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Frontal fibrosing alopecia severity index (FFASI): a validated scoring system for assessing frontal fibrosing alopecia

Running title: Frontal fibrosing alopecia severity index (FFASI)

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The incidence of frontal fibrosing alopecia (FFA) appears to be increasing¹⁻⁶ and response to treatment has been largely disappointing¹⁻³. 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 FFA severity index (FFASI), which provides a standardised 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 band width 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 (figure 1). FFASI utilises clinical images of the entire hairline, divided into 4 sections. Alopecia severity is graded 1-5 based on hairline recession similar to criteria proposed by Vano Galvan⁴. 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. Non-scalp hair loss⁵ (eyebrow, eyelash, limb and flexural) are scored, as are associated features (facial papules^{2,9}; cutaneous^{2,4}, nail¹⁰ and mucosal LP^{1,2,4}; and generalised scalp LPP^{1,4}). Scores for hairline recession, inflammatory band, non-scalp loss and associated features may be combined to give a maximum score of 100. FFASIB uses the same format but rather than grading alopecia, permits user-defined measurement of each hairline section. FFASIB was not validated in this exercise.

FFASI validation was undertaken by two methods. Firstly, the clinical images used in FFASI were evaluated by panel of 11 BHNS consultant dermatologists. Each graded 30 FFA patient photographs using FFASI. The exercise was undertaken twice to assess intra-observer agreement. Secondly, a clinical assessment of 3 FFA patients was undertaken by 6 dermatologists (3 consultant dermatologists (2 with alopecia special interest), 2 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” hairs¹¹ and, where band width was not uniform, to grade to the most representative image. Inter and intra-observer 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 Schmidt¹². Where assessment ratings resulted in 2 or fewer categories, Kappa statistics were computed. Calculations were performed using Minitab(v17).

For assessment of patient photographs using FFASI, intra-observer concordance showed strong to very strong agreement for all hairline areas, indicating consistency in assessments by individual consultants (supplementary table 1a). The results of inter-observer agreement indicated overall agreement between consultants was very strong, with all values >0.85 for each hairline area assessed (supplementary table 1b). Thus, all consultants consistently assessed FFA patient photographs using FFASI. In the clinical evaluation, Kendall's coefficient demonstrated intra-observer reliability was very strong for frontal, right, left and posterior hairlines, and frontal band assessments (supplementary table 2a). 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 inter-observer agreement for clinical assessments showed very strong agreement between observers for frontal, right and left hairlines and eyebrows (supplementary table 2b). For the posterior hairline and frontal band, values were slightly >0.5 , indicating moderate agreement. Agreement for eyelash and limb assessment was poorer with kappa values 0.037 and 0.345 respectively. However, concordance between consultants with an alopecia interest was very strong, suggesting 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. Initially considered a scalp disorder, both facial and body hair are frequently lost⁵ and may sometimes predate onset of scalp loss⁴. Facial vellus follicle involvement results in facial papules^{2,9}. Cutaneous, mucosal and nail LP, and generalised scalp LPP 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 universal⁴. Loss of eyelashes and facial papules are associated with more severe disease⁴.

FFA treatments need to be assessed by clinical trials. Many treatments have been used but as evidence is weak (no RCTs, variable outcome measures etc.), it is difficult to assess superiority of efficacy^{7,13}. To have confidence in trial results, a standardised, validated and objective assessment method is required. To date, several non-standardised and non-validated methods have been used. The most frequent method is measurement from nasal crease to 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 pre-morbid hairline positions. Detailed photographic images are an accurate means of monitoring disease however, they do not permit statistical analysis. LPPAI was devised as an assessment tool for LPP activity¹⁴. It includes scoring of symptoms and signs of inflammation, positive anagen-pull and disease spreading, with results calculated using a devised formula. It has been criticised for being based on subjective data calculated using an arbitrary formula¹⁵, and gives no account of 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 standardised format allows comparison between patients. One weakness of FFASI is that it relies upon a “best-fit” model for grading alopecia band width: bands of recession are not entirely uniform and clinical judgement is required. However, more precise assessment can be made by recording actual band width measurements using FFASI B. In conclusion, we have developed a validated scoring system for

FFA which allows global disease assessment for individuals over time and permits comparison between patients.

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References

1. Holmes S, Clark C, MacDonald A. 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. Vano-Galvan 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, Fenton DA, Stefanato CM. 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, Paus R, Harries MJ. Primary cicatricial alopecias: a UK survey. *Br J Dermatol* 2012;167:692-705
7. Racz E, Cho C, Moorman PW, Noordhoek Hogt V, Neumann HAM. Treatment of frontal fibrosing alopecia and lichen planopilaris: a systematic review. *J EADV* 2013;1461-70
8. Fernandez Crehuet P, Rodrigues Barata AR, Vano Galvan 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, Valente NS, Romiti R. Facial papules in frontal fibrosing alopecia. *Arch Derm* 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 Derm 2011;147:1240

12. Schmidt RC. Managing Delphi Surveys Using Nonparametric Statistical Techniques. Decision Sciences 1997;28:763-74

13. Harries MJ Messenger A. Treatment of frontal fibrosing alopecia and lichen planopilaris. JEADV 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

Frontal Fibrosing Alopecia Severity Index (FFASI)



Date														
Scalp margin	Grade: 1 - 5		No loss - Score 0		Grade 1 - Score 4		Grade 2 - Score 8		Grade 3 - Score 12		Grade 4 - Score 16		Grade 5 - Score 20	
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														
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