What you should know about
CICATRICIAL ALOPECIA

CICATRICIAL ALOPECIA RESEARCH FOUNDATION

www.carfintl.org
The Foundation

- provides funds for research to find effective treatments and a cure
- supports education and advocacy
- raises public awareness

CARF is a non-profit, tax exempt organization under Section 501(c)(3) of the Internal Revenue Code.
What is cicatrical alopecia or scarring alopecia?

The term “cicatrical alopecia” refers to a diverse group of rare disorders that destroy the hair follicle, replace it with scar tissue, and cause permanent hair loss. The clinical course is highly variable and unpredictable. Hair loss may be slowly progressive over many years, without symptoms, and unnoticed for long periods. Or the hair loss may be rapidly destructive within months and associated with severe itching, pain and burning. The inflammation that destroys the follicle is below the skin surface and there is usually no “scar” seen on the scalp. Affected areas of the scalp may show little signs of inflammation, or have redness, scaling, increased or decreased pigmentation, pustules, or draining sinuses. Cicatrical alopecia occurs in otherwise healthy men and women of all ages, is not contagious, not hereditary and is seen worldwide.

Are there different kinds of cicatrical alopecia?

Yes, cicatrical alopecias are classified as primary or secondary. This discussion is confined to the primary cicatrical alopecias in which the hair follicle is the target of the destructive inflammatory process. In secondary cicatrical alopecias, destruction of the hair follicle is incidental to a non-follicle-directed process or external injury, such as severe infections, burns, radiation, or tumors.
Primary cicatricial alopecia is currently classified by the type of inflammatory cells seen on scalp biopsy. The inflammatory cells may be primarily lymphocytes, neutrophils, or sometimes the inflammation has mixed cells. Cicatricial alopecias that involve predominantly lymphocytic inflammation include lichen planopilaris, frontal fibrosing alopecia, central centrifugal cicatricial alopecia, pseudopelade (Brocq), chronic cutaneous lupus erythematosus, and keratosis follicularis spinulosa decalvans. Cicatricial alopecias that involve predominantly neutrophilic inflammation include folliculitis decalvans and tufted folliculitis. Sometimes the inflammation shifts from a predominantly neutrophilic process to a lymphocytic process, or visa versa. Cicatricial alopecias with a mixed inflammatory infiltrate include dissecting cellulitis and folliculitis keloidalis, both of which are secondary to follicular rupture.

What causes cicatricial alopecia?

The cause of the various cicatricial alopecias is poorly understood. However, all cicatricial alopecias involve inflammation directed at the hair follicle, usually the upper part of the follicle where the stem cells and sebaceous gland (oil gland) are located. If the stem cells and the sebaceous gland are destroyed, there is no possibility for regeneration of the hair follicle and permanent hair loss results.

New research suggests that there is a loss of function of a “master regulator” called the peroxisome proliferator-activated receptor gamma, or PPAR gamma. PPAR gamma plays an important role in the preservation of hair follicle cells, including stem cells, and sebaceous glands.
Decreased PPAR gamma leads to sebaceous gland dysfunction, which causes abnormal processing and buildup of “toxic” lipids. This abnormal buildup of lipids triggers inflammation that ultimately destroys the hair follicle.

**Who is affected by cicatricial alopecia?**

Cicatricial alopecias affect both men and women of all ages and are rare in children. Some disorders may manifest in the teenage years. Epidemiologic studies have not been performed to determine the incidence of cicatricial alopecias. In general, they are not common.

There have been a few reports of cicatricial alopecia occurring in a family. However, the majority of patients with cicatricial alopecia have no family history of a similar condition.

Central centrifugal cicatricial alopecia affects women of African ancestry most commonly and may occur in several family members. Frontal fibrosing alopecia is seen most commonly in post-menopausal women but also occurs in young women and men. While it is possible to have more than one type of hair loss condition, non-scarring types of hair loss do not turn into scarring forms of hair loss.

**Are cicatricial alopecias associated with other illnesses?**

In general, cicatricial alopecias are not associated with other illnesses and usually occur in healthy men and women.

Patients with chronic cutaneous lupus erythematosus may have an increased personal and family history of autoimmune disorders.
How are cicatricial alopecias diagnosed?

A scalp biopsy is essential for the diagnosis of cicatricial alopecia and is the necessary first step. Findings of the scalp biopsy, including the type of inflammation present, location and amount of inflammation, and other changes in the scalp, are necessary to diagnose the type of cicatricial alopecia, to determine the degree of activity, and to select appropriate therapy.

Clinical evaluation of the scalp is also important. Symptoms of itching, burning, pain or tenderness usually signal ongoing activity. Signs of scalp inflammation include redness, scaling, and pustules. However, in some cases there are few symptoms or signs and only the scalp biopsy demonstrates the active inflammation. The overall extent and pattern of hair loss is noted and sometimes photographed for future comparison. A hair “pull test” is performed to identify areas of active disease in which hairs are easily pulled out. The pulled hairs are mounted on a slide and the hair bulbs are viewed with a microscope to determine how many are growing hairs and how many are resting hairs. In addition, if pustules are present, cultures may be performed to identify which microbes, if any, may be contributing to the inflammation.

A thorough evaluation that includes all of these parameters is important in diagnosing a cicatricial alopecia and in identifying features in individual patients that will help the selection of therapy.

Diagnosis and treatment of cicatricial alopecias is often challenging. For this reason, it is helpful to be evaluated by a dermatologist with a special interest or expertise in scalp and hair disorders, and who is familiar with current diagnostic methods and therapies. A hair specialist who is experienced in the evaluation and treatment of
patients with cicatricial alopecias may be found by contacting the Cicatricial Alopecia Research Foundation (www.carfintl.org), the American Academy of Dermatology (www.aad.org), or the North American Hair Research Society (www.nahrs.org)

**How are cicatricial alopecias treated?**

As mentioned above, primary cicatricial alopecias are classified by the predominant type of inflammatory cells that attack the hair follicles: i.e., lymphocytes, neutrophils, or mixed inflammatory cells. Treatment strategies are different for each of these three subtypes and detailed treatment options are beyond the scope of this discussion. However, certain general principals are reviewed below.

Treatment of the lymphocytic group of cicatricial alopecias (including lichen planopilaris, frontal fibrosing alopecia, central centrifugal cicatricial alopecia, pseudopelade (Brocq), chronic cutaneous lupus erythematosus, and keratosis follicularis spinulosa decalvans) involves use of anti-inflammatory medications. The goal of treatment is to decrease or eliminate the lymphocytic cells that are attacking and destroying the hair follicle. Oral medications may include hydroxychloroquine, doxycycline, mycophenolate mofetil, pioglitazone, cyclosporine, or corticosteroids. Topical medications may include corticosteroids, topical tacrolimus, Derma-Smoothe/FS scalp oil; triamcinolone acetonide (a corticosteroid) may be injected into inflamed, symptomatic areas of the scalp.
Treatment of the neutrophilic group of cicatricial alopecias (folliculitis decalvans and tufted folliculitis) is directed at eliminating the predominant microbes that are invariably involved in the inflammatory process.

Culture and sensitivities are essential for the selection of oral antibiotics, which are the mainstay of therapy. Topical antibiotics may be used to supplement the oral antibiotic and triamcinolone acetonide (a corticosteroid) may be injected into inflamed areas.

Treatment of the mixed group of cicatricial alopecias (dissecting cellulitis and folliculitis keloidalis) may include antimicrobials although culture often does not grow a pathogen. Isotretinoin, anti-inflammatory medications such as corticosteroids, and tumor necrosis factor inhibitors may be used. In dissecting cellulitis, incision and drainage of nodules may be needed.

You should discuss any treatment with your dermatologist who will also explain potential side effects, as well as laboratory tests that are needed before starting medications and sometimes are monitored during treatment.

The course of cicatricial alopecia is usually prolonged and treatments are often given for 6 to 12 months or longer. Treatment is continued until the symptoms and signs of scalp inflammation are controlled. In other words, itching, burning, pain and tenderness have cleared and scalp redness, scaling, and/or pustules are no longer present.
Treatment may then be stopped. However, with the exception of pioglitazone, current treatments do not usually influence the underlying process and may not arrest progression of hair loss even when symptoms and signs have cleared. Clinical activity often recurs and treatment frequently needs to be repeated.

Because of the above last statement, surgical treatment is not an option in most cases except under certain circumstances. If the disease has been inactive for one or two years, then surgical removal of the scarred scalp and/or hair restoration surgery may be considered for cosmetic benefit. Folliculitis keloidalis is one exception in that excision of the affected scalp (scalp reduction) may provide relief for the patient.

**Will my hair grow back?**

Hair will not regrow once the follicle is destroyed. However, it may be possible to treat the inflammation in and around the surrounding follicles before they are destroyed and for this reason it is important to begin treatment as early as possible to halt the inflammatory process. Minoxidil solution (2% or 5%) applied twice daily to the scalp may help to stimulate any small, remaining, unscarred follicles. The progression of hair loss is unpredictable. In some cases, progression is slow and there is sufficient hair remaining to cover the affected scalp areas; in other cases, progression can be rapid and extensive.
How should I care for my hair?

Hair care products and shampoos can generally be used with any frequency desired, as long as the products are not irritating to the scalp. Dermatologists may recommend specific shampoos and products to decrease scalp symptoms, scaling, and inflammation. Hair pieces, wigs, hats, scarves may be used freely.

Does CARF have a website?

Yes, the website is: www.carfintl.org and is updated on a regular basis to include newsworthy items, information about the annual patient conference and resources.

CARF does not endorse any of the medications, treatments, or products. Please consult with your physician before considering any of the drugs or treatments.
YES I want to help myself and others with cicatrical alopecia. Enclosed in my donation to help the Cicatricial Alopecia Research Foundation with its programs of research, support, and education.

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