Fast Facts: Disorders of the Hair and Scalp

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Second edition
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List of abbreviations

ANA: antinuclear antibody
DHT: dihydrotestosterone
DLE: discoid lupus erythematosus
FPHL: female pattern hair loss
FSH: follicle-stimulating hormone
H&E: hematoxylin & eosin (staining)
Ig: immunoglobulin
LH: luteinizing hormone
LPP: lichen planopilaris
MPHL: male pattern hair loss
PCOS: polycystic ovarian syndrome
PSA: prostate specific antigen
PUVA: psoralen combined with ultraviolet A
SHBG: sex hormone-binding globulin
SLE: systemic lupus erythematosus
VDRL: Venereal Disease Research Laboratory
Introduction

From the Biblical image of Samson’s power residing in his hair, to the present day vogue of hair extensions, colors and styling, human hair has played a key role in sexual and social communication throughout history. Hair is inextricably linked with an individual’s sense of worth, personal style and self confidence. If this is overlooked by clinicians, patients presenting with hair disorders may feel undertreated and despair that the devastating effects of their hair loss are underappreciated.

This fully updated second edition of Fast Facts: Disorders of the Hair and Scalp covers basic hair biology and pathology in enough detail to help understand the mechanism of hair loss and how symptoms relate to pathogenesis. Its primary focus, however, is one of practical application, including a number of ‘top tips’ to aid clinical diagnosis, such as the value of the dermatoscope in examination of the hair and scalp.

It should be noted that randomized controlled trials are virtually non-existent in hair disease. Clinicians treating patients with hair loss often rely on evidence which is at best ‘Strength of Recommendation B’ and with quality of evidence very often level II or III. For this reason we have not routinely included ‘evidence strength data’ throughout the text.

Overall, this concise handbook will provide healthcare professionals with the ability to diagnose a range of common and less common hair and scalp disorders, including different types of hair loss, to perform further investigations where appropriate, and to provide comprehensive and appropriate therapeutic, cosmetic and supportive advice. It is an ideal resource for primary care practitioners, practice nurses, trainee dermatologists and medical students, as well as consultant dermatologists and trichologists wanting to keep up to date and informed on hair and scalp disorders.

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Hair development

The first follicles appear on the eyebrows, upper lip and chin at about 9 weeks of embryonic development. By 22 weeks the entire body, except for the palms and soles, glans penis and labia minora, is covered by 5 million follicles at regular intervals of 274 to 350 µm. The first hairs produced by these follicles are fine, soft, non-pigmented lanugo hairs with no central medulla. About 1 month before birth, the entire coat of lanugo hair is shed in utero cephalocaudally over 7 days, and is replaced by a second coat of lanugo. This second lanugo is then shed 3–4 months after birth.

At birth, some babies are born with a crop of hair on their head, but many have no discernible difference between the hair on the scalp and body. This lack of differentiation may persist for up to 12 months. Over time the differences between scalp, eyebrow and body hair accentuate. With progressive hair cycles (see ‘The hair cycle’, page 11) the scalp becomes covered with longer and coarser terminal hairs, while hair follicles on the body miniaturize and produce barely visible vellus hairs. On the scalp, primary follicles develop secondary hairs that together form follicular units. Multiple hairs emerge on the scalp to form a single follicular infundibulum and contribute to scalp hair volume (Figure 1.1) (see ‘Follicular units’, page 9).

At puberty, vellus hairs in the axilla and pubis in both sexes, and the beard and chest in men, transform into terminal hairs under the influence of androgens.

The hair follicle

An adult scalp contains on average 100 000 hair follicles.

Formation. Mesenchymal cells in the dermis aggregate just below the epidermis to form a dermal papilla. This aggregate in turn entices the epidermis to migrate down into the dermis where it expands into the hair bulb and engulfs the dermal papilla. Melanocytes migrate from
the neural crest into the hair bulb, and are responsible for hair pigmentation (Figure 1.2). Active cell division only occurs in the matrix within the bulb, and only in a single direction – upwards towards the skin surface. Cells within the matrix rapidly differentiate into the six distinctive layers of the hair: at the center, the fiber medulla, cortex and cuticle, surrounded by the inner root sheath cuticle, and Huxley’s then Henle’s layers. The medulla is absent in vellus hairs and often intermittent rather than continuous in terminal hairs (Figure 1.3).

The outermost inner root sheath layers harden and keratinize first, and provide support and structure to the undifferentiated fiber in the center. Once the fiber keratinizes and hardens, the inner root sheath becomes redundant and is dissolved and digested by sebum entering the upper follicles at the level of the sebaceous gland duct.

The fiber cuticle consists of 5–10 overlapping cell layers, each 350–450 nm thick, which appear to be imbricated (overlapping like the tiles on a roof) (Figure 1.4). The scale margins are intact over the newly formed part of the hair, but as the hair emerges from the skin the margins become jagged and progressively break off (‘weathering’).
The ‘environmental’ outer surface of each cuticular cell has a highly inert A-layer, rich in ultra-high-sulfur protein, which occupies the region just below the cell membrane and helps to protect the cells from premature breakdown due to chemical and physical insults.

**Follicular units.** In most mammals, hairs exist in the skin in follicular units, with 2–5 hairs emerging from a single pore or in very close proximity. These units have a single arrector pili muscle that attaches to all the hairs within the unit (Figure 1.5a). Compound units with multiple hairs are best seen on the adult scalp, while most units on the body and male beard are single units with a solitary hair (Figure 1.5b). These units are not fully developed at birth, and scalp hair in newborns is generally very light and fine with only a single hair emerging from each pore. By school age, secondary hairs develop alongside the primary hair, adding volume and density to the hair. In androgenetic alopecia, secondary hairs are preferentially lost from these compound follicular units, leading to increased hair shedding and a diffuse loss of hair volume that precedes the development of baldness, often by a number of years (see Chapter 4).
Figure 1.3 (a) Section through a hair follicle, showing six distinctive layers – at the center the medulla, cortex and cuticle, surrounded by the inner root sheath (IRS) cuticle and Huxley’s and Henle’s layers. (b) Section of a non-medullated hair follicle stained with toluidine blue. Reproduced courtesy of Professor Les Jones, Melbourne University.
Androgenetic alopecia is the process whereby both circulating and locally produced androgen hormones bind to and activate hair follicle androgen receptors and induce loss of scalp terminal hairs through a process of hair follicle miniaturization (Figure 3.1). The inherent sensitivity of each follicle to androgens and hence the age of onset, rate of progression, overall severity and pattern of hair loss is genetically determined. The clinical outcome of androgenetic alopecia is a bald scalp, although the pattern of hair loss is manifestly different in men (male pattern hair loss; MPHL) and women (female pattern hair loss; FPHL).

**Figure 3.1** Circulating androgens (e.g. testosterone) and locally produced androgens bind directly to cytoplasmic androgen receptors in the dermal papilla cells of the hair follicle. The androgens can also be metabolized by the enzyme 5α-reductase into dihydrotestosterone (DHT). DHT binds to the same cytoplasmic androgen receptors but five times more avidly, making it more bioactive than its precursor testosterone. Although the process is not yet fully understood, androgen activity leads to hair follicle miniaturization.
Pathophysiology

**Changes in the hair follicle.** The specific events within the follicle after androgen binding that produce miniaturization are not understood. Our understanding of these downstream events is complicated by the fact that compared with scalp follicles, beard follicles have the exact opposite response to androgen hormones. One clue is that the size of the dermal papilla determines the size of the hair bulb and subsequently the shaft. Dermal papilla cells travel out into the dermal sheath during catagen and return from the dermal sheath into the papilla when the subsequent anagen phase of the hair cycle starts (see Chapter 1). Regulation of cell traffic into and out of the dermal sheath could determine terminal-to-vellus and vellus-to-terminal transitions. In cell culture experiments, dermal papilla cells derived from beard follicles secrete insulin growth factor type 1 (IGF1) in response to androgen stimulation, while balding scalp follicles produce transforming growth factor beta (TGFβ). Whether these cytokines regulate dermal papilla cell traffic is not known.

**Changes in the hair cycle.** In addition to hair follicle miniaturization, changes in the dynamics of the hair cycle are seen in androgenetic alopecia. The anagen phase of the cycle progressively shortens, making the hair shorter in length, until it becomes so short that it is no longer visible at the follicular ostia (openings). The telogen phase remains the same or is prolonged, resulting in increased shedding and an increase in the number of empty follicles on the scalp.

**Changes in the follicular units.** Initially, miniaturization affects some but not all of the hairs within an individual follicular unit, so that on affected zones of the scalp each unit has a reduced number of hairs rather than a total absence of hairs (Figure 3.2). The sparing of one or two hairs within each follicular unit results in diffuse thinning and increased hair shedding that precedes the development of baldness (Figure 3.3). Eventually, when all the hairs within a follicular unit are lost, a bald scalp becomes apparent.

In contrast, in alopecia areata, which also demonstrates follicular miniaturization, all the hairs within each unit are lost simultaneously leading to instantaneous baldness within affected areas.
Figure 3.2 Hair follicle miniaturization. The number of hairs emerging from a follicular unit reduces as miniaturization occurs. The sparing of one or two hairs within each follicular unit results in diffuse thinning before total baldness. Note, the hairs closest to the arrector pili muscle are the most resistant to miniaturization, while the most distal hair follicles separate from the arrector pili muscle and are unable to maintain their stem cell niche.
Figure 3.3 Progressive androgenetic alopecia. As Figure 3.2 shows, there is a reduction in the number of hairs emerging from each follicular unit, resulting in diffuse thinning across the scalp. In (b) several miniature hairs are visible emerging from the follicular units (arrows), while in (c) there are fewer follicular units, and several with only single hairs remaining, resulting in sparser hair coverage.