



Angioedema

Frequently Asked Questions

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Q 1: What is angioedema?

Angioedema is a condition where small blood vessels leak fluid into the tissues under the skin, causing swelling in different parts of the body. It can develop because of an allergic reaction and is usually accompanied by hives (urticaria). Up to 20% of people will develop hives at some time in their life, and around one in three of these will have angioedema as well. It is possible to develop angioedema without also developing hives.

Although the swelling can be quite severe, it is temporary. Angioedema is not usually caused by a serious underlying disease and is not likely to cause illness or damage to vital organs such as kidneys, liver, or lungs. There is no known cure, but with the right diagnosis and management with appropriate medication, it can usually be prevented.

Q 2: What does angioedema look like?

The signs and symptoms of angioedema are not always the same.

Angioedema with hives (urticaria)

- Pink or red itchy rashes, that may appear as blotches or raised red lumps (wheals) on the body with swelling under the skin that feels itchy, hot, tingly, or burning.

Angioedema without hives

- Large swollen areas under the skin, that look red and are itchy, hot, tingly, burning or generally uncomfortable.
- In some people, skin-coloured swellings, that are not itchy, red, or uncomfortable may appear.

Symptoms are temporary in most people but can reappear due to infection, stress, or unknown reasons.

Q 3: Where does angioedema occur?

The most common areas of the body affected by angioedema are the face, lips, tongue, throat, and genital areas. Swelling of other organs like the oesophagus (muscular tube that takes food to the stomach), abdomen (stomach area), or bowel can sometimes cause pain in the chest. Swelling can sometimes develop over and around joints which can be painful and last for many days.

The swelling from angioedema usually resolves in a few days. Swelling lasting longer than this may be due to another cause and should be seen by a doctor.

Q 4: What causes angioedema?

Infection

A viral infection is usually the most common cause of hives (urticaria) and angioedema in children, especially if they last for more than 24 hours.

Food or drug allergy

Swellings due to allergic reactions to foods or drugs are sometimes severe and dramatic, but usually resolve within 24 hours.

ACE (angiotensin converting enzyme) inhibitor medications

Around 1 in 200 people who take these medications for heart and blood pressure, develop angioedema, usually appearing within the first few months of treatment. In some cases, it will not appear until a few months or years after treatment has started, or it may only appear if the dose of medication is increased. It is not clear why some people develop angioedema after taking this medication and others don't, and unfortunately, switching to another brand or type of ACE inhibitor does not seem to help.

The swelling that happens after taking these medications usually affects the face, tongue, and throat, is not itchy or painful, and hives do not develop. This is not an allergic reaction so will not show up on skin or blood tests. A diagnosis is usually confirmed if the symptoms disappear once the person stops taking the medication. Swelling will resolve, but it can take a few weeks after stopping the medication.

Hereditary angioedema (HAE)

HAE is a rare genetic condition occurring in approximately 1 in 50,000 people, who have low levels (deficiency) or reduced effectiveness of C1-inhibitor enzyme. Swelling usually happens in the upper airway (mouth, tongue, and throat) and the digestive tract (oesophagus, stomach, and bowel). In some cases, upper airway swelling can be so severe that it becomes life threatening.

The swellings are not itchy or painful and the colour of the skin is usually normal. Although HAE is not associated with hives (urticaria), sometimes a red circular rash can occur. Swelling of the internal digestive organs without any visible swelling on the outside of the body is common, and people will often present to hospital with severe stomach or chest pains. Episodes of HAE can be triggered by emotional stress, alcohol, hormonal changes, or trauma (such as dental surgery).

HAE is genetic, meaning it can be passed on to children and sometimes genetic changes may appear that were not present in the parents. HAE can be detected from blood testing and usually presents in adolescence rather than in infancy.

Acquired C1-inhibitor deficiency

In even rarer cases, low levels of C1-inhibitor occur with some cases of lymphoma, malignancies and in autoimmune diseases such as Systemic Lupus Erythematosus (SLE). The swellings are like those described above.

Other causes

Thyroid gland disease and some types of inflammatory arthritis are more common in people with hives (urticaria) and angioedema. In some people whose swellings persist for years without an obvious cause, an autoimmune process may be responsible. This means that part of the body's immune system may be attacking the skin. In most cases a single cause is not found. Allergic disease is almost never the cause of swellings that last for days or recur for weeks at a time.

Q 5: Is angioedema dangerous?

Angioedema does not cause damage to vital organs like the kidneys, liver, or lungs. It can be dangerous if it affects the throat or the tongue as swelling in these areas can cause difficulty breathing.

Severe throat swelling requires the early use of medication (adrenaline for anaphylaxis or medication for HAE) followed by immediate transfer to hospital by ambulance. Swelling on the outside of the neck is uncomfortable but does not affect breathing. Swelling that interferes with breathing is uncommon, even in people with recurrent angioedema. People with repeated angioedema should be referred by their doctor to a clinical immunology/allergy specialist for diagnosis and management.

Q 6: When is an allergic cause of angioedema suspected?

Angioedema that develops without any other skin symptoms is rarely due to allergy. If swelling occurs after exposure to a known allergen such as foods or medications, allergy may be the cause.

Food allergy reactions can be severe, causing symptoms such as trouble breathing, a drop in blood pressure, stomach upset, hives (urticaria) as well as angioedema. Symptoms usually occur within one to two hours of eating the food. Allergy tests can be useful for this type of reaction, but it is important to note that food allergy by itself rarely causes angioedema.

Some people think that food intolerance (which is different to allergy) plays a role in angioedema but there is no proof that it does. People with a suspected food intolerance should seek advice from a medical practitioner and/or accredited dietitian before making any changes to their diet.

Q 7: What types of swellings looks like angioedema?

- **Contact allergy** from animals or plants causes itchy rashes at the site of contact that develop into blisters that weep then peel after a few days. Although the rash may be swollen, it is limited to the affected area and there is no internal swelling.
- **Animal allergy** can cause itchy hives (urticaria) and angioedema, but this usually occurs with severe hay fever (allergic rhinitis) and/or asthma.
- **Insect stings** from bees, wasps and some ants can cause severe, localised (around where the sting occurred), temporary swelling.
- **Palindromic rheumatoid arthritis** is a rare form of arthritis that causes affected joints and limbs to swell. The swelling often lasts a few days at a time and the areas affected become painful and hot to the touch, rather than itchy.
- **Other less common conditions that look like angioedema** include the following:
 - Dermatomyositis - muscle weakness, facial redness and swelling.
 - Blockage of the superior vena cava - fluid accumulation in the neck, face, and arms due to the blockage of a major vein in the chest that does not resolve.
 - Underactive thyroid gland - causing puffiness of the face and lips known as myxoedema.
 - Facial rosacea - causing puffiness of the face, redness, flushing and pimples.
 - Orofacial granulomatosis - facial and lip swelling that does not resolve. Sometimes associated with inflammation of the bowel.
 - Subcutaneous emphysema - leakage of air into the soft tissues, often occurring after chest trauma or surgery.
 - Cluster headache - severe one-sided headache which can cause puffiness around the eye on the same side.

Q 8: How long does angioedema last?

The amount of time angioedema takes to resolve will depend on the cause. If the cause of the angioedema is found, then steps can be taken to fix the issue and the episodes of swelling should stop. An example of this would be stopping ACE (angiotensin converting enzyme) inhibitor medications, which are known to cause angioedema. If no cause is found, there is no way to predict how long the swelling will last. It may stop after a few weeks or months, or it may continue for years.

Q 9: When is testing for angioedema needed?

Testing is important for people where angioedema is suspected to help rule out other possible causes of symptoms. This usually includes a physical examination by a doctor, and some blood tests. Significant or recurring episodes of angioedema may need further tests. There are some conditions that look like angioedema at first but develop into other conditions later.

Q 10: How is angioedema managed?

- **Tests can confirm or exclude a cause** but in most cases test results are normal.
- **Symptoms may disappear over time.** Some people only have a single episode, and others may develop symptoms many times. In most cases, symptoms will resolve on their own.
- **Avoid the triggers that make symptoms worse.** It may be possible to manage angioedema by avoiding exposure to triggers that include:
 - Excessive heat, eating spicy foods, and alcohol consumption.
 - Pain relief medications - an alternative such as paracetamol may reduce symptoms.
 - ACE (angiotensin converting enzyme) inhibitors - angiotensin 2 receptor blockers are usually considered safe.
- **Antihistamines are commonly used to treat angioedema.**
 - Antihistamines block the release of histamine within the skin, which seems to be the trigger for swelling in most people.
 - Symptoms usually start to settle within one to two hours after taking antihistamines and it is best to take them on a regular basis rather than when symptoms appear.
 - The aim of treatment is to stop the episodes of angioedema from happening, make them less frequent or make the symptoms less severe.
 - Non-drowsy antihistamines can be purchased without a prescription, and these are effective in treating angioedema with hives (urticaria). They do not treat the swelling itself and are not useful for the treatment of angioedema that happens without hives.
- **Other medications** may be used if antihistamines are unsuccessful in managing angioedema. Due to potential side effects, they are usually only given under specialist supervision.

Q 11: How does aspirin and other pain medications relate to angioedema?

If people are allergic to aspirin or similar pain medications, taking them may cause swelling. One in three people with angioedema may have increased symptoms after taking these medications, even if they are not allergic to them. For these people, it may be better to take paracetamol for pain management. If taking aspirin or similar pain medications regularly without symptoms, there is no reason to stop taking them.

Q 12: Can medication for angioedema be taken during pregnancy and breastfeeding?

Most medications, such as antihistamines, used to treat angioedema are generally considered safe during pregnancy and while breastfeeding. The use of any medication in pregnancy and while breastfeeding should always be discussed with a doctor or pharmacist.

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