PATIENT INFORMATION LEAFLET

CENTRAL CENTRIFUGAL CICATRICIAL ALOPECIA

WHAT ARE THE AIMS OF THIS LEAFLET?

This leaflet has been written to help you understand more about central centrifugal cicatricial alopecia. It tells you what this condition is, what causes it, what can be done about it, and where you can find out more about it.

WHAT IS CENTRAL CENTRIFUGAL CICATRICIAL ALOPECIA?

Central centrifugal cicatricial alopecia (CCCA) is a type of hair loss (alopecia) that destroys the hair follicles and replaces them with scar tissue, causing permanent hair loss.

In the past, CCCA was also called hot comb alopecia, follicular degeneration syndrome, and chemically induced cosmetic alopecia.

WHO IS AFFECTED BY CENTRAL CENTRIFUGAL CICATRICIAL ALOPECIA?

CCCA occurs almost exclusively in women of African descent aged 30 to 40 years with hair that grows in tight curls. Men and children of African descent may also be affected but it is less frequently seen.

WHAT CAUSES CENTRAL CENTRIFUGAL CICATRICIAL ALOPECIA?

The hair loss in CCCA is due to inflammation developing around the hair follicles (the roots within the scalp that produce the hair). These hair follicles are damaged and destroyed by the inflammation, eventually being replaced by fibrosis (scar tissue). Therefore, in the areas where the hair is lost there is no potential for the hair to regrow.

What causes this inflammation/hair damage in CCCA is not yet known, but it is likely to be due to multiple factors, including:

- Genetic factors: CCCA may occur in several family members.
- Hair care practices: hair straightening using hot combs (straighteners) and chemical relaxers (lotion or cream to alter the hair texture), tight hairstyles, braids and weaves have all been suggested as increasing the risk of developing CCCA; however, not all studies have found this link. Some people may have another cause of hair loss at the same time such as traction alopecia due to tight hairstyles.
- Autoimmune factors: when your own immune system does not recognise your hair follicles and tries to fight against them causing inflammation.
- Increased tendency to develop fibrosis (scar tissue); keloid scars (raised scars) and uterine fibroids (benign growths in a woman's womb) are more common in women affected by CCCA.
- CCCA may also be associated with type 2 diabetes. In this case there can be a higher risk of developing bacterial scalp infections.
- Emerging evidence found an association with CCCA and low vitamin D levels.
- Furthermore, obesity, as measured by body mass index (BMI), has been associated with an increased risk of CCCA.

More research is needed to understand how these factors may contribute to the development of CCCA.

IS CENTRAL CENTRIFUGAL CICATRICIAL ALOPECIA HEREDITARY?

Studies have shown a genetic link (PADI3 gene) in about 25% of patients affected by CCCA. A change in this gene leads to

abnormalities of the hair structure resulting in increased hair fragility and breakage.

WHAT DOES CENTRAL CENTRIFUGAL CICATRICIAL ALOPECIA FEEL AND LOOK LIKE?

Often there are no symptoms. Some people may experience burning, tingling, itching, soreness, or tenderness of the scalp.

Hair breakage may be an early sign before CCCA patches develop. CCCA typically begins as a slight patch of hair thinning on the midline of the crown (top) of the scalp. Over time, the patch expands outwards in all directions (described as a centrifugal pattern) and the severity of hair loss increases, with the most severe hair loss remaining in the center. This progression is usually very slow.

Hair loss and hair breakage may also be seen at the front and sides of the hairline due to traction alopecia (e.g. pulling the hair into a tight ponytail, braids, etc.). This is often also seen in women affected by CCCA, due to the association of tight hairstyles and traumatic haircare in both these conditions.

The scalp that is exposed may appear shiny, especially when the hair follicles have gone. It is also common to see shorter, finer hairs within the area of hair loss due to shrinkage of the hair follicle. Usually the scalp skin looks normal, although in some people the skin may be more inflamed (identified by redness, pigment change and scaliness).

HOW IS CENTRAL CENTRIFUGAL CICATRICIAL ALOPECIA DIAGNOSED?

Diagnosis is based on the history and clinical features (appearance), including the pattern of hair loss and appearance of the scalp skin and hair. If the diagnosis is unclear, a scalp biopsy (taking a skin sample from the scalp) may be required.

CAN CENTRAL CENTRIFUGAL CICATRICIAL ALOPECIA BE CURED?

No, there is currently no cure for CCCA. Early recognition and treatment can help with symptoms and limit the extent of hair loss.

HOW CAN CENTRAL CENTRIFUGAL CICATRICIAL ALOPECIA BE TREATED?

The aim of CCCA treatment is to stop hair loss and maintain the remaining hair by reducing the inflammation responsible for damaging the hair follicles. The basic treatments are anti-inflammatory medications such as those listed below.

However, hair regrowth is unlikely to occur. Sometimes, hair regrowth may occur, but the hair will not regrow from follicles that have been permanently damaged. Starting treatment in the very early stages of the condition is critical for maximising the likelihood of successful results.

Response to CCCA treatment is slow and will usually take at least six months before any effect can be seen.

Stop (or reduce) damaging hair-grooming practices: Although there is no consensus regarding hair styling recommendations, women are encouraged to consider natural hairstyles.

Consider less use of all heated hair devices (hooded dryers, blow dryers, hot combs, and flat irons), avoid tight braids, weaves or extensions or any other hair practice that causes scalp discomfort or irritation. Hardening gels and sprays should be avoided as they increase the hair's fragility. Relaxers should only be performed by a professional, no more frequently than every 6-8 weeks and to new hair growth only. The scalp should not burn when using a relaxer.

Recent research has found that patients wearing natural hairstyles have higher chances of CCCA improving compared with patients not wearing natural hairstyles.

The pharmacological treatment of CCCA can be divided into 2 approaches: the anti-inflammatory approach and the hair regrowth approach.

Anti-inflammatory topical treatment options

Topical steroids: Strong steroid based preparations, (e.g. lotions, gels, creams, ointments, mousses), can be applied to the scalp to help improve any symptoms of itch

or redness and to dampen any inflammation within the skin. This would not be prescribed for long as prolonged use of topical steroids can cause thinning, spots, and lightening of the skin.

Steroid injections (known as 'intralesional steroids') may be offered to you by your doctor to treat a small area on your scalp. These can be uncomfortable and may cause thinning or dimpling of the skin, known as atrophy, or lighter coloured patches of skin at the site of injection.

Topical calcineurin inhibitors: These topical treatments can settle inflammation of the affected area of skin. They do not have the potential to cause thinning of the skin as seen with topical steroids. Side-effects include stinging on initial application which usually improves with time. Excessive sun exposure, sunbathing and sunbeds should be avoided while using this treatment (see Patient Information Leaflet on calcineurin inhibitors).

Anti-dandruff shampoos: These agents help to ease itching, discomfort, and control scalp flaking. Seborrhoeic dermatitis (dandruff) has been identified as the most common hair problem for people who are affected by CCCA at the same time. Dandruff may play a role in worsening CCCA, therefore, CCCA patients are encouraged to use anti-dandruff shampoo every 1 to 2 weeks.

Anti-inflammatory tablet treatments

Tetracycline antibiotics: These are antibiotic tablets that also have anti-inflammatory effects and aim to slow the progression of CCCA. They are usually taken daily for 6 months, after which your response to this therapy should be assessed by you and your doctor before deciding on whether to continue.

Hydroxychloroquine: This is another anti-inflammatory tablet commonly used to treat other conditions like arthritis. It usually takes 6 months to see whether the drug is effective. If helpful it may be continued for longer until the condition stops getting worse. Very rarely, hydroxychloroquine may damage the retina (the light sensitive layer of cells at the back of the eye - retinopathy). Around seven patients out of every 100 taking hydroxychloroquine for more than 5 years may develop retinopathy and it is much higher in people taking the drug for 20 years or longer. The risk of this is generally prevented by keeping the dose low and limiting the overall length of time you are on this treatment. While you are taking hydroxychloroquine, a baseline specialist eye examination, followed by annual eye tests, are recommended.

Immunosuppressive drugs: Several different tablets have been suggested for the treatment of scarring alopecias and may rarely be needed in CCCA. These drugs work by suppressing the immune system, with varying degrees of success. These are usually safer than taking steroid tablets in the long-term but do have side effects and therefore require close monitoring, with periodic clinical reviews and regular blood tests. It is not recommended for women to become pregnant whilst taking immunosuppression drugs. These drugs include ciclosporin, methotrexate and mycophenolate mofetil.

Hair regrowth therapy

Topical minoxidil solution or foam: You can apply 2% or 5% minoxidil solution/foam to the affected areas on the scalp once or twice a day alongside the above treatments to help stimulate hair growth. Topical Minoxidil can sometimes cause irritation to the skin and may stimulate facial hair growth in some women (although this is not permanent). It is not available on prescription but can be bought over the counter or online.

In advanced cases when there has been a wide area of CCCA, it may be preferable not to offer any medical therapy in view of the poor likelihood of improvement. You should be offered psychological support at all stages of the process if you feel this would help. Interventions that assist in camouflaging hair loss, can be very useful for patients with advanced CCCA, including:

Wigs and hair pieces: These can either be bought privately or obtained through the support of the NHS with a consultant's prescription (please note, prescription charges apply).

Cosmetic camouflage: Preparations containing small, pigmented fibres or coloured sprays are available from the internet and may help to disguise the hair loss. These preparations are not waterproof and may wash away if the hair gets wet (for example, through swimming, perspiration, or rain) needing to be reapplied regularly.

Surgical treatments

Surgical treatments are not offered routinely under the NHS. These can be sought privately and include:

- Hair transplantation. This is a procedure whereby hair follicles are taken from other places on the scalp and transplanted into the bald areas. This option is only available if the inflammatory process has been well-controlled with treatment for at least a year. This should be done with caution due to the high risk of keloid (raised) scar formation in people affected by CCCA.
- Scalp reduction surgery. This involves the surgical removal of the bald area and stretching the remaining hair-baring scalp skin to cover the removed area. However, this may leave a visible scar.

SELF-CARE (WHAT CAN I DO?)

- Join a hair loss support group. Studies found that CCCA has a significant negative impact on quality of life. Supportive social network and strong relationships (friends and relatives) have a protective effect on psychological wellbeing.
- Seek unbiased medical help and be wary of online solutions, especially those that offer instant or quick remedies.
- Avoid chemicals (such as hair relaxers) if possible. Please see BAD leaflet on Afro-textured hair care.
- Avoid traumatic tight hair practices such as tight braids, weaves, or extensions. However, we understand that changing hairstyling techniques can be difficult for patients because of strong personal preference or cultural, religious, or professional requirements. If you are unable to completely discontinue a traction hairstyle, we encouraged you to:
    1. Alter the hairstyle in a manner that minimizes tension on the hair.
    2. Be advised that the development of pain, tenderness, or pimples from a hairstyle should not be tolerated. If such symptoms occur, the hairstyle should be removed or adjusted immediately.
    3. Take down any traction hairstyle whenever possible. Try wearing such hairstyles infrequently and for brief periods.
- "Protective hairstyles" have been advertised on social media with the goal of reducing daily manipulation of the hair and harsh environmental exposures. These hairstyles range from cornrows and bantu knots to braided lace wigs and crotchet styling. Be aware, the use of hair extensions and tugging on the hair for braiding may inadvertently apply excess tension to the hair and may promote inflammation.
- Avoid exposing hair to excessive heat (such as hooded dryers, blow dryers, hot combs, and flat irons) and increased friction (such as vigorous brushing).
- It is important to protect any bald areas of your scalp from the sun to prevent sunburn and long-term sun damage. You should cover any bald patches with sun block or a hat if you are going to be exposed to sunlight for prolonged periods.

WHERE CAN I GET MORE INFORMATION ABOUT 5-FU CREAM?

Patient support groups providing information:
Alopecia UK
Web: www.alopecia.org.uk

E-mail: info@alopecia.org.uk

Scarring Alopecia Foundation
Web: www.scarringalopecia.org/

Weblinks to other relevant sources:

British Hair and Nail Society:
https://bhns.org.uk

DermNet:
https://dermnetnz.org/topics/central-centrifugal-cicatricial-alopecia/

Jargon Buster:
https://www.skinhealthinfo.org.uk/support-resources/jargon-buster/

Please note that the British Association of Dermatologists (BAD) provides web links to additional resources to help people access a range of information about their treatment or skin condition. The views expressed in these external resources may not be shared by the BAD or its members. The BAD has no control of and does not endorse the content of external links.

BRITISH HAIR & NAIL SOCIETY
Specialists in hair and nail disorders

This leaflet aims to provide accurate information about the subject and is a consensus of the views held by representatives of the British Association of Dermatologists and the British Hair and Nail Society: individual patient circumstances may differ, which might alter both the advice and course of therapy given to you by your doctor.

This leaflet has been assessed for readability by the British Association of Dermatologists' Patient Information Lay Review Panel

BRITISH ASSOCIATION OF DERMATOLOGISTS

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