



CICATRICAL ALOPECIA OVERVIEW

Cicatricial alopecia, also called scarring alopecia, refers to a group of rare disorders that destroy hair follicles. The follicles are replaced with scar tissue, causing permanent hair loss.

WHAT ARE THE DIFFERENT FORMS?

Cicatricial alopecia has two forms. In the primary form, the hair follicle is the target of the destructive process. In the secondary form, the hair follicle is an innocent bystander, destroyed by another cause. This can be a severe burn, an infection, radiation, or a tumor.

This fact sheet focuses on the primary form. The primary form is defined by the type of immune cells involved. These immune cells inflame and destroy the hair follicle. They are called either lymphocytes or neutrophils. Sometimes, the process shifts between the two kinds of cells.

WHAT IS THE CAUSE?

The cause of cicatricial alopecia is not well understood. What is known is that redness, heat, pain, or swelling occurs at the upper part of the hair follicle. That is the place where stem cells and sebaceous (oil) glands are located. Stem cells are cells that can develop into different kinds of cells. If the stem cells and oil glands are destroyed, the hair follicle cannot regrow, and hair is permanently lost.

WHO GETS CICATRICAL ALOPECIA?

Cicatricial alopecia is not contagious. It occurs worldwide in otherwise healthy men and women. It affects all ages, but is not common in children.

This condition usually affects only one family member. One exception is central centrifugal alopecia. It most commonly affects women of African ancestry, and may occur in more than one family member.

FORMS OF CICATRICAL ALOPECIAS

Those involving mostly lymphocytes:

- lichen planopilaris (LPP)
- frontal fibrosing alopecia
- central centrifugal alopecia
- pseudopelade of Brocq

Those involving mostly neutrophils:

- folliculitis decalvans
- tufted folliculitis

Those involving both (called mixed inflammatory infiltrate):

- dissecting cellulitis
- folliculitis keloidalis

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WHAT ARE THE SYMPTOMS?

In some cases, hair loss happens quickly, and there is severe itching, pain, and burning. In other cases, hair loss is gradual, and there are no other symptoms.

HOW IS CICATRICAL ALOPECIA DIAGNOSED?

A doctor may see a loss of hair follicles on the scalp and ask for a scalp biopsy. The biopsy provides information about which type of cell is involved, where and how much inflammation there is, and whether the oil gland is still there. It can also show other changes in the scalp. The biopsy helps the doctor diagnose the type of cicatricial alopecia, and how much there is.

Then he or she can decide with the patient on treatment or therapy.

Other things the doctor will do:

- A clinical evaluation of the scalp, noting symptoms such as itching, burning or tenderness; signs of inflammation including redness, scaling, and pustules; and overall extent and pattern of hair loss.
- A hair-pull test to identify areas of active disease where hairs are easily pulled out.
- A microscopic evaluation of the hair bulbs—expansions at the base of the hair from which the shaft develops—to determine if the hairs pulled out in a hair-pull test are resting hairs or growing hairs.
- Take cultures of pustules, if present, to identify which microbes may be contributing to inflammation.

HOW IS CICATRICAL ALOPECIA TREATED?

Because hair loss in cicatricial alopecia is permanent and the hair does not grow back, it is important to begin aggressive treatment early before extensive hair is lost. The specific treatment used varies, depending largely on whether lymphocytes, neutrophils, or both are predominantly responsible for the hair follicle destruction.

The form of the disorder caused mainly by lymphocytes is usually treated with medicines to remove the inflammatory cells. Oral treatments may include antimalarials, antibiotics, immunosuppressive

drugs, and a class of diabetes medications called thiazolidinediones. Medicines applied directly to the skin include corticosteroids, topical tacrolimus, and other agents that suppress or modulate the immune system. Corticosteroids may be injected directly into inflamed, symptomatic areas of the scalp.

The form of the disorder caused mainly by neutrophils is treated with oral and topical antibiotics and anti-inflammatory medications. For cases involving both neutrophils and lymphocytes, treatment may include antimicrobials, anti-inflammatory medications, and retinoids. Retinoids are a class of medicines chemically related to vitamin A. An agent that blocks tumor necrosis factor-alpha (TNF- α), a molecular messenger involved in systemic inflammation, may be helpful for dissecting cellulitis that does not respond to other treatments.

Although hair cannot grow back after a follicle has been destroyed, it may be possible to stimulate follicles in the affected area before permanent damage occurs. Applying an antihypertensive medicine that promotes hair growth may stimulate viable follicles.

Treatment usually lasts a long time. It continues until the symptoms and signs are controlled, and progression of hair loss has been slowed or halted. Unfortunately, hair loss may continue silently even after the symptoms and signs have been removed. After cicatricial alopecia stabilizes, it may start up again after a period of years, and treatment may need to be restarted.

IS THERE A ROLE FOR SURGICAL TREATMENT?

After the disease has been inactive for 1 or 2 years, surgical hair restoration or scalp reduction may be useful for restoring hair to bald areas. In hair restoration surgery, also known as hair transplantation or follicular micrografting, follicles from the back of the head are surgically removed and transplanted into bare areas of the scalp. In scalp reduction, a bald area of the scalp is removed, and the part of the scalp next to it is pulled together to cover the gap. It is important to understand that cicatricial alopecia may recur even after surgical intervention.

WHAT TYPES OF DOCTORS DIAGNOSE AND TREAT CICATRICIAL ALOPECIA?

Dermatologists are the specialists who diagnose and treat cicatricial alopecia. Dermatologists are doctors trained to diagnose and treat diseases of the skin, hair, and nails.

WHAT RESEARCH IS BEING DONE?

Researchers are working to better understand the inflammation and inflammatory cells that target and destroy the hair follicles. One of the most promising areas of research in the form of the disorder called lichen planopilaris (LPP) suggests that inflammation may result from changes in the lipid and metabolism in the oil glands.

Specifically, the loss of function of a protein called peroxisome proliferator-activated receptor gamma (PPAR- γ), which regulates fatty acid storage and glucose metabolism. This leads to the abnormal processing and buildup of lipids, triggering inflammation, which results in scarring and destruction of the hair follicle. A new approach to treatment for LPP may be drugs to block PPAR- γ . These medications are already in wide use for the treatment of type 2 diabetes.

RESOURCES

Genetic and Rare Diseases (GARD) Information Center

Website: www.rarediseases.info.nih.gov/GARD

American Academy of Dermatology

Website: www.aad.org

Cicatricial Alopecia Research Foundation

Website: www.carfintl.org

North American Hair Research Society

Website: www.nahrs.org

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FOR YOUR INFORMATION

This publication contains information about medications used to treat the health condition discussed here. When this publication was printed, we included the most up-to-date (accurate) information available. Occasionally, new information on medication is released.

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