Medical and Environmental Risk Factors for the Development of Central Centrifugal Cicatricial Alopecia

A Population Study

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Objective: To investigate medical and environmental risk factors for central centrifugal cicatricial alopecia (CCCA), the most common type of scarring alopecia in African American women.

Design: A population study involving a quantitative cross-sectional survey of risk factors for CCCA. Survey results are then correlated with a clinical evaluation for CCCA using a standardized, previously published central scalp alopecia photographic scale.

Setting: Two African American churches and a health fair for African American women in Cleveland, Ohio.

Participants: A total of 326 African American women who participated in the hair study.

Main Outcome Measures: Prevalence of CCCA in the general African American population and risk factors associated with CCCA.

Results: Of the 326 responders, 28% received a grade of 2 or higher using a standardized, previously published central scalp alopecia photographic scale, a score consistent with clinically evident central hair loss. Advanced central hair loss with clinical signs of scarring (grade ≥3) was seen in 59% of these respondents and was interpreted as clinically consistent with CCCA. Diabetes mellitus type 2 was significantly higher in those with CCCA (P = .01), as were bacterial scalp infections (P = .045) and hair styles associated with traction (eg, from braids and weaves) (P = .02).

Conclusions: Our survey results suggest that there is a high prevalence of central hair loss among African American women. Hair styles causing traction as well as inflammation in the form of bacterial infection may be contributing to the development of CCCA. The increase in diabetes mellitus type 2 among those with CCCA is in line with the recent theory that cicatricial alopecia may be a manifestation of metabolic dysregulation.


CENTRAL CENTRIFUGAL CICATRICAL ALOPECIA (CCCA) is a term coined by the North American Hair Research Society (NAHRS) to describe a scarring hair loss, centered on the vertex of the scalp, that spreads peripherally. It is almost exclusively used to describe this type of hair loss in African American women and replaces previously used terms such as hot comb alopecia, coined by Lopresti et al in 1968 and follicular degeneration syndrome, coined by Sperling and Sau in 1992. It is thought to be the most common pattern of scarring hair loss seen in African American women, yet so little is known about its true prevalence among them. Moreover, the etiology and risk factors, including environmental, medical, and genetic risk factors, remain to be elucidated.

Most of the prevalence data on this entity come from Khumalo et al, who have conducted population studies in Africa looking at hairdressing and the prevalence of scalp diseases commonly seen in black children and adults. They found a surprisingly low prevalence of CCCA (1.9%) in adults, most of whom were older than 50 years, and no CCCA in children, a surprising finding given that the common hair grooming practices, such as chemical relaxer use and braids linked to CCCA in African American women, are also used in this African population. In contrast, African American women in the United States commonly present with this entity, but true prevalence data in this population are lacking.

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Lopresti et al were the first to propose an environmental risk factor, namely, the use of hot combs in combination with scalp oil to straighten the naturally curly African hair in order to increase manageability. Since then, most other forms of hair grooming methods used by African Americans, including the use of braids, weaves, and chemical relaxers, have been linked to the development of CCCA. Gathers and Lim found an association between CCCA and hair weaves and braids but not relaxers. Khumalo et al, however, reported 5 cases of acute chemical relaxer–associated scarring hair loss in African women. Bulengo-Ransby and Bergfeld also reported a case of chemical relaxer–associated scarring hair loss. There are other reports of relaxers causing clinically significant chemical burns and hair loss, but it is unclear if those affected went on to develop CCCA. Thus, it is still uncertain whether hair grooming practices are a risk factor.

While several studies have addressed environmental risk factors by examining hair grooming practices, few have addressed medical and genetic risk factors for this disorder. Given that CCCA has a distinct clinical presentation and seems to predominantly affect African Americans, it is important to ask whether risk factors unique to this population might contribute to the pathogenesis of this disorder. Could CCCA be linked to other common medical conditions found in African Americans, such as diabetes mellitus (DM), and other autoimmune conditions, such as lupus? Could it be linked to common skin conditions found in this population, such as hypertrophic scars and keloids, fungal and bacterial scalp infections, and seborrheic dermatitis? Is there a family history of this type of hair loss in patients with CCCA? These are important questions that remain unanswered. Given the lack of epidemiologic data, the main goal of this study was to elucidate environmental as well as medical risk factors that may be associated with CCCA as well as to estimate the prevalence of this disorder in African Americans.

### METHODS

This study, which was approved by the Cleveland Clinic’s institutional review board, involved the administration of a questionnaire about risk factors for CCCA to 326 African American women at 2 churches and a health fair for African American women at the Cleveland Clinic. The questionnaire consisted of questions about demographic data, such as age and participants’ level of education; questions about genetic susceptibility, such as family history of male- and female-pattern hair loss; questions about medical history, such as personal history of bacterial and fungal infections, autoimmune conditions, such as thyroid disease and DM; and questions about hormonally driven conditions, such as unwanted and excessive hair growth, acne, and difficulty conceiving (eTable, http://www.archdermatol.com). Finally, data about methods of hair grooming, such as the age at which chemical relaxers were first used and use of braids and weaves, were compiled and analyzed to determine if there was any association with CCCA.

The questionnaire was followed by a scalp examination using the standardized central scalp alopecia photographic scale to grade hair loss (previously published by Olsen et al). Using this scale, Olsen et al interpreted a central hair loss grade (CHLG) of 0 as normal hair; a CHLG of 1 to 2 as possibly early CCCA, androgenetic alopecia, or telogen effluvium; and a CHLG of 3 to 5 as probable CCCA given the usual characteristics of scarring such as a shiny scalp and loss of follicular ostia found in this group. The scalp examination was conducted by dermatologists well trained in the area of hair loss, assessing for the characteristics of scarring. Standardized photographs of participants’ central scalp and anterotemporal scalp were obtained using professional photography. Validation of the CHLGs assigned by each evaluator was ensured through a follow-up group review of the scalp photographs. Answers to the questionnaire were compared with the CHLG to determine if there was a relationship between the development of CCCA and these various risk factors. The data were analyzed using frequencies and percentages. Spearman correlation was used for associations; t tests, χ² tests, and Fisher exact test were used when appropriate.

### RESULTS

#### DEMOGRAPHIC DATA AND CLINICAL FINDINGS

There were 326 African American responders, with a mean age of 50 years (Table 1). From the 326 responders, 16 were excluded from analysis because of a history of areata, lupus, and/or other hair loss pattern that was consistent with either of these types of hair loss. The NAIRS central hair loss scale was used to grade central hair loss in all responders. A CHLG of 0 was interpreted as normal central hair density without obvious hair loss. A CHLG of 1 was interpreted as minimal central hair loss and thus unlikely to have CCCA; a CHLG of 2 was interpreted as clinically evident central hair loss and possibly early evolving CCCA, although a diagnosis of androgenetic alopecia must be entertained. A CHLG of 3 to 5 was interpreted to be consistent with clinically evident CCCA (Figure 1 and Figure 2). A total of 86 of 310 respondents (28%) received a CHLG of 2 or higher using this scale, a score consistent with clinically evident central hair loss. Central centrifugal cicatricial alopecia (CHLGs 3-5) was seen in 59% of these respondents (Table 1).

#### SYSTEMIC METABOLIC DISEASE

There was a low prevalence of type 2 DM in this population (8%) but a statistically significant increase (P = .01) in the prevalence of type 2 DM was observed in those

<p>| Table 1. Central Hair Loss Grades (CHLGs) at Examinationa |
|---------------------------------|----------------|------------|</p>
<table>
<thead>
<tr>
<th>Physician-assigned CHLGs</th>
<th>Respondents, No. (%) of Total Population</th>
<th>Mean Age, y</th>
</tr>
</thead>
<tbody>
<tr>
<td>0-1</td>
<td>224 (72)</td>
<td>40</td>
</tr>
<tr>
<td>2-5</td>
<td>86 (28)</td>
<td>53</td>
</tr>
<tr>
<td>3-5</td>
<td>52 (17)</td>
<td>58</td>
</tr>
</tbody>
</table>

*Grades: 0, no central scalp hair loss; 1, minimal central scalp loss; 2, clinically evident central scalp hair loss; 3-5, advanced central scalp hair loss.*
with CCCA (CHLGs 3-5) when comparing those with no CCCA (CHLGs 0-1). Type 2 DM was defined in the questionnaire as requiring diet modification or medication for control. Data on weight or body mass index, which is highly correlated with type 2 DM and also a large issue among African American women, were not collected. Only 9% of the study population reported thyroid abnormalities. Most of them (74%) were classified as having minimal central hair loss (CHLGs 0-1); however, there was no statistically significant difference when this group is compared with those with advanced CCCA ($P = .99$) (Table 2 and Figure 3).

**INFECTION**

There was a low prevalence of bacterial skin infections (11.4%), but there was a statistically significant increase in bacterial skin infections in those with CCCA compared with those without CCCA. There was no such trend with fungal infections of the scalp, ringworm, or vaginal yeast infections (Table 2 and Figure 4).
HORMONAL DYSREGULATION

There was also an increased rate of adult acne and difficulty conceiving in those with CCCA (CHLGs 3-5) compared with those without CCCA (CHLGs 0-1), but this was not statistically significant (data not shown) (Table 2).

HORMONAL DYSREGULATION

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HAIR GROOMING

Of the 310 respondents, 286 (91%) relaxed their hair by chemical means. All participants began using chemical relaxer at an early age: 10.3 years on average for those with a CHLG of 0 to 2, and 13.0 years on average for those with a CHLG of 3 to 5. Also, all participants had onset of hair loss at a later age, with average ages of 44.9 years for those with a CHLG of 0 to 2 and 45.5 for those with a CHLG of 3 to 5 (Table 3 and Figure 5). Ninety-four percent of those with clinically evident central hair loss (CHLG 3-5) had a history of chemical relaxer use. There was no statistically significant difference (P = .39) in the use of chemical relaxer in those without central hair loss (CHLGs 0-1) and those with clinically significant central hair loss (CHLGs 3-5).

There was an overrepresentation of use of hot comb and traction hair styles in those with clinically evident central hair loss (CHLG 3-5) compared with those with minimal to no central hair loss (CHLGs 0-1). There was no statistically significant difference (P = .35) in the use of hot comb in those without central hair loss (CHLGs 0-1) and those with CCCA (CHLGs 3-5). There was a statistically significant (P = .02) increase in the use of traction hair styles (braids, weaves) in those with CCCA compared with those with less severe central hair loss (CHLG 2).

FAMILY HISTORY OF HAIR LOSS

Participants were asked to estimate central hair loss in their male relatives using the Norwood pictorial male-pattern baldness scale. Males relatives included participants’ fathers and maternal and paternal grandfathers. They were also asked to grade central hair loss in their female relatives using the central hair loss scale developed.
oped by Olsen et al. Female relatives included participants' mothers and maternal and paternal grandmothers. There was a statistically significant increase in central hair loss of the participant's maternal grandfather among those who had CCCA (CHLGs 3-5) (data not shown).

**SCAR FORMATION AND CCCA**

Given the increase in keloid formation among African Americans and the fact that CCCA is a scarring disorder, participants were asked about their history of thick scars (keloids). Looking at this variable, only 6% of participants reported a history of thick scars, and it was not significantly increased in those with CCCA (CHLGs 3-5) (data not shown).

**DERMATITIS AND CCCA**

Participants were asked about their history of seborrheic dermatitis (scaling on the scalp) and eczema (atopic dermatitis) as well as the development of contact dermatitis from chemical relaxers, shampoos, and conditioners. All groups, regardless of CHLG, reported high incidence of seborrheic dermatitis (24% of total participants). This also held true for dermatitis from relaxer use (9%), which was highly reported in all groups. Eczema was reported by 13% of the participants and seemed to be inversely related to CCCA with most having little or no hair loss (Table 2).

**COMMENT**

Central centrifugal cicatricial alopecia is the most common pattern of scarring hair loss seen in African American women, yet so little is known about the etiology and risk factors associated with this condition. Furthermore, the historical association of CCCA with hot comb use is weak at best given the fact that there have been few epidemiological studies to back this claim. In conducting this study, the main goal was to elucidate environmental as well as medical and genetic risk factors that may be associated with CCCA as well as estimate the prevalence of this disorder.

The results of this study suggest that hair grooming practices that cause traction, such as weaves and braids, may be contributing to the development of CCCA because these styles are more commonly used in those with the most severe central hair loss to increase hairstyle versatility while camouflaging hair loss (Table 2 and Figure 5B). This has some clinical bearing because traction can clinically produce folliculitis of the scalp, which can cause scarring if this inflammation is prolonged. Given the fact that many African American women pay hundreds of dollars to have their hair braided and weaved, they often maintain these hair styles for weeks to months at a time to justify the money spent. The resulting prolonged traction can produce chronic folliculitis, which can eventually lead to scarring.

The relationship between chemical relaxer use and the development of CCCA continues to be murky. While it is clear that chemical relaxers weaken the hair shaft, which can result in increased breakage, it is unclear whether it is related to the development of CCCA. The fact that most African Americans use chemical relaxers in combination with braiding and other hair grooming practices makes it even more difficult to tease out a relationship. Moreover, it is difficult to find a chemical relaxer–naive comparison group because of the early age at which African American children have their hair chemically relaxed and braided (Table 3). A younger population, however, would still be legitimate as a comparison group because presumably they have had fewer years of exposure than their adult counterparts. Overall, we feel that it is not unreasonable to assume that the scalp may absorb some of the caustic chemicals found in relaxers and in time lead to damage of the scalp in the form of scarring.
Our study also demonstrates that inflammation in the form of bacterial infection and acne may also be contributing to the development of CCCA, a finding consistent with the histopathologic characteristics of this disease, which show a lymphocytic perifollicular infiltrate in its early stages. The fact that fungal scalp infection was not associated with CCCA was surprising given how prone this population is to fungal infection.13

One of the most surprising findings of our data was the overrepresentation of type 2 DM in those with CCCA. This group demonstrated that peroxisome proliferator of activated t-cell receptor gamma (PPAR-gamma), a transcription factor important in lipid metabolism by sebaceous glands was aberrant in the hair follicles of patients with the scarring hair disorder, lichen planopilaris.14-16 These are important data that need further study because CCCA may be a marker of metabolic dysfunction and, when present, can prompt clinicians to do further testing for DM in those affected.

On the one hand, genetics may play a role, as is demonstrated by the fact that a history of male-pattern baldness in the maternal grandfather was found to be a risk factor for CCCA. On the other hand, hormonal dysregulation was not a risk factor. This study has raised many interesting questions about the risk factors for CCCA, and further studies are needed to explore these associations.

This study has several limitations. Because this is a survey, self-report bias must be considered. Also, no biopsy specimens were taken to confirm the diagnosis of CCCA, a pattern of scarring hair loss that can be difficult to differentiate clinically from female-pattern hair loss. However, the diagnosis of CCCA is often made clinically without biopsy; thus, this study reflects current clinical practice. Dermatoscopy may assist in clinical diagnosis. Moreover, the age of presentation of CCCA and female-pattern hair loss overlaps; thus, it may be that CCCA is being overdiagnosed clinically, especially when the patient is African American. Again, this potential limitation reflects current practice. Also, an assumption is made that the population in this survey represents the general African American population, but this may not be true, as is evidenced by the demographic data showing that most of those surveyed achieved a higher level of education (some college education or higher) than the average American.

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Author Contributions: All authors had full access to all of the data in the study and take responsibility for the integrity of the data and the accuracy of the data analysis. Study concept and design: Kyei and Bergfeld. Acquisition of data: Kyei, Bergfeld, Piliang, and Summers. Analysis and interpretation of data: Kyei and Bergfeld. Drafting of the manuscript: Kyei, Bergfeld, and Piliang. Critical revision of the manuscript for important intellectual content: Kyei, Bergfeld, and Summers. Statistical analysis: Kyei.

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REFERENCES