

CCCA (Central Centrifugal Cicatricial Alopecia)

What is CCCA? CCCA belongs to a group of disorders called cicatricial (scarring) alopecias that cause progressive, often irreversible destruction of the hair follicle secondary to inflammation and scarring.

Whom does CCCA affect? CCCA primarily affects middle-aged women of African descent. It often runs in families.

What are the symptoms and signs of CCCA? Hair loss typically begins on the crown of the scalp and moves outward, often with an insidious progression. Some people do not experience any symptoms. Others may have itching, burning, and tenderness of the scalp early in their disease. Hair breakage may be an early clinical sign of CCCA, but can often be confused for traction alopecia (loss of hair at the hairline). Traction alopecia can be a result of tight hair styling and lack of hair hydration, but some studies have shown that it may be related to CCCA.

What causes CCCA? The cause of CCCA is thought to be multifactorial, including genetic, autoimmune, infective, and environmental factors. The hair follicle becomes damaged (exactly how is still not understood), causing persistent irritation and mild inflammation. This eventually causes scarring of the hair follicle, meaning a new hair cannot grow.

Is CCCA associated with other illnesses? CCCA is associated with type 2 diabetes mellitus. A small pilot study found that there is a nearly three-fold increased risk for breast cancer in Black CCCA patients. This was a small study and larger studies are needed to assess this finding to the general population. CCCA has been shown to have an increased expression of genes involved in fibroproliferative disorders like keloids scars and uterine fibroids. One research study evaluated whether CCCA was associated with fibroproliferative disorders, specifically of the lungs (interstitial lung disease), arteries (atherosclerosis of the aorta), liver (non-alcoholic steatohepatitis), kidney (end stage renal disease), and uterus (uterine leiomyoma). This study found no association between these disorders and CCCA.

How should I care for my hair?

Gentle hair care practices are recommended for patients with CCCA. Recent research shows that patients who showed improvement in their CCCA were more likely to use hooded dryers and natural hairstyles.

What tests are done to confirm CCCA? Obtaining a thorough medical history and history of hair styling practices is important when diagnosing CCCA. Decreased hair density and follicular dropout are clinical clues that can point to a diagnosis of CCCA; however, only a scalp biopsy can provide a confirmatory diagnosis. This is a minimally invasive procedure performed in-office. Local anesthetic is given and a small sample of skin is sent for microscopic examination. The biopsy can provide information such as the number of hair follicles, the amount of fibrotic tissue, and the extent of inflammation. Additionally, the dermatologist may order labs to assess for any associated conditions

How is CCCA treated? The goals of treatment are to relieve symptoms and to halt disease progression. Regrowth of hair is not possible if the hair follicle has been replaced by scar tissue. CCCA is treated by targeting the inflammatory response. Treating CCCA earlier has a better response.

- **Oral anti-inflammatory medications** are utilized when inflammation is severe. Doxycycline, an antibiotic with anti-inflammatory properties, is a first-line oral medication. If doxycycline is not sufficient to halt the inflammatory process, hydroxychloroquine, an antimalarial with anti-inflammatory properties, is considered.
- Potent **corticosteroids** may be used for symptom control in the form of topical solutions, oils, foams, lotions, and sprays. Non-steroidal **calcineurin inhibitors** such as tacrolimus may also be used for symptom control. If there is concurrent seborrheic dermatitis (dandruff), treatment of the seborrheic dermatitis is imperative to attenuate the inflammatory response. In women of African American descent, the antifungal shampoo ciclopirox 1% is prescribed more often than ketoconazole 2%, due to increased hair fragility.
- **Corticosteroid injections** are administered on a monthly basis to decrease symptoms and inflammation.
- **Topical minoxidil 5%**, foam or solution, can be used to stimulate remaining hair follicles and thicken the existing hair.
- **Platelet rich plasma (PRP)** which has shown to be efficacious in women with androgenetic alopecia (aka female pattern hair loss) is currently being studied in women with CCCA.
- Other novel medications include **topical metformin 10%**.
- Once hair loss has stabilized and the inflammation has been controlled, **hair transplantation** may be considered.

Frontal Fibrosing Alopecia (FFA) and Lichen Planopilaris (LPP)

What are LPP and FFA? LPP is a form of primary immune-mediated scarring alopecia in which lymphocytes, a type of white blood cell, attack the hair follicle and can cause irreversible hair loss. While classic LPP usually affects the vertex (crown) of the scalp, FFA is a version of LPP that affects the frontal hairline and occasionally the eyebrows.

Whom does LPP and FFA affect? LPP and FFA affect mostly Caucasian women between the ages of 40-60, but has been seen in men as well.

What are the signs and symptoms of LPP and FFA? Symptoms of FFA include pain and itching at the frontal hairline and loss of eyebrow and sideburn hair. It may be mistaken for traction alopecia, especially in women with a history of tight hair styles. FFA sometimes has facial papules early in the disease course. LPP is associated with variable patterns of hair loss and a painful, itchy scalp. Around half of the cases of LPP will have signs of lichen planus (purple itchy bumps) on the skin.

What causes LPP and FFA? Similar to CCCA, the exact cause of LPP and FFA are not known. FFA is thought to have an autoimmune and hormonal component, but it is believed that genetics and environmental exposures play a role as well. FFA and LPP autoimmunity can be triggered by stress, infection, metal exposure, or pollution. Some controversial studies have noted an association with chemical sunscreen use. LPP is thought to be mostly an autoimmune problem and does not have a genetic component.

What tests are done to diagnose LPP and FFA? A thorough clinical history and physical exam are conducted. Your dermatologist may use a special skin magnifier, called a dermatoscope, to look closely at the scalp for signs of inflammation. A scalp biopsy may be necessary to confirm the diagnosis.

How are LPP and FFA treated? LPP can be difficult to treat and has the best results when treated early. Generally, patients start with steroid injections in the scalp and progress to oral or topical steroids or immunosuppressant medications. FFA is treated very similarly to LPP. (Further descriptions of these treatments can be found under "CCCA.") For both FFA and LPP, doctors may additionally prescribe low-dose oral minoxidil, or dutasteride or finasteride to treat the hormonal component. Laser treatments have also had promising preliminary results in FFA.

Are LPP and FFA associated with other diseases? FFA has been associated with rosacea, hypothyroidism, LPP, and other autoimmune conditions. Notably, there is a high association in women taking hormone replacement therapy. LPP is sometimes associated with lichen planus elsewhere on the body and Lassueur Graham-Little Piccardi Syndrome.

How should I care for my hair? Reduce heat and harsh chemicals used in the hair. Use gentle skincare and talk to your dermatologist about the products you are using.

Other Types

What is Folliculitis Decalvans (FD)? A rare form of primary immune-mediated scarring alopecia caused by neutrophils. The bacteria *s. aureus* and poor immune response are also thought to play a role in causing this disease.

Whom does FD affect? Typically middle-aged adults.

How is FD treated? FD is treated with oral and topical antibiotics. Treatments involving photodynamic therapy have been recently studied with favorable results.

What is Discoid Lupus Erythematosus (DLE)?

Categorized as a chronic form of cutaneous lupus erythematosus. Ears, face, and scalp are most commonly involved.

Whom does DLE affect? 20% of patients with DLE with scalp involvement will exhibit scarring alopecia.

How is DLE treated? Early diagnosis and treatment are important. Establishing goals of treatment is important since hair loss is permanent once the hair follicle is completely scarred. Sunscreens and smoking cessation may be helpful in prevention of new areas of hair loss. Antimalarial medications, topical and injected steroids, and non-steroidal agents called calcineurin inhibitors are first-line treatments.

What is Dissecting Cellulitis of the Scalp (DCS)? A rare neutrophilic scalp condition affecting the scalp.

Who does DCS affect? DCS tends to appear in young males of African descent, has a preference for the vertex (crown) of the scalp, and is associated with tobacco use and obesity.

How is DCS treated? DCS is challenging to treat but many options are available including oral antibiotics, isotretinoin, dapsone, TNF-alpha inhibitors, and surgical excision.

Visit www.scarringalopecia.org for information on all types of scarring alopecia.

Where can I go for more information about scarring alopecia?

Diagnosis and treatment of scarring alopecia is often challenging and can take an emotional toll on patients. For this reason, it is helpful to be evaluated by a dermatologist with a special interest or expertise in scalp and hair disorders who is familiar with current diagnostic methods and therapies. A hair specialist who is experienced in the evaluation and treatment of patients with scarring alopecias may be found on our website (scarringalopecia.org) or the American Academy of Dermatology (aad.org). You can also use these websites to learn more about your condition. Clinical research trials in your area may be found on clinicaltrials.gov by searching “scarring alopecia.” Information on support groups and resources can also be found on our website.

You are not alone!

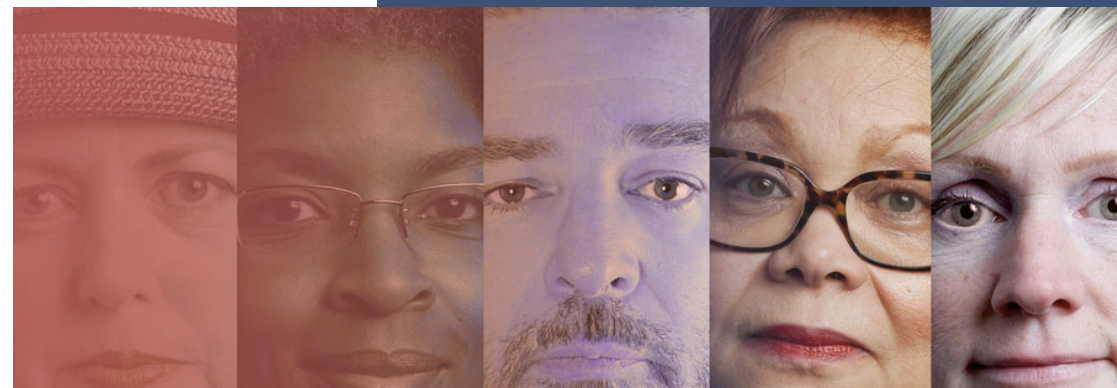
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About the Scarring Alopecia Foundation (SAF)

Our mission is to provide education and patient support, raise public awareness and advance and promote research into scarring alopecia.

SAF is the exclusive patient advocacy organization dedicated to meeting the unique needs of the scarring alopecia community. We provide reliable and up-to-date information, serving as the driving force for this patient population. Our commitment is motivated by our desire to understand the causes of this permanent hair loss disease, find effective treatments, and enhance the lives of those affected. SAF operates as a 501(c)(3) non-profit organization, and our tax identification number is 20-2049037.



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SCARRING ALOPECIA INFORMATION

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Lichen Planopilaris (LPP)

Frontal Fibrosing Alopecia (FFA)

Folliculitis Decalvans (FD)

Discoid Lupus Erythematosus (DLE)

Dissecting Cellulitis of the Scalp (DCS)



What Is Scarring Alopecia?

The term “scarring alopecia” (also known as cicatricial alopecia) refers to a diverse group of inflammatory disorders that destroy the hair follicle, replace it with scar tissue, and cause permanent hair loss.

There are distinct types of cicatricial alopecia, classified as primary or secondary. In **primary cicatricial alopecia**, the hair follicle is the target of the destructive inflammatory process. In **secondary cicatricial alopecia**, 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 a scalp biopsy. The inflammatory cells may be primarily lymphocytes or neutrophils, or sometimes the inflammation has mixed cells.

Due to the nature of these inflammatory diseases, the immune system plays a significant role in its pathology, clinical presentation, and response to treatment.

Each type of scarring alopecia is unique. Hair loss may occur over a short or long period of time and may be accompanied by symptoms such as pain, itching, or burning, while some types of scarring alopecias may present with no symptoms outside of hair loss. The inflammation that destroys the follicle is below the skin surface and there is usually no “scar” seen on the scalp.