

## Alopecia areata investigational assessment guidelines\*

Elise Olsen, MD,<sup>a</sup> Maria Hordinsky, MD,<sup>b</sup> Susan McDonald-Hull, MB, FRCP,<sup>c</sup> Vera Price, MD,<sup>d</sup> Janet Roberts, MD,<sup>e</sup> Jerry Shapiro, MD,<sup>f</sup> and Kurt Stenn, MD<sup>g</sup>  
*Durham, North Carolina; Minneapolis, Minnesota; Pontefract, United Kingdom; San Francisco, California; Portland, Oregon; Vancouver, British Columbia, Canada; Skillman, New Jersey*

### I. PURPOSE

To establish criteria for selecting and assessing subjects for both clinical and laboratory studies of alopecia areata, thereby facilitating collaboration, comparison of data, and the sharing of patient-derived tissue

### II. DEFINITION OF ALOPECIA AREATA

Alopecia areata is a dermatologic disease characterized in its limited form by circumscribed round or oval patches of alopecia with well-demarcated borders between normal and affected scalp. There is no scale or induration of the scalp and no loss of follicular markings. Disease extent may progress from this limited form to complete loss of hair on the scalp and/or body.

### III. INCLUSION CRITERIA

These guidelines apply only to terminal hair loss or growth on the scalp.

These guidelines focus on the major forms of alopecia areata (patchy alopecia areata, alopecia totalis, and alopecia universalis) and their expression on the scalp. The terms *alopecia totalis* and *alopecia universalis* imply 100% scalp hair loss.

Areas of hair loss other than on the scalp may be assessed and documented in the data collected on each patient. (See Section IV. B)

### IV. CRITERIA FOR MEASURING EXTENT OF INVOLVEMENT

Those data items in boldface type should be filled out in toto. Those data items *not* in bold are optional and can be filled out as desired by the investigator.

**A. The proportion of scalp involvement is determined by dividing the scalp into 4 quadrants and estimating the percentage of the scalp surface that all the alopecic areas would occupy if placed together. The following groups will be used:**

**S: Scalp hair loss**

\_\_\_\_\_ **S<sub>0</sub> = No hair loss**

\_\_\_\_\_ **S<sub>1</sub> = ≤ 25% hair loss**

\_\_\_\_\_ **S<sub>2</sub> = 26%-50% hair loss**

\_\_\_\_\_ **S<sub>3</sub> = 51%-75% hair loss**

\_\_\_\_\_ **S<sub>4</sub> = 76%-99% hair loss**

\_\_\_\_\_ **a = 76%-95% hair loss**

\_\_\_\_\_ **b = 96%-99% hair loss**

\_\_\_\_\_ **S<sub>5</sub> = 100% hair loss**

**B. Other areas of alopecia or involvement by alopecia areata may be noted:**

**B: Body hair loss**

\_\_\_\_\_ **B<sub>0</sub> = No body hair loss**

\_\_\_\_\_ **B<sub>1</sub> = Some body hair loss**

\_\_\_\_\_ **B<sub>2</sub> = 100% body (excluding scalp) hair loss**

**N: Nail involvement**

\_\_\_\_\_ **N<sub>0</sub> = No nail involvement**

\_\_\_\_\_ **N<sub>1</sub> = Some nail involvement**

\_\_\_\_\_ **a. Twenty-nail dystrophy/trachyonychia (must be all 20 nails)**

From Duke University Medical Center, Durham<sup>a</sup>; University of Minnesota, Minneapolis<sup>b</sup>; Pontefract General Infirmary<sup>c</sup>; University of California at San Francisco<sup>d</sup>; Northwest Cutaneous Research Specialists, Portland<sup>e</sup>; The University of British Columbia, Vancouver<sup>f</sup>; and Johnson & Johnson, Skin Biology Research Center, Skillman.<sup>g</sup>

Reprint requests: Elise A. Olsen, MD, Professor of Medicine, Duke University Medical Center, Box 3294, Durham, NC 27710.

J Am Acad Dermatol 1999;40:242-6.

\*Developed from the Alopecia Areata Consensus meeting sponsored by the National Alopecia Areata Foundation at the First Tricontinental Meeting of the Hair Research Societies, Brussels, Belgium, Oct 8, 1995. Participants are listed at the end of the guidelines.

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C. Terminology

The term *alopecia totalis* is  $S_5B_0$  only and is 100% scalp terminal hair loss.

The term *alopecia totalis/alopecia universalis* (AT/AU) is  $S_5B_0-B_2$  and implies 100% scalp terminal hair loss with variable body hair loss.

The term *alopecia universalis* is  $S_5B_2$  and is 100% scalp terminal hair and 100% body hair loss.

V. ALOPECIA AREATA DATABASE

A. Essential background data

Patient's initials \_\_\_\_\_

Date of intake \_\_\_\_\_

Age \_\_\_\_\_

Date of birth \_\_\_\_\_

Age at onset of first episode of alopecia areata \_\_\_\_\_

First episode of alopecia areata (month/year of onset) \_\_\_\_\_

Current episode alopecia areata

Age of onset \_\_\_\_\_

Month/year of onset \_\_\_\_\_

Duration of current episode (months) \_\_\_\_\_

Extent of hair loss \_\_\_\_\_ ( $S_0-S_5$ ,  $B_0-B_2$ : as defined in IVA and IVB)

Sex

Male \_\_\_\_\_

Female \_\_\_\_\_

Racial group

American Indian or Alaskan native \_\_\_\_\_

Asian or Pacific Islander \_\_\_\_\_

Black, not of Hispanic origin \_\_\_\_\_

Hispanic \_\_\_\_\_

White, not of Hispanic origin \_\_\_\_\_

Predominant hair color

Black \_\_\_\_\_

Brown \_\_\_\_\_

Red \_\_\_\_\_

Blond \_\_\_\_\_

Gray \_\_\_\_\_

White \_\_\_\_\_

B. Prior history of alopecia areata

Number of prior episodes of alopecia areata \_\_\_\_\_

History of alopecia totalis or alopecia totalis/alopecia universalis at any time

1. > 2 years' duration \_\_\_\_\_

2. ≤ 2 years' duration \_\_\_\_\_

C. Pertinent immediate past history

History of infections within 6 months before onset of hair loss

a. Initial episode of alopecia areata

Site of infection \_\_\_\_\_

Type of infection \_\_\_\_\_

b. Current episode of alopecia areata

Site of infection \_\_\_\_\_

Type of infection \_\_\_\_\_

History of vaccination within 6 months before onset of hair loss

a. Initial episode of alopecia areata

Type of vaccination \_\_\_\_\_

b. Current episode of alopecia areata

Type of vaccination \_\_\_\_\_

Patient's or parent's perception of trigger for hair loss

Initial episode \_\_\_\_\_

Current episode \_\_\_\_\_

D. Patient and family medical history

Patient and family medical history	Patient	Mother	Father	Son	Daughter	Brother	Sister	Maternal Grandmother	Maternal Grandfather	Paternal Grandmother	Paternal Grandfather	Maternal Aunt	Maternal Uncle	Paternal Aunt	Paternal Uncle
Atopic dermatitis															
Allergic rhinitis															
Asthma															
Thyroid disease															
Hashimoto's thyroiditis															
Graves' disease															
Vitiligo															
Diabetes															
Insulin-dependent diabetes															
Non-insulin-dependent diabetes															
Unknown type															
Lupus erythematosus															
Pernicious anemia															
Rheumatoid arthritis															
Ulcerative colitis															
Celiac disease															
Psoriasis															
Other autoimmune disease															
Type															
Down syndrome															
Immunodeficiency															
Type															
Other															

In attendance were Sam Alaiti, MD, Brigitte Almond, MD, Ulrike Blume-Peytavi, MD, Jean-Claude Bystry, MD, Nigel Hibberts, MD, Maria Hordinsky, MD, Satoshi Itami, MD, Vicki Kalabokes, Ulrike Lichti, PhD, Takashi Masuai, MD, Andrew McDonagh, MD, Susan McDonald-Hull, MB, FRCP, Andrew Messenger, MD, Kazoo Murato, MD, Michael Nutbrown, PhD, Roy Oliver, PhD, Elise Olsen, MD, Giovanni Orrechia, MD, Ralf Paus, MD, Vera Price, MD, Valerie Randall, PhD, Janet Roberts, MD, Marty Sawaya, MD, PhD, Jerry Shapiro, MD, Kurt Stenn, MD, John Sundberg, DVM, PhD, Susan Takayasu, MD, Julie Thornton, PhD, Dominique Van Neste, MD, and David Whiting, MD.

## COMMENTARY

Alopecia areata is a disorder with many clinical presentations, from a single patch of hair loss on the scalp or body to complete scalp and/or body hair loss to diffuse scalp hair thinning without discrete patches of loss. The hair loss may be the single obvious clinical abnormality or there may be associated nail abnormalities. Less commonly, there may be evidence of associated conditions (primarily categorized as autoimmune) such as Hashimoto's thyroiditis and vitiligo. Alopecia areata clearly has a genetic component, but the specific genetic linkage may vary with the hair loss phenotype. Other potentially hereditary factors such as atopy may be linked to severity of disease and refractoriness to therapy. The importance of environmental factors in triggering hair loss or modifying the subsequent course is uncertain.

Previously, variable attention was paid to the clinical aspects of patients with alopecia areata whose tissue or blood was used for clinical investigation. However, the type, extent, and duration of hair loss, presence of other clinical abnormalities, genetic factors, and other concurrent or preceding immunologic events may greatly (and without taking these into account, unknowingly) influence the test results. Moreover, clinical studies that have attempted to track the clinical course of alopecia areata in relation to a variety of factors or to determine a response to treatment have been hampered by the lack of a method to clearly depict the type and extent of hair loss that would, in turn, facilitate pooling of data. The guidelines developed and presented herein are a studied attempt to correct this situation for clinical and bench researchers alike by providing a means by which to communicate the specifics of the clinical, genetic, and pertinent environmental factors specific to a study patient with alopecia areata. It is also a first step toward establishing a fruitful alopecia areata database.

To facilitate usage of the aforementioned guidelines, the following clarifications are offered:

1. There are several ways to determine the percentage of scalp hair loss and whichever way is easiest for a particular investigator is fine. However, whatever method is chosen, it should be used uniformly for all patients at a given site. If repetitive assessments are done over time in a given patient, the same investigator should do all of the assessments or if two or more investigators are rating a patient's hair loss at different time

points, then there must previously have been found a good reproducibility of assessments between investigators.

To determine percentage scalp hair loss, one can:

- a. Mentally divide the scalp into 4 quadrants (Fig 1) and estimate the percentage of the scalp surface that all the alopecic areas would occupy if placed together.

- b. Use the visual aid provided for determining percentage hair loss (Fig 2), which gives the investigator a graphic representation of the percentage surface area of the scalp covered by the quadrants demarcated which, taken together, equal 100% of the scalp. One can then determine the percentage of scalp hair lost in a given quadrant and multiply this by the total scalp area delineated by that quadrant and add the resultant numbers for each quadrant. For example, if one fourth (25%) of the back area of the scalp was devoid of hair, then  $0.25 \times 24\% = 6\%$  scalp hair loss in this quadrant. In this same patient, if the top of the scalp had 40% loss, then  $0.4 \times 40\% = 16\%$  scalp hair loss in this section. If the sides of the scalp in this patient had no loss, the total scalp hair loss for this person would thus be  $6\% + 16\% = 22\%$ , or  $S_1$  by the guidelines classification.

- c. Where scalp hair loss is extensive (ie, where  $> 90\%$  of the scalp hair is lost), it is often easier to reverse this procedure and to determine the amount of hair remaining versus lost on the scalp. If one determines that 8% of scalp hair remains, obviously this is also 92% loss or  $S_4$  ( $S_{4a}$ , to be more specific) according to the guidelines classification.

- d. The investigator may also draw the alopecic areas directly on the diagrams for a general means of documenting location and extent of loss or, if the areas of loss are very discrete, to determine percentage of loss by an image analyzer. Drawing the lesions directly on the diagrams is not necessary to determine percentage of hair loss using methods *a-c*, as given above.

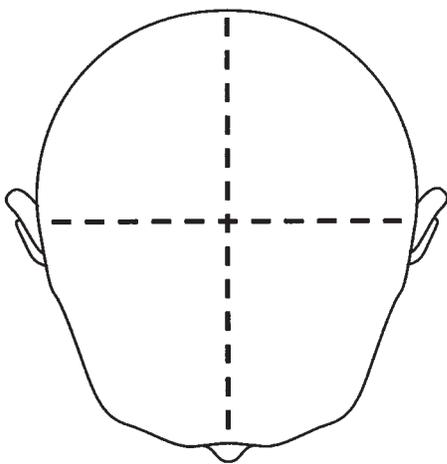
2. Vellus hair is not taken into account in this system, only terminal hair. A fine downy hair growth on the scalp that is otherwise devoid of terminal hair would be characterized as alopecia totalis (AT) or  $S_5$ .

3. We encourage investigators to examine all of the alopecia areata patient's body for hair and nail abnormalities so that body hair loss and nail

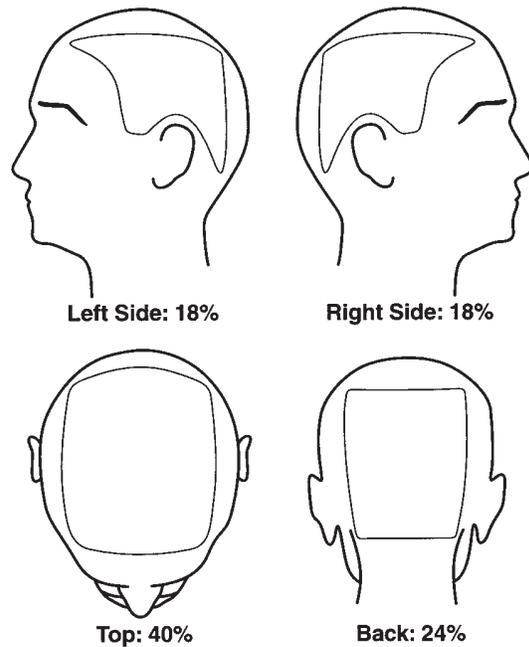
involvement can be most accurately determined and recorded. Determination of body hair loss and nail involvement can be done without examining the patient in toto; a patient who only exposes his or her arms, face, and scalp to examination can be determined to have B<sub>1</sub> and N<sub>1</sub> classification (ie, some body hair loss and some nail involvement). However, this same patient, if undressed, may more accurately have B<sub>2</sub> and N<sub>1a</sub> (total body hair loss and 20-nail dystrophy) classification. In this case, B<sub>1</sub> and N<sub>1</sub> rating, although not totally accurate, is still correct and is superior to not answering these questions at all. Please note that a patient must be checked for total body hair loss to designate that person as having alopecia universalis (S<sub>5</sub>B<sub>2</sub>).

4. For reporting purposes, the terms *alopecia totalis* (AT) and *alopecia universalis* (AU) may still be used, but they are defined narrowly (ie, AT is 100% terminal scalp hair loss without any body hair loss and AU is 100% terminal scalp hair and body hair loss). *Alopecia totalis/alopecia universalis* (AT/AU) is the term now recommended to use to define AT with variable amounts of body hair loss. A patient with extensive, but not complete, terminal scalp hair loss and extensive or complete body hair loss is not defined by AT, AU, or AT/AU.

Elise A. Olsen, MD  
Duke University Medical Center



**Fig 1.** Visual aid for estimating percentage scalp hair loss. (Described by Price VH. In: Khoury EL, Price VH, Abdel-Salam MM, Stern M, Greenspan JS. Topical minoxidil in alopecia areata: no effect on the perifollicular lymphoid infiltration. *J Invest Dermatol* 1992;99:40-7.)



% = percentage of scalp surface area in region outlined.

**Fig 2.** Visual aid for estimating percentage scalp hair loss. (Olsen EA, Canfield DC. Canfield Scientific; Fairfield, NJ.)