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 (CCCA). It tells you what this condition is, what it is caused by, what can be done about it, and where you can find out more about it.

What is Central Centrifugal Cicatricial Alopecia?

Alopecia is a general term for hair loss. Central centrifugal cicatricial alopecia (CCCA) is a type of permanent hair loss that often starts on the top of the scalp and spreads outwards. Other names for CCCA include ‘Follicular Degeneration Syndrome’ and ‘Hot-comb alopecia’.

CCCA is the most common form of permanent hair loss seen in women of African descent, mainly beginning around the ages of 30-40 years. Very occasionally, it may also affect men.

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

- A genetic predisposition.
- 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.

- Autoimmune predisposition (where your own immune system does not recognise your hair follicles and tries to fight against them causing inflammation).
- Increased tendency to develop fibrosis; keloid scars (raised scars) and uterine fibroids (abnormal growths in a woman’s womb) are more common in women affected by CCCA.
- CCCA may also be associated with type 2 diabetes mellitus and with a higher risk of developing bacterial scalp infections

Is Central Centrifugal Cicatricial Alopecia hereditary?

Studies have proposed that in some people with affected by CCCA there is a genetic link identified (i.e. PADI3 gene mutation) that stops the hair developing normally, predisposing the person to this type of hair loss.

What are the symptoms of Central Centrifugal Cicatricial Alopecia?

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

What does Central Centrifugal Cicatricial Alopecia look like?

Hair breakage may be an early sign before CCCA patches develop. Hair loss typically begins at the top of the scalp and extends outwards (described as a centrifugal pattern). 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 (see Patient Information Leaflet on Traction Alopecia for further information).

The scalp that is exposed may appear shiny, especially when the hair follicles are lost. We also commonly see shorter, finer ‘minaturised’ 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, 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 diagnosis is important in order to start treatment to control any symptoms and prevent further hair loss.

How can Central Centrifugal Cicatricial Alopecia be treated?

The aim of CCCA treatment is to improve the inflammation that is responsible for damaging the hair follicle, as well as control any symptoms and slow the progression of hair loss in order to minimise the size of the permanent area of hair loss. The basic treatment is anti-inflammatory medications. Unfortunately, hair regrowth is unlikely to occur in places where there has been permanent damage to the hair follicle.

Treatment options for CCCA include:

Stop (or reduce) damaging hair-grooming practices: Women are encouraged to consider natural hairstyles, minimise the use of 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 only to new hair growth only. The scalp should not burn when using a relaxer.

Topical steroids: Potent steroid based preparations, (e.g. lotions, gels, or mousses), can be applied to help improve any symptoms of itch or redness of the scalp and to dampen any inflammation within the skin. 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 at the site of injection.

Topical calcineurin inhibitor creams and ointments: These topical treatments can settle local inflammation. 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).

*Topical Minoxidil solution or foam:* Applying 2% or 5% minoxidil solution/foam to the affected areas on the scalp once or twice a day may be used 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.

*Anti-dandruff shampoos:* These can help to ease itching, discomfort and control scalp flaking.

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 the patient’s response to this therapy should be objectively evaluated before deciding on whether to continue.

*Hydroxychloroquine:* This is another anti-inflammatory tablet commonly used to treat other conditions like arthritis. It usually needs 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. Around seven patients out of every 100 taking hydroxychloroquine for more than 5 years may develop this retinopathy and it’s 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. (see Patient Information Leaflet on hydroxychloroquine for further information).

*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 these medications. The immunosuppressive drugs include ciclosporin, methotrexate and mycophenolate mofetil. (Please see the relevant Patient Information Leaflets for further information).
In advanced cases, it may be preferable not to offer any medical therapy in view of the poor prognosis. You should be offered psychological support at all stages of the disease process if you feel this would help.

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 (e.g. through swimming, perspiration, or rain,) and needs reapplying, between washing.

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

(i) 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 scar formation in people affected by CCCA.

(ii) 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 still result in a visible scar.

Self care (What can I do?)

- Join a hair loss support group
- Seek unbiased medical help and be wary of online solutions, especially those that offer instant, or quick, remedies.
- Avoid traumatic hair practices such as tight braids, weaves, or extensions.
- Avoid exposing hair to excessive heat (such as hooded dryers, blow dryers, hot combs, and flat irons), chemicals (such as hair relaxers) 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 long periods.

Where can I get more information about Central Centrifugal Cicatricial Alopecia?
DermNet NZ:
https://dermnetnz.org/topics/central-centrifugal-cicatricial-alopecia/

Other websites you may find helpful:

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

Cicatricial Alopecia Research Foundation: http://www.carfintl.org/

Links to patient support groups:

Alopecia UK. Website: www.alopecia.org.uk

E-mail: mailto:info@alopecia.org.uk

For details of source materials used please contact the Clinical Standards Unit (clinicalstandards@bad.org.uk).

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: 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

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