HEALTH BASICS

C1-INH

C1-INH is a blood protein that controls systems involved in inflammation and swelling.

WHAT IT IS
A blood protein that limits swelling and immune-system reactions

WHERE IT COMES UP
Angioedema blood tests and C1-INH medicine labels

WHY IT MATTERS
Helps control bradykinin-driven fluid leakage and swelling

WHAT IT IS NOT
Not a disease or a diagnosis

WHO TO ASK
An allergist or clinical immunology service

HOW IT IS CHECKED
C1-INH antigen level and functional blood tests

Overview

C1-INH stands for C1 inhibitor . It is also called C1 esterase inhibitor . It is a protein found in the blood, mainly produced by the liver.

C1-INH helps control several linked systems in the body. These systems affect inflammation, blood clotting, and fluid movement through blood vessel walls. Its best-known role is limiting the production of bradykinin , a chemical that can make blood vessels leak fluid into nearby tissue.

The term may appear in several places:

These meanings are related, but they are not interchangeable. A C1-INH test result is not a diagnosis by itself. C1-INH concentrate is a prescription medicine, not a routine supplement.

- A blood test measuring the amount of C1-INH
- A test measuring how well C1-INH works
- An evaluation for certain forms of angioedema
- The name of a medicine that replaces C1-INH

How it works

C1-INH belongs to a group of proteins called serine protease inhibitors , or serpins. These proteins slow particular enzymes before the enzymes create an excessive chain reaction.

One system controlled by C1-INH is the contact system . Within this system, enzymes including factor XIIa and plasma kallikrein help produce bradykinin. Bradykinin widens blood vessels and allows fluid to pass through their walls.

A controlled amount of bradykinin supports normal body functions. Too much activity, however, can cause fluid to collect under the skin or in tissues lining the mouth, throat, or digestive tract.

C1-INH also regulates part of the complement system . Complement is a network of blood proteins involved in immune defence and inflammation. The "C1" in the name refers to its effect on early complement proteins.

C1-INH does not completely switch off these systems. It keeps their activity within useful limits.

Two laboratory measurements provide different information:

A person may have too little C1-INH, or they may have a near-normal amount that does not function correctly. This is why the two tests are often considered together.

- C1-INH antigen level measures how much of the protein is present
- C1-INH functional activity measures whether the protein works properly

Why it matters for your health

C1-INH testing is most often used when a clinician is evaluating episodes of angioedema , meaning swelling in deeper layers of the skin or body tissues.

Testing may be considered when swelling keeps returning, affects the abdomen or airway, occurs without itchy hives, or has a family pattern. These features can occur for several reasons, so they do not prove that C1-INH is involved.

Results are usually interpreted with other information. This can include your medical history, examination, family history, medicines, and other blood tests.

A C4 complement test is commonly used alongside C1-INH tests. C4 is often low when C1-INH is deficient or not working, but C4 alone cannot confirm the cause. A normal C4 result does not always exclude a C1-INH-related condition.

A C1q test may help when clinicians are considering acquired C1-INH deficiency, especially

when swelling began later in life and there is no family history. C1q is often reduced in acquired deficiency and usually normal in hereditary deficiency, although exceptions occur.

Each laboratory has its own methods and reference ranges. A result marked "low" should be interpreted using the range printed on that report. Sample handling, timing, age, and illness can also affect interpretation.

What can go wrong

Several named conditions and testing situations relate to C1-INH.

Hereditary angioedema due to C1-INH deficiency is usually caused by a harmful change in the `SERPING1` gene. In type I, the amount and activity of C1-INH are low. In type II, the amount may be normal or increased, but its activity is low.

Other forms of hereditary angioedema can occur with normal C1-INH levels and function. These may involve other genes or have no identified genetic cause. Normal C1-INH testing therefore does not answer every question about recurring swelling.

Acquired C1-INH deficiency is not inherited. It can occur when C1-INH is used up too quickly or blocked by antibodies. It may be associated with certain immune-system or blood-cell disorders. Finding acquired deficiency can lead to further testing for an underlying condition.

C1-INH results can also be misleading if a sample is stored or transported incorrectly. A functional test is particularly sensitive to handling. Clinicians may repeat an unexpected result before relying on it.

Most swelling is not caused by C1-INH deficiency. Allergic or histamine-related angioedema is more common and may occur with hives or itching. Some medicines can also cause angioedema through different pathways. The pattern, timing, examination, and test results help distinguish these situations.

Swelling of the tongue or throat, trouble breathing, a change in voice, difficulty swallowing saliva, faintness, or rapidly worsening symptoms requires emergency care now . Do not wait for a C1-INH test or a routine appointment.

Recurring swelling without breathing difficulty usually calls for routine medical assessment , although severe abdominal pain, dehydration, or symptoms that are getting worse may need more urgent evaluation.

What you can do

There is no home test that can reliably measure C1-INH. There is also no food or non-prescription supplement proven to correct a low level or poor function.

If C1-INH testing has been ordered, ask whether both the antigen level and functional activity are being measured. You can also ask whether C4 or C1q would help interpret the result.

Keep copies of the complete laboratory reports, including reference ranges. A label such as "normal" or "abnormal" without the test method and range may not provide enough information for a later review.

If you have recurring swelling, keep a record of:

If a result is unexpected, ask whether it should be repeated when you are well and whether the laboratory handled the sample correctly. Genetic testing may be useful in some families, but it is not required or informative in every case.

If you already have a diagnosed angioedema condition, follow the care plan supplied by your specialist. Ask how to recognize an airway emergency, when to use prescribed treatment, and what information to carry for emergency staff.

- Where the swelling occurred and how long it lasted
- Whether hives or itching were present
- Any abdominal pain, vomiting, voice change, or breathing difficulty
- Medicines taken before the episode
- Dental work, injury, illness, stress, or hormonal changes near the episode
- Similar episodes in biological relatives

Common questions

Is C1-INH the same as C1?

No. C1 is part of the complement system. C1-INH is a regulatory protein that limits C1 and several other enzymes.

Does a low C1-INH result confirm hereditary angioedema?

Not by itself. The amount, function, C4 level, clinical history, and sometimes C1q or genetic testing need to be considered together. Acquired deficiency and laboratory problems can also produce abnormal results.

Can C1-INH be normal even when hereditary angioedema is present?

Yes. Some forms of hereditary angioedema occur with normal C1-INH amount and function.

They are evaluated using the clinical pattern, family history, and sometimes genetic testing.

Is C1-INH concentrate the same as the natural protein?

C1-INH medicines replace or supplement the protein's activity. Products may be made from screened human plasma or produced using recombinant technology. Their approved uses and routes of administration are product-specific.

Does a low C4 result mean C1-INH is low?

Not necessarily. C4 can be reduced for several reasons. It is a supporting test rather than a stand-alone diagnosis.

Can an allergy test check C1-INH?

No. Standard allergy skin tests and allergy-antibody blood tests measure different processes. C1-INH requires specific antigen and functional blood tests.

Sources

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