

# Central Centrifugal Cicatricial Alopecia in Males: Analysis of Time to Diagnosis and Disease Severity

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## PRACTICE POINTS

- Most males with central centrifugal cicatricial alopecia (CCCA) experience considerable diagnostic delays and typically present to dermatology with late-stage disease.
- Dermatologists should consider CCCA in the differential diagnosis for adult Black males with alopecia.
- More research is needed to explore advanced CCCA in males, including factors limiting timely diagnosis and the impact on quality of life in this population.

To the Editor:

Central centrifugal cicatricial alopecia (CCCA) is a chronic progressive type of scarring alopecia that primarily affects women of African descent.<sup>1</sup> The disorder rarely is reported in men, which may be due to misdiagnosis or delayed diagnosis. Early diagnosis and treatment are the cornerstones to slow or halt disease progression and prevent permanent damage to hair follicles. This study aimed to investigate the time to diagnosis and disease severity among males with CCCA.

We conducted a retrospective chart review of male patients older than 18 years seen in outpatient clinics

at an academic dermatology department (Philadelphia, Pennsylvania) between January 2012 and December 2022. An electronic query using the *International Classification of Diseases, Ninth and Tenth Revisions*, code L66.9 (cicatricial alopecia, unspecified) was performed. Patients were included if they had a clinical diagnosis of CCCA, histologic evidence of CCCA, and scalp photographs from the initial dermatology visit. Patients with folliculitis decalvans, scalp biopsy features that limited characterization, or no scalp biopsy were excluded from the study. Onset of CCCA was defined as the patient-reported start time of hair loss and/or scalp symptoms. To determine alopecia severity, the degree of central scalp hair loss was independently assessed by 2 dermatologists (S.C.T., T.O.) using the central scalp alopecia photographic scale in African American women.<sup>2,3</sup> This 6-point photographic scale displays images with grades ranging from 0 (normal) to 5 (bald scalp); higher grades indicate probable and more severe CCCA. The scale also divides the central hair loss in a frontal-accentuation or vertex-predominant pattern, which corresponds to the A or B designations, respectively; thus, a score of 5A indicates probable severe CCCA with a frontal accentuation pattern, while 5B indicates probable severe CCCA with hair loss focused on the vertex scalp. This study was approved by the University of Pennsylvania institutional review board (approval #850730).

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Cutis. 2024 June;113(6):246-248. doi:10.12788/cutis.1031

Of 108 male patients, 12 met the eligibility criteria. Nearly all patients (91.7% [11/12]) had a CCCA severity grade of 3 or higher at the initial dermatology visit, indicating extensive hair loss (Table). The clinical appearance of severity grades 2 through 5 is demonstrated in the Figure. Among patients with a known disease duration prior to diagnosis, 72.7% (8/11) were diagnosed more than 1 year after onset of CCCA, and 45.4% (5/11) were diagnosed more than 5 years after onset. On average (SD), it took 6.4 (5.9) years for patients to receive a diagnosis of CCCA after the onset of scalp symptoms and/or hair loss.

Randomized controlled trials evaluating treatment of CCCA are lacking, and anecdotal evidence posits a better treatment response in early CCCA; however, our results suggest that most male patients present with advanced CCCA and receive a diagnosis years after disease onset. Similar research in alopecia areata has shown that 72.4% (105/145) of patients received their diagnosis within a year after onset of symptoms, and the mean time from onset of symptoms to diagnosis was 1 year.<sup>4</sup> In contrast, male patients with CCCA experience considerable diagnostic delays. This disparity indicates the need for clinicians to increase recognition of CCCA in men and quickly refer them to a dermatologist for prompt treatment.

Androgenetic alopecia (AGA) commonly is at the top of the differential diagnosis for hair loss on the vertex of the scalp in males, but clinicians should maintain a high index of suspicion for CCCA, especially when scalp symptoms or atypical features of AGA are present.<sup>5</sup> Androgenetic alopecia typically is asymptomatic, whereas the symptoms of CCCA may include itching, tenderness, and/or burning.<sup>6,7</sup> Trichoscopy is useful to evaluate for scarring, and a scalp biopsy may reveal other features to lower AGA on the differential. Educating patients, barbers, and hairstylists about the importance of early intervention also may encourage earlier visits before the scarring process is advanced. Further exploration into factors impacting diagnosis and CCCA severity may uncover implications for prognosis and treatment.

### Time to Diagnosis and Disease Severity in Males With CCCA

Patient no.	Time from CCCA onset to diagnosis	CCCA severity score <sup>a</sup>	
		Reviewer 1	Reviewer 2
1	1 y 10 mo	4B	4B
2	5 y	2A	2A
3	9 mo	3A	3A
4	Unknown	4B	4B
5	3 y	4B	4A
6	9 y	3A	3B
7	12 y	5B	5B
8	6 y	3B	3B
9	8 mo	3B	3B
10	1 y	4B	4A
11	15 y 2 mo	5A/B	5A/B
12	16 y 6 mo	4B	3B

Abbreviation: CCCA, central centrifugal cicatricial alopecia.

<sup>a</sup>The 6-point photographic scale displays images with scores ranging from 0 (normal) to 5 (bald scalp); higher grades indicate probable and more severe CCCA. The scale also divides the central hair loss in a frontal-accentuation or vertex-predominant pattern, which corresponds to the A or B designations, respectively; thus, a score of 5A indicates probable severe CCCA with a frontal accentuation pattern, while 5B indicates probable severe CCCA with hair loss focused on the vertex scalp.



A–D, Clinical appearance of central centrifugal cicatricial alopecia grades 2A, 3A/B, 4B, and 5B, respectively, based on comparison of the patients' hair loss to the images in the scale.

This study was limited by a small sample size, retrospective design, and single-center analysis. Some patients had comorbid hair loss conditions, which could affect disease severity. Moreover, the central scalp alopecia photographic scale<sup>2</sup> was not validated in men or designed for assessment of the nonclassical hair loss distributions noted in some of our patients. Nonetheless, we hope these data will support clinicians in efforts to advocate for early diagnosis and treatment in patients with CCCA to ultimately help improve outcomes.

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