



PAIN IN FABRY DISEASE

A common—but invisible—burden

- Thousands of people around the world live with Fabry disease, but because its symptoms can vary significantly, no two people experience it in exactly the same way. And, as with many other chronic diseases, some of the symptoms of Fabry disease are “invisible”—meaning that they are not directly apparent to others and may be difficult to measure with medical tests.
- Pain is one of those symptoms that’s invisible to others. It’s also one of the earliest symptoms of Fabry disease (it often begins in early childhood) and one of the most common: pain affects about three out of four people who live with Fabry disease.^{1,2}
- Because pain is “invisible” and may be hard to measure, it can be hard to discuss or explain to others, even medical professionals. This can cause a lot of frustration, and it can even damage trust between people with Fabry disease and their health-care providers (HCPs).^{3,4}
- **Even though most people who have Fabry disease have pain, their experiences may be very different. This fact sheet describes some of the different ways that people with Fabry disease experience pain, and provides information and ideas designed to help those who live with Fabry disease have more productive discussions with HCPs about pain.**



What does Fabry pain feel like?

→ Burning. Stabbing. Tingling. These are some of the most common ways that people who have Fabry disease describe their pain.^{2,5} Others may describe their pain as pressing, prickling, squeezing, electrifying or feeling sore.⁵ People who have Fabry disease experience more than one type of pain, and they may have different types of pain at different times, or may sometimes have several types at once.⁵

Researchers have described four main patterns of pain associated with Fabry disease:⁵

PAIN ATTACKS

Sudden, intense pain that does not last very long

Experienced by almost 47% of people with Fabry disease who have pain

PAIN CRISIS

Intense pain that can last for hours or even days

Experienced by more than 17% of people with Fabry disease who have pain

EVOKED PAIN

Pain caused by something that would normally cause no pain or only slight pain, like a light touch or hot water

Experienced by 66% of people with Fabry disease who have pain

CHRONIC PAIN

Pain that is always present

Experienced by more than 17% of people with Fabry disease who have pain

→ How often people with Fabry disease experience pain also varies: most report experiencing pain one to four times a month, but some experience pain daily, weekly, once a month or only a few times a year.^{5,6} This also may change over time: for example, some people find that their pain becomes less severe with age. This may be because the longer someone has Fabry disease, the more damage may be done to their nerves, which can make them less able to feel pain.^{3,6}

What makes pain happen?

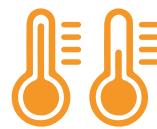
Pain attacks and pain crises can be set off—or “triggered”—by a number of things in everyday life. These may include:^{2,5,7}



fever and/or acute illness (such as the flu)



emotional stress



high or low temperatures or rapid temperature changes



exercise



fatigue

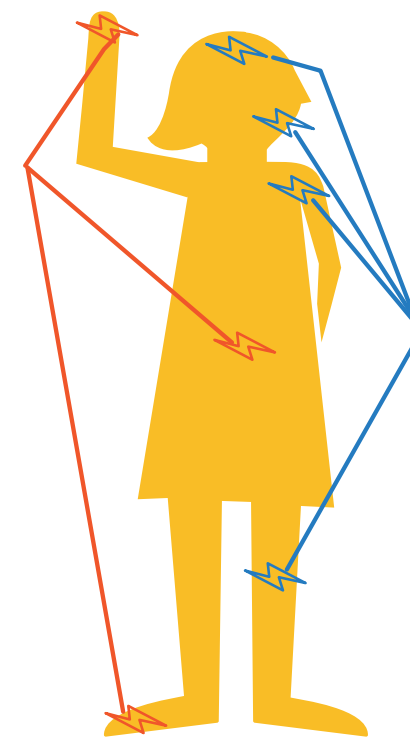


certain foods, such as coffee and alcohol

Where do people with Fabry feel pain?

The location of pain is another thing that can vary among people who have Fabry disease. Pain in the hands and/or feet affects almost 85% of people who have pain associated with Fabry disease (this type of pain is sometimes called acroparesthesia).^{2,6} But people with Fabry disease also report having pain in many other places—sometimes all over their bodies.^{2,5-7}

Pain in the **hands and feet** and **abdominal pain** are very common in people with Fabry disease



However, some people also report pain in other locations, including the **head, teeth, shoulders and joints**

Pain can hurt in many different ways



→ More than just hurting physically, pain may affect the well-being of people with Fabry disease in other important ways. Research has shown that pain can have a negative effect on almost every aspect of life in people with Fabry disease, including mood, relationships, work, sleep and life enjoyment, as well as the ability to walk and engage in ordinary daily activities. Unsurprisingly, the greater the frequency and intensity of the pain, the more negative its effects tend to be.⁶



→ People who are experiencing significant amounts of pain might need to limit physical activities, or limit daily activities such as working, socializing or even going to school.^{1,7} Pain can also be so bad and so constant that it has significant effects on mood and state of mind: about half of those who have Fabry disease experience depression, and almost 40% experience anxiety.⁶⁻⁹

What can be done to help manage pain?



---> Paying attention to what triggers pain—and avoiding those triggers—can be helpful in managing pain.¹⁰ For example, a person who notices that getting overheated worsens their pain could avoid hot environments or wear a cooling vest. Or, someone who finds that fatigue or too much exercise triggers pain could try scheduling their school, work or social activities differently to help avoid overexertion. Keeping a simple pain diary to note changes in pain levels and possible triggers may be useful.



---> HCPs can provide medications and pain management plans that are customized to individual patients' needs.^{10,11} And the more they know about the pain their patients are experiencing, the more they may be able to help. This is why one of the most important things people with Fabry disease can do to help manage their pain is to tell HCPs exactly *how, when, and where* they are experiencing pain.



---> To help people with Fabry disease keep track of and describe their own pain, researchers have developed a self-administered evaluation tool called the Fabry Pain Questionnaire. Information about this questionnaire was published in an article in *Orphanet Journal of Rare Diseases*, available online at <https://www.ncbi.nlm.nih.gov/pmc/articles/PMC4573689/>.¹² (A link to the questionnaire itself is provided at the end of the article). Reading this article and questionnaire may help people with Fabry disease learn how to describe their pain to HCPs more clearly and completely. And remember—better communication with HCPs can contribute to better care.

---> **To find out more about Fabry disease and the Fabry community, talk to an HCP, and consider visiting fabrynetwork.org/members to find a local patient organization.**

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