXE. Children do not need intensive vascular testing unless they have symptoms or another medical reason for evaluation. Sometimes children are misdiagnosed and could have Generalized Arterial Calcification of Infancy (GACI) instead. These children should be seen by someone familiar with GACI. (Contact PXE International for referrals.)

For children, the most important steps are healthy habits, regular primary care, and attention to symptoms if they arise. Encourage ordinary physical activity, play, and sports with appropriate eye protection when needed.

PXE should not become the center of a child’s daily life. The same habits that support health in any child—movement, good nutrition, avoiding smoking exposure, and regular medical care—also support vascular health in PXE.

Key Points

- PXE can affect medium-sized arteries.
- Tiny mineral deposits can build up in the elastic part of the artery wall.
- Some people develop reduced pulses or leg pain with walking, called intermittent claudication.
- Blood pressure, cholesterol, diabetes, smoking, and other vascular risk factors should be managed carefully.
- Regular physical activity is generally important and should not be avoided unnecessarily.
- Testing should be guided by symptoms, examination findings, and medical history.
- Seek prompt care for chest pain, stroke symptoms, severe limb pain, non-healing wounds, or black tarry stools.
- Serious circulation problems are uncommon in children with PXE.

PXE International

PXE International provides information, support, and research opportunities for individuals and families affected by pseudoxanthoma elasticum.

Email: info@pxe.org • Phone: 202.362.9599 • Website: pxe.org • © 1995–2026 PXE International.

Except where otherwise noted, educational content in this guide is licensed under a Creative Commons Attribution-NonCommercial-ShareAlike 4.0 International License.