Research letter

Frontal Fibrosing Alopecia Severity Index (FFASI): a validated scoring system for assessing frontal fibrosing alopecia

DOI: 10.1111/bjd.14445

Dear Editor, The incidence of frontal fibrosing alopecia (FFA) appears to be increasing,[1–6] and response to treatment has been largely disappointing.[1–3] However, assessment of treatment interventions is confounded by slow disease progression and lack of robust means of assessing disease severity and activity. To address the latter, we have developed a validated clinical scoring system – the Frontal Fibrosing Alopecia Severity Index (FFASI), which provides a standardized framework for FFA assessment and patient stratification.

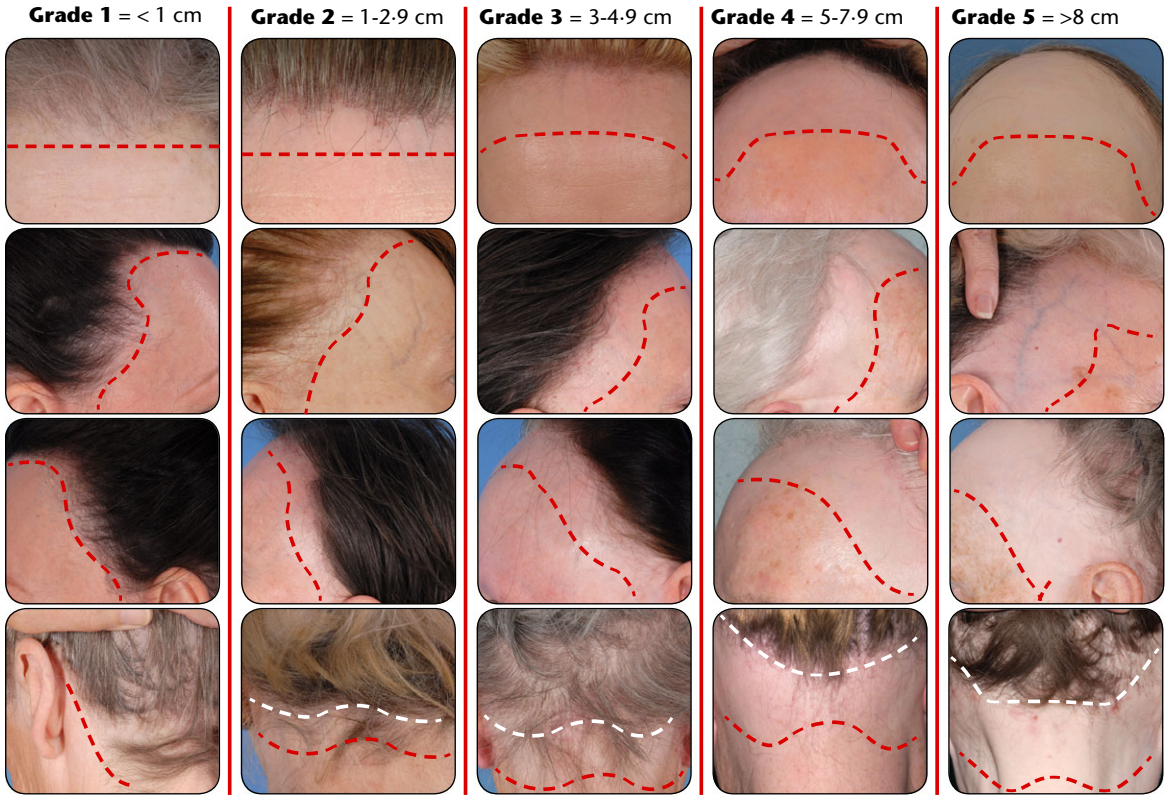
A British Hair and Nail Society (BHNS) subgroup considered clinical methods of assessing FFA severity and activity. In agreement with other authors,[4,7] assessment of alopecia bandwidth was deemed the most appropriate and objective measurement of severity, with changes in extent over time reflecting disease activity. FFASI was compiled in two forms: FFASI and FFASI B (Fig. 1). FFASI utilizes clinical images of the entire hairline, divided into four sections. Alopecia severity is graded 1–5 based on hairline recession, similarly to the criteria proposed by Vañó-Galván *et al.*[4] In order that hairline recession comprises the greatest proportion of the assessment, each grade is weighted. Although of uncertain significance,[1,2,8] frontal band inflammation is also assessed. Nonscalp hair loss[5] (eyebrow, eyelash, limb and flexural) are scored, as are associated features (facial papules;[2,9] cutaneous,[2,4] nail[10] and mucosal lichen planus;[1,2,4] and generalized scalp lichen planopilaris).[1,4] Scores for hairline recession, inflammatory band, nonscalp loss and associated features may be combined to give a maximum score of 100. FFASI B uses the same format, but rather than grading alopecia it permits user-defined measurement of each hairline section. FFASI B was not validated in this exercise.

FFASI validation was undertaken by two methods. Firstly, the clinical images used in FFASI were evaluated by a panel of 11 BHNS consultant dermatologists. Each graded 30 FFA patient photographs using FFASI. The exercise was undertaken twice to assess intraobserver agreement. Secondly, a clinical assessment of three patients with FFA was undertaken by six dermatologists – three consultant dermatologists (two with alopecia special interest), two dermatology trainees and a staff grade – using FFASI. Each patient was examined on two occasions by each dermatologist. Assessors were instructed to grade to the main hairline, not to 'lonely' hairs,[11] and, where the bandwidth was not uniform, to grade to the most representative image. Inter- and intraobserver agreement were assessed using Kendall's coefficient of concordance *W*. Values range from 0 (no agreement) to 1 (complete agreement), and agreement levels were classified according to Schmidt.[12] Where assessment ratings resulted in two or fewer categories, kappa statistics were computed. Calculations were performed using Minitab v17 (Minitab Inc., Coventry, U.K.).

For assessment of patient photographs using FFASI, intraobserver concordance showed strong to very strong agreement for all hairline areas, indicating consistency in assessments by individual consultants (Table S1a; see Supporting Information). The results of interobserver agreement indicated that overall agreement between consultants was very strong, with all values $> 0{\cdot}85$ for each hairline area assessed (Table S1b; see Supporting Information). Thus, all consultants consistently assessed FFA patient photographs using FFASI. In the clinical evaluation, Kendall's coefficient demonstrated that intraobserver reliability was very strong for frontal, right, left and posterior hairlines, and frontal band assessments (Table S2a; see Supporting Information). Scores for flexural hair loss were strong to very strong. Concordance for eyebrow and eyelash scores showed complete agreement; however, limb scores showed poorer agreement for assessors without an alopecia special interest. The results for interobserver agreement for clinical assessments showed very strong agreement between observers for frontal, right and left hairlines and eyebrows (Table S2b; see Supporting Information). For the posterior hairline and frontal band, values were slightly above 0·5, indicating moderate agreement. Agreement for eyelash and limb assessment was poorer, with kappa values of 0·037 and 0·345, respectively. However, concordance between consultants with an alopecia interest was very strong, suggesting that experience in clinical assessment resulted in greater consistency.

We have developed a validated scoring system for FFA assessment. FFASI permits assessment of the entire hairline, inflammatory frontal band, facial and body hair loss, and associated features. FFASI is weighted in favour of hairline assessment, as alopecia is the principle feature. However, a total score out of 100 can be calculated, representing global disease severity. Although FFA was initially considered a scalp disorder, both facial and body hair are frequently lost,[5] and this may sometimes predate onset of scalp loss.[4] Facial vellus follicle involvement results in facial papules.[2,9]

Frontal Fibrosing Alopecia Severity Index (FFASI)



Date														
Scalp margin	**Grade: 1 - 5** **Frontal Band:**													
	No loss = Score 0 Grade 1 = Score 4 Grade 2 = Score 8 Grade 3 = Score 12 Grade 4 = Score 16 Grade 5 = Score 20 Score 0 if not inflamed, normal density; Score 2 if inflamed or reduced density; Score 4 if inflamed and reduced density													
Frontal														
R lateral														
L lateral														
Posterior														
Frontal Band														
Total	/84	/84	/84	/84	/84	/84	/84	/84	/84	/84	/84	/84	/84	/84
Other Hair loss	**No loss Score 0** **Partial loss Score 1** **Complete loss Score 2**													
eyebrow loss														
eyelash loss														
flexural loss (axillary, pubic)														
upper limb hair loss														
lower limb hair loss														
Additional features	**Absent Score 0** **Present Score 1**													
typical scalp LPP														
facial papules														
cutaneous LP / LP variants														
oral mucosal LP														
genital mucosal LP														
nail LP														
Total	/16	/16	/16	/16	/16	/16	/16	/16	/16	/16	/16	/16	/16	/16
Combined Total	/100	/100	/100	/100	/100	/100	/100	/100	/100	/100	/100	/100	/100	/100

Fig 1. Frontal Fibrosing Alopecia Severity Index (FFASI).

FFASI B

Date														
Scalp margin	**Measurement** **Frontal Band:** **Score 0 if not inflamed, normal density; Score 2 if inflamed or reduced density; Score 4 if inflamed and reduced density**													
Frontal														
R lateral														
L lateral														
Occipital														
Frontal Band														
Total														
Other Hair loss	**No loss Score 0**			**Partial loss Score 1**			**Complete loss Score 2**							
eyebrow loss														
eyelash loss														
flexural loss (axillary, pubic)														
upper limb hair loss														
lower limb hair loss														
Additional features	**Absent Score 0**			**Present Score 1**										
typical scalp LPP														
facial papules														
cutaneous LP / LP variants														
oral mucosal LP														
genital mucosal LP														
nail LP														
Total	/16	/16	/16	/16	/16	/16	/16	/16	/16	/16	/16	/16	/16	/16

Definitions

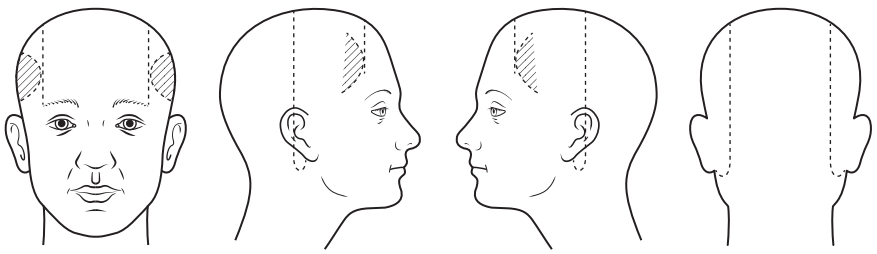
Scalp Margins

Frontal:
Intertemporalis

R lateral:
R mastoid to anterior
R temporalis

L lateral:
L mastoid to anterior
L temporalis

Posterior:
Intermastoid



Notes

mi • 259134

Fig 1. continued.

Cutaneous, mucosal and nail lichen planus and generalized scalp lichen planopilaris are infrequently associated.[1,2,4,10] The natural history is unclear and it is uncertain how the condition progresses. Involvement of the frontal hairline seems universal.[4] Loss of eyelashes and facial papules are associated with more severe disease.[4]

FFA treatments need to be assessed by clinical trials. Many treatments have been used, but as evidence is weak (no randomized controlled trials, variable outcome measures etc.), it is difficult to assess superiority of efficacy.[7,13] To have confidence in trial results, a standardized, validated and objective assessment method is required. To date, several nonstandardized and nonvalidated methods have been used. The most frequent method is measurement from the nasal crease to the frontal hairline or other forehead/frontal hairline measures.[1,2] Although helpful for measuring change in a patient over time, this is less helpful when comparing between patients due to differing premorbid hairline positions. Detailed photographic images are an accurate means of monitoring disease; however, they do not permit statistical analysis.

The Lichen Planopilaris Activity Index was devised as an assessment tool for lichen planopilaris activity.[14] It includes scoring of symptoms and signs of inflammation, positive anagen pull and disease spreading, with results calculated using a devised formula. However, it has been criticized for being based on subjective data calculated using an arbitrary formula,[15] and gives no account of the extent of hair already lost. FFASI offers a more complete assessment of the hairline than point measurement(s) and provides numerical data that can be analysed statistically. It does not rely unduly on measures of uncertain significance (symptoms, erythema or anagen pull), but measures the cardinal disease feature: extent of alopecia. Additionally, it allows global disease assessment by including facial and body hair and associated features.

Change in FFASI grade over time reflects disease activity, and the standardized format allows comparison between patients. One weakness of FFASI is that it relies upon a 'best-fit' model for grading alopecia bandwidth: bands of recession are not entirely uniform and clinical judgement is required. However, more precise assessment can be made by recording actual bandwidth measurements using FFASI B.

In conclusion, we have a developed a validated scoring system for FFA that allows global disease assessment for individuals over time and permits comparison between patients.

Acknowledgments

We would like to thank the members of the BHNS for their contribution to the development of the FFASI and Mr Alun Bevan, Medical Illustration Department, Glasgow Royal Infirmary. We would also like to thank the patients and clinical colleagues who participated in the clinical assessment exercise.

S. Holmes[1]
T. Ryan[2]
D. Young[2]
M. Harries[3]
On behalf of the British Hair and Nail Society

[1]*Alan Lyell Centre for Dermatology, Queen Elizabeth University Hospital, 1345 Govan Road, Glasgow G51 4TF, U.K.*
[2]*Department of Mathematics and Statistics, University of Strathclyde, Glasgow, U.K.*
[3]*The Dermatology Centre, The University of Manchester, Salford Royal NHS Foundation Trust, Manchester, U.K.*
E-mail: susan.holmes2@ggc.scot.nhs.uk

References

1 MacDonald A, Clark C, Holmes S. Frontal fibrosing alopecia: a review of 60 cases. *J Am Acad Dermatol* 2012; **67**:955–61.
2 Tan KT, Messenger AG. Frontal fibrosing alopecia: clinical presentations and prognosis. *Br J Dermatol* 2009; **160**:75–9.
3 Ladizinski B, Bazakas A, Selim MA, Olsen EA. Frontal fibrosing alopecia: a retrospective review of 19 patients seen at Duke University. *J Am Acad Dermatol* 2013; **68**:749–55.
4 Vañó-Galván S, Molina Ruiz AM, Serrano Falcon C *et al.* Frontal fibrosing alopecia: a multicentre review of 355 patients. *J Am Acad Dermatol* 2014; **70**:670–8.
5 Chew AL, Bashir SJ, Wain EM *et al.* Expanding the concept of frontal fibrosing alopecia: a unifying concept. *J Am Acad Dermatol* 2010; **63**:653–60.
6 Griffen LL, Michaelides C, Griffiths CEM *et al.* Primary cicatricial alopecias: a U.K. survey. *Br J Dermatol* 2012; **167**:692–705.
7 Rácz E, Cho C, Moorman PW *et al.* Treatment of frontal fibrosing alopecia and lichen planopilaris: a systematic review. *J Eur Acad Dermatol Venereol* 2013; **27**:1461–70.
8 Fernández Crehuet P, Rodrigues Barata AR, Vañó-Galván S *et al.* Trichoscopic features of frontal fibrosing alopecia: results in 249 patients. *J Am Acad Dermatol* 2015; **72**:357–9.
9 Donati A, Molina L, Doche I *et al.* Facial papules in frontal fibrosing alopecia. *Arch Dermatol* 2011; **147**:1424–7.
10 MacPherson M, Hohendo RF, Ansari P, Trueb RM. Nail involvement in frontal fibrosing alopecia. *Int J Trichology* 2015; **7**:64–6.
11 Tosti A, Miteva M, Torres F. Lonely hair: a clue to the diagnosis of frontal fibrosing alopecia. *Arch Dermatol* 2011; **147**:1240.
12 Schmidt RC. Managing Delphi surveys using nonparametric statistical techniques. *Decision Sci* 1997; **28**:763–74.
13 Harries MJ, Messenger A. Treatment of frontal fibrosing alopecia and lichen planopilaris. *J Eur Acad Dermatol Venereol* 2014; **28**:1404–5.
14 Chiang C, Sah D, Cho B *et al.* Hydroxychloroquine and lichen planopilaris: efficacy and introduction of Lichen Planopilaris Activity Index scoring system. *J Am Acad Dermatol* 2010; **62**:387–92.
15 Sperling LC, Nguyen JV. Commentary: treatment of lichen planopilaris. *J Am Acad Dermatol* 2010; **62**:398–400.

Supporting Information

Additional Supporting Information may be found in the online version of this article at the publisher's website:

Table S1. Kendall's coefficients of concordance *W* for intraobserver assessments of frontal, right lateral and left lateral hairline images (a) and for interobserver assessment of frontal, right lateral and left lateral hairline images (b) using the Frontal Fibrosing Alopecia Severity Index.

Table S2. Kendall's coefficients of concordance *W* for intraobserver clinical assessments (a) and for interobserver clinical assessments (b) using the Frontal Fibrosing Alopecia Severity Index.

Funding sources: none.

Conflicts of interest: none declared.