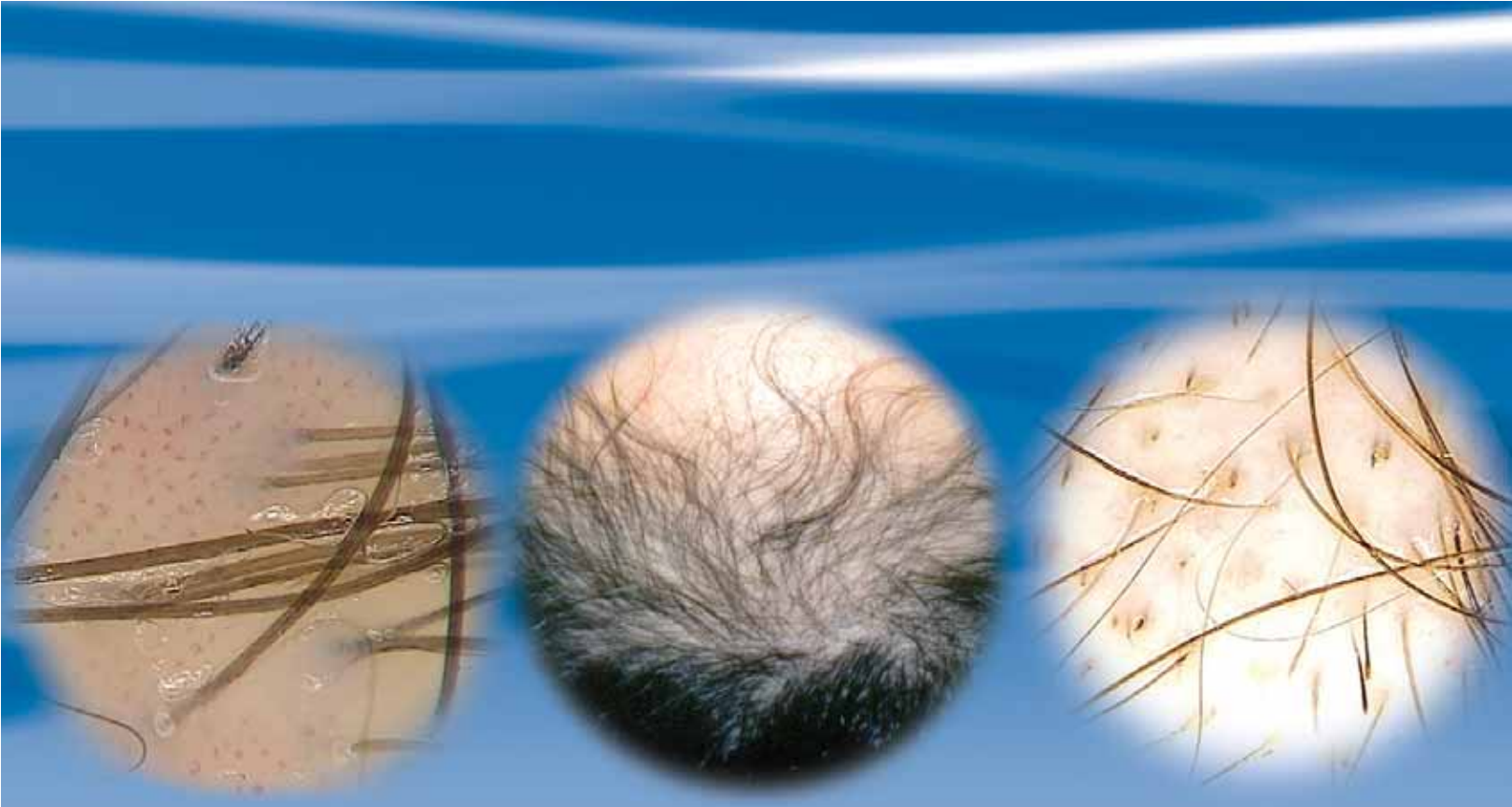

DIAGNOSIS AND TREATMENT OF HAIR DISORDERS

An Evidence Based Atlas

Antonella Tosti
Bianca Maria Piraccini

Taylor & Francis
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Diagnosis and Treatment of Hair Disorders

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Contents

Preface

Hair science is one of the most captivating topics of dermatology that goes on evolving over the years. Hair disorders are numerous and the genetic aid has permitted to classify most of the hereditary hair defects. Thanks to molecular biology, current understanding of hair follicle science is improving.

Although hair is not essential for health and survival in humans, changes in hair growth density, pattern or alterations in its color and texture can often lead to distress. In addition, these types of changes can indicate underlying systemic disease, including endocrine, genetic, metabolic and psychiatric disorders.

This book is a practical atlas, complete with color pictures and tables, to present the reader with the broad spectrum of hair and scalp disorders and to help in the differential diagnosis and treatment strategies.

Several hair disorders are very difficult to diagnose and, for this reason, it is necessary to have a practical guide helping the clinician to avoid the most common mistakes.

In this book the authors describe hair anatomy and physiology, hair and scalp disorders and methods useful to perform a correct diagnosis. One chapter is also dedicated to abnormalities induced by cosmetics and styling, another to the specific hair and scalp disorders of African-Americans, as well as to hypertrichosis and hirsutism together with their management.

Evidence levels in therapy

A: Double blind study
B: Clinical trial > 20 subjects
C: Clinical trial < 20 subjects or very large numbers of case reports
D: Series ≤ 5 subjects (5 cases in literature)
E: Anedoctical < 5 cases in literature

1 The normal hair

Introduction

There are three types of hair:

- Lanugo hairs are long and non-medullated (Figure 1.1). They are present in utero and shed after birth.
- Vellus hairs are short and non-pigmented (Figure 1.2). They constitute 6–25% of the hair population even in the areas considered to have only terminal hair.
- Terminal hairs are long, pigmented and medullated (Figure 1.3). The terminal hair shaft is wider than the inner root sheath of the follicle.

Hair density considerably varies in the different areas of the body with 1/5 (1 000 000) of the hair follicles (5 000 000) being on the head (Table 1.1). The scalp by itself contains about 100 000 follicles.

Maximum length and growth rate of terminal hair varies in the different body regions (Table 1.2). Scalp hair grow about 0.4 mm/day and may reach a length of more than 1 meter. Maximum hair length decreases with age.

Thickness of scalp hair ranges from 50–90 μm.

Table 1.1 Follicular density in the different body regions

Scalp		200–400/cm²*
Face	Nasal fold	1600/cm²
	Anterior forehead	800/cm²
	Lateral forehead	455/cm²
	Preauricolar area	466/cm²
Trunk		50–100/cm²
Lower limbs		50/cm²

*Hair density decreases with aging.

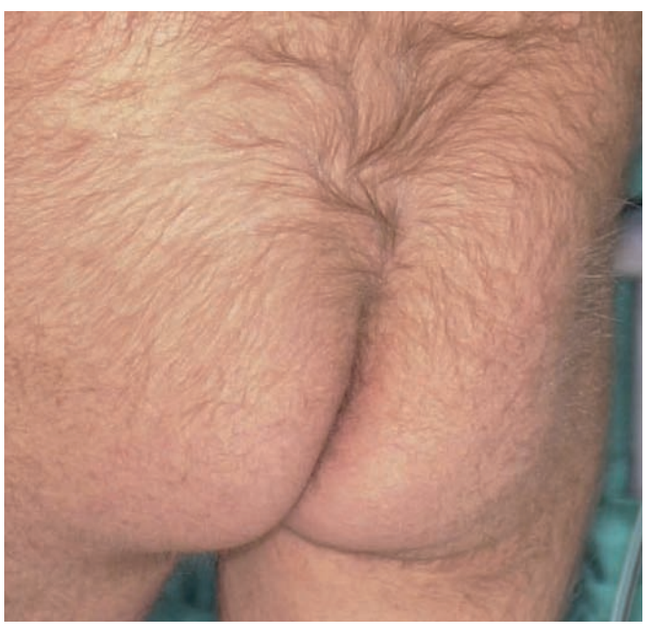

Figure 1.1
Lanugo hair in a newborn.

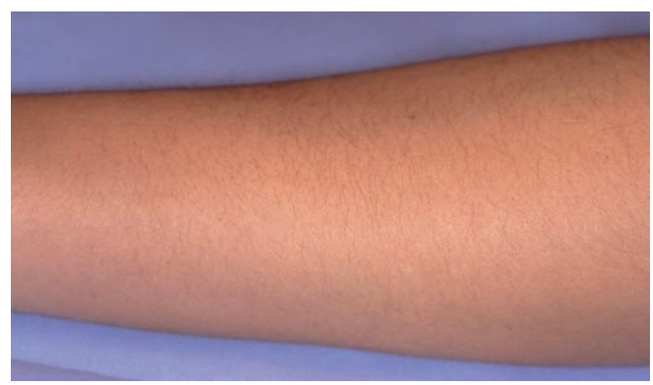

Figure 1.2
Vellus hair.

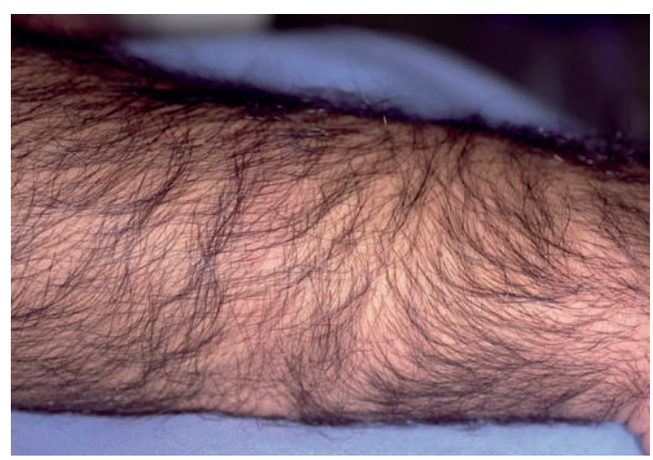

Figure 1.3
Terminal hair.

Table 1.2 Hair growth rate

Scalp	0.35 mm/day
Chin	0.38 mm/day
Brows, eyelashes	0.16 mm/day
Axillae	0.30 mm/day
Thighs	0.20 mm/day
Legs	0.20 mm/day

Table 1.3 Vellus to terminal conversion according to age

Pubis	Adrenarche (7–9 years)
Axillae	Adrenarche (7–9 years)
Face	12–14 years

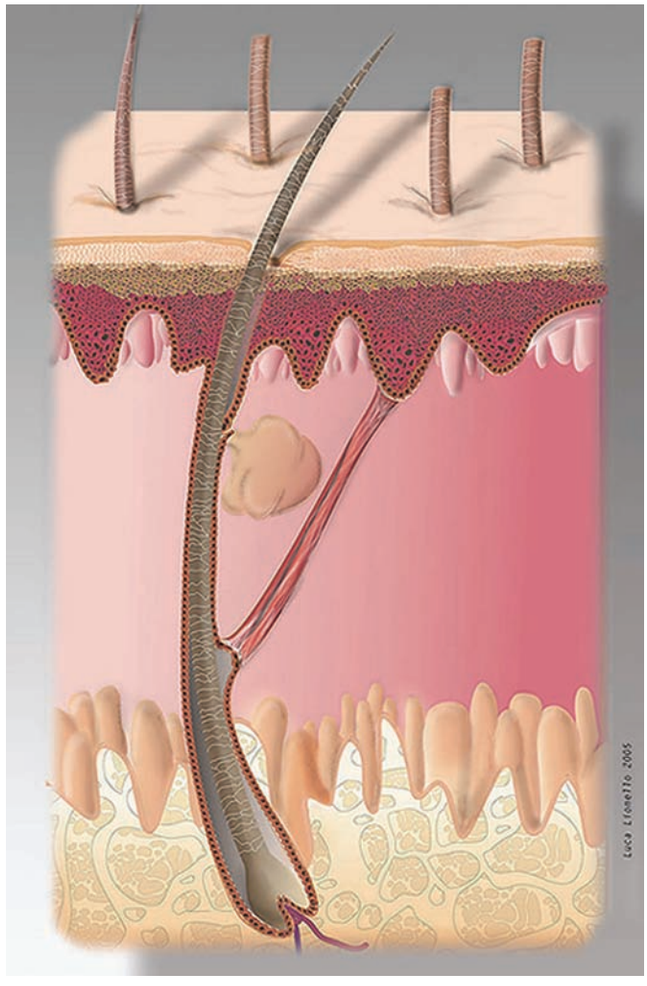

Figure 1.4
Hair follicle.

At puberty, vellus hair of androgen-dependent areas transforms into terminal hair under the effect of androgens (Table 1.3).

The hair follicle

The hair follicle (Figure 1.4) is divided in two portions:

- The upper follicle, permanent
- The lower follicle, dynamic.

The hair bulb, which is the deep bulbous portion of the hair follicle that surround the dermal papilla, contains the hair matrix which produces the hair shaft and its sheets (Figure 1.5).

The hair cycle

Hair follicles have a cyclic activity characterized by alternance of periods of hair shaft production and of resting (Figure 1.6). Adjacent follicles are not synchonized. The hair cycle consists of three phases:

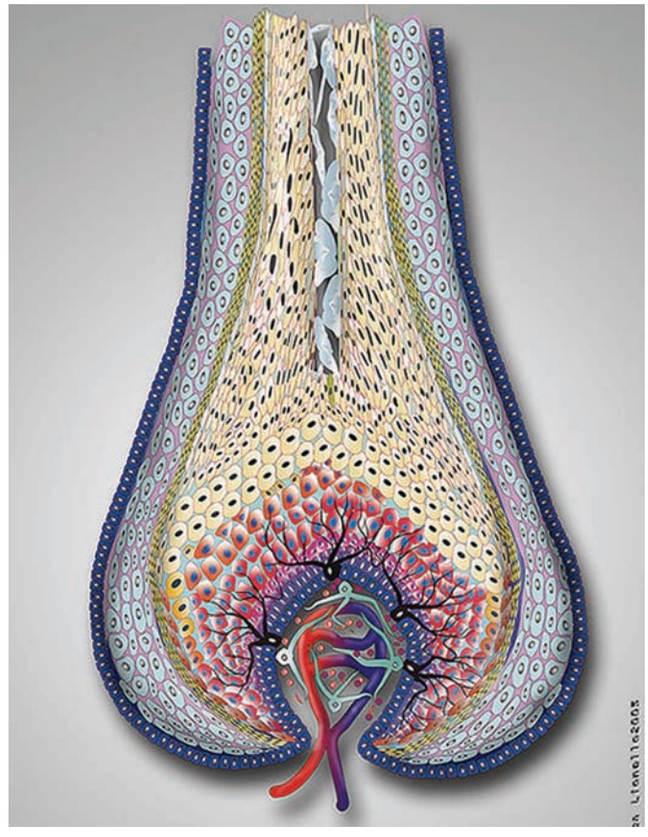

Figure 1.5
Hair matrix.

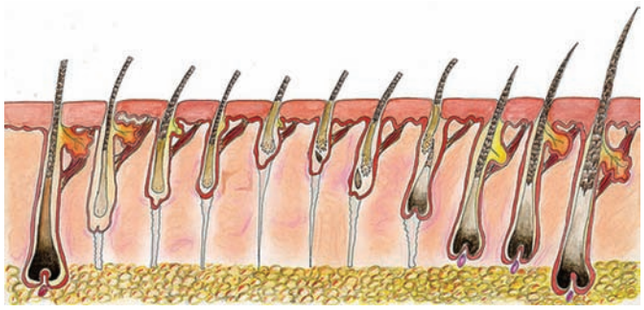

Figure 1.6
Hair cycle.

1. Anagen (hair shaft production). The hair follicle actively produces the hair shaft (Table 1.4). This phase is the longest of the hair cycle and its duration determines the length of the hair shaft. Due to the high mytotic rate of follicular matrix cells, the anagen phase is very sensitive to noxious insults.

Table 1.4 Duration of anagen in different body areas

Scalp	2–6 years
Chin	1 year
Brows, eyelashes	1–2 months
Axillae	4 months
Arms	3 months
Pubis	4 months
Thighs	1–2 months
Legs	4–6 months

2. Catagen. This is a transitional phase which lasts 2–3 weeks and is characterized by apoptosis of the hair matrix cells and involution of the lower part of the follicle. During catagen, the hair bulb migrates from the hypodermis to the mid dermis.
3. Telogen (resting phase). During this phase hair shaft production is absent and the hair bulb is completely keratinized. Telogen phase, in the scalp follicles, lasts about 3 months but it is considerably longer in other body regions such as the lower limbs. The hair shaft remains anchored to the follicle during telogen. Hair shedding (teloptosis/exogen) usually occurs when the follicle re-enters a new anagen phase (Figure 1.7). In some follicles teloptosis occurs before the end of telogen and the follicle remains empty for a certain period (kenogen) (Figure 1.8). This may be due to a premature teloptosis or prolonged telogen duration.

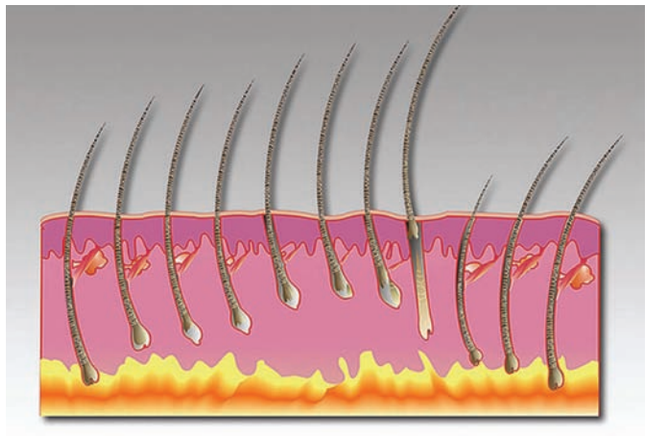

Figure 1.7
Teloptosis.

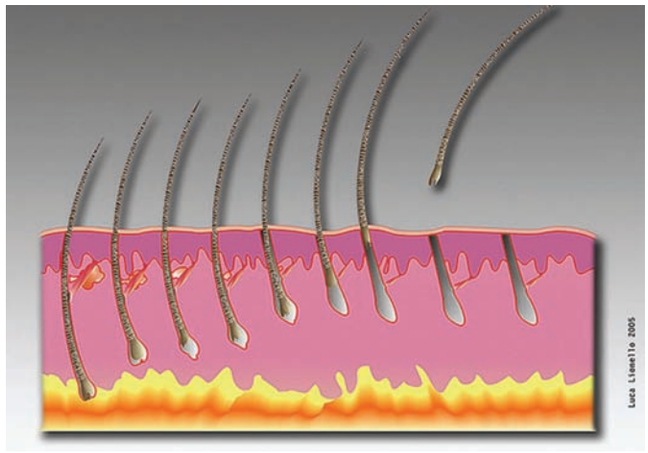

Figure 1.8
Sometimes after telogen the hair follicle remains empty (kenogen).

Factors that regulate the hair cycle are still unknown, but several cytokines and growth factors are possibly involved (Table 1.5).

Follicular stem cells are localized in the outer root sheath at the isthmus level within the follicle bulge (Figure 1.9). These cells repopulate the hair matrix at each cycle. Destruction of follicular stem cells results in cicatricial alopecia.

The hair shaft

The hair shaft (Figure 1.10) is made up of three structures.

Table 1.5 Factors regulating hair growth

Family	*Effect on hair cycle*
IGF-I	Essential for follicular growth in vitro
VEGF	Essential for anagen-associated angiogenesis
FGF7	Induces anagen
SHH	Initiates anagen
Estrogen receptor antagonist	Induces anagen
PTH rp	Induces anagen
Calcineurin	Induces anagen Blocks onset of catagen
Prolactin	Stimulates anagen and catagen onset
HGF receptor	Retards entry to catagen
FGF5	Terminates anagen
TGF-β-RI	Promotes catagen
TGF-β1	Promotes catagen
Neurotrophin 3, neurotrophin 4, BDNF	Promote catagen

BDNF = brain-derived neutrophic factor; FGF = fibroblast growth factor; HGF = hepatocyte growth factor; IGF-I = insulin-like growth factor; PTHrp = parathyroid hormone-related peptide; SHH = sonic hedgehog; TGF = transforming growth factor; VEGF = vascular endothelial growth factor.

- *The cuticle:* this is the outer part of the hair shaft and it is composed of 5–8 layers of flattened cells with an overlapping pattern and with the free edge oriented upward.
- *The cortex:* this is composed of flattened and tight cells, known as hair fibers, that are elongated and contain melanin pigment.
- *The medulla:* this is present only in terminal hairs and it is made up of polyedric cells that are scarcely cornified and rich in glycogen and air bubbles.

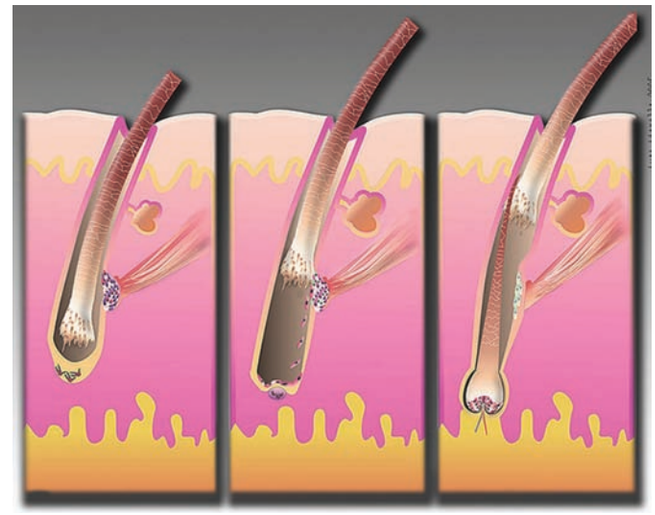

Figure 1.9
The bulge area contains hair follicle stem cells.

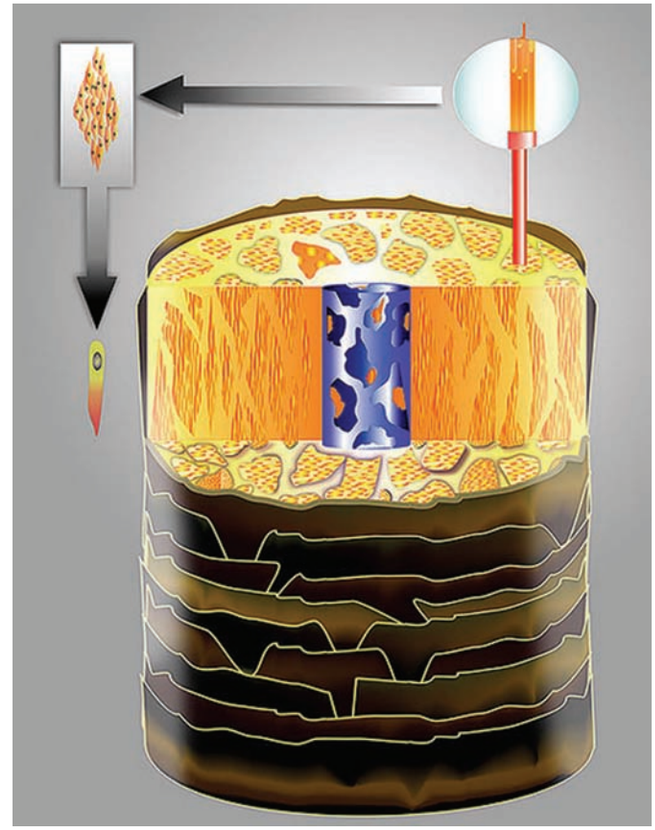

Figure 1.10
Different constituents of the hair shaft.

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2 Evaluation of hair loss

Examine topographic pattern of hair loss

Normal hair density

- Look at the temporal region: temporal thinning with short hair suggests chronic telogen effluvium (Figure 2.1).

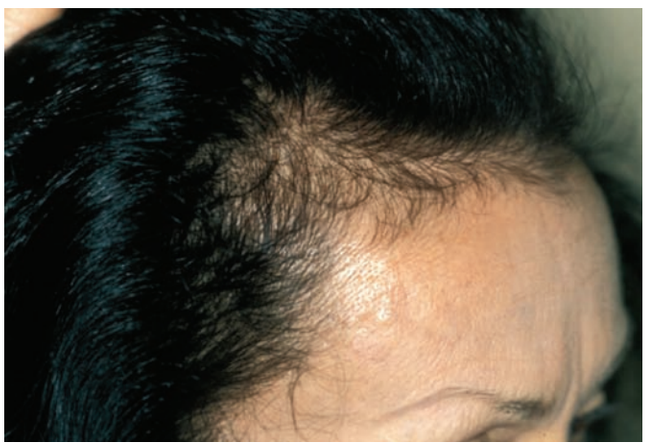


Figure 2.1
Temporal thinning in chronic telogen effluvium.

- Perform a pull test: if more than two roots (Figures 2.2, 2.3) for pull site
  - acute telogen effluvium (telogen roots in different stages of maturation) (Figures 2.4–2.6).
  - anagen effluvium (telogen and dystrophic roots (Figure 2.7). Pol-Pinkus mark can be evident (Figure 2.8)).
  - acute alopecia areata (dystrophic roots (Figure 2.9) and telogen roots or telogen and catagen roots (Figure 2.10)).

Figure 2.2
Pull test.

Figure 2.3
Positive pull test.

Figure 2.4a
Telogen hair roots at pull test.

Figure 2.4b
Mature telogen root.

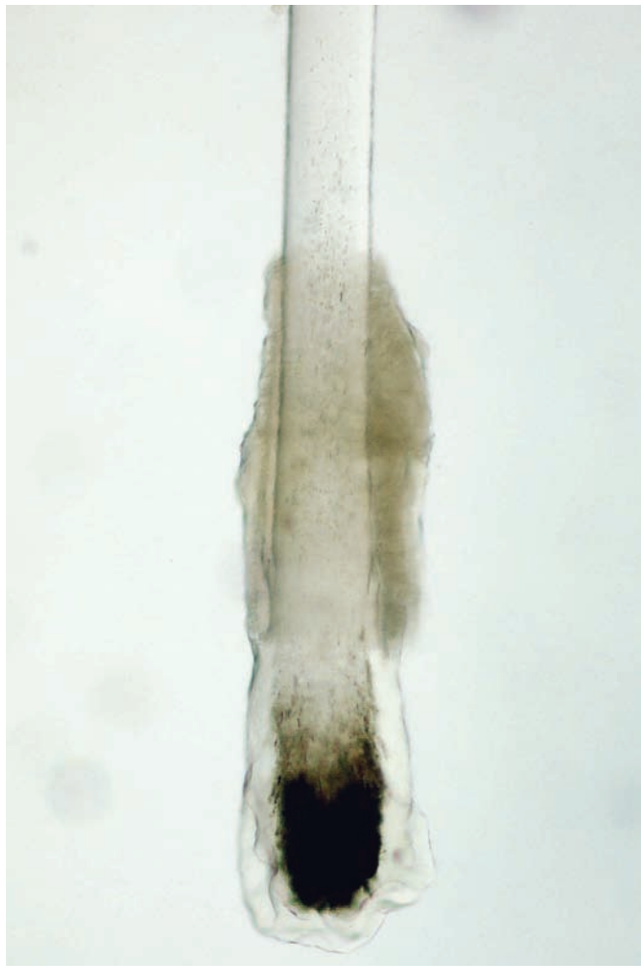


Figure 2.5
Early telogen root in acute telogen effluvium: note presence of pigment in the club.

Figure 2.6
Keratinized telogen hair surrounded by hair cast in acute telogen effluvium.

Figure 2.7a,b
Dystrophic hair roots in anagen effluvium.

Figure 2.8a,b
Pol-Pinkus mark.

Figure 2.9a,b
Dystrophic hair roots in alopecia areata.

Figure 2.10
Telogen and catagen hair roots at the pull test in alopecia areata incognita.

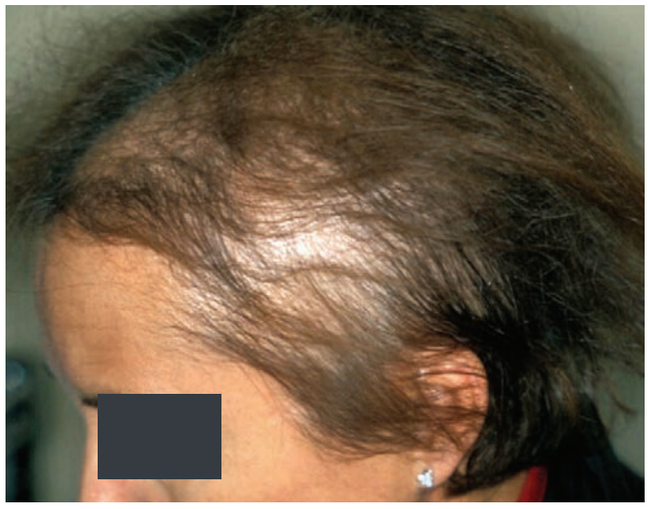


Figure 2.11
Diffuse hair loss due to AGA in woman.

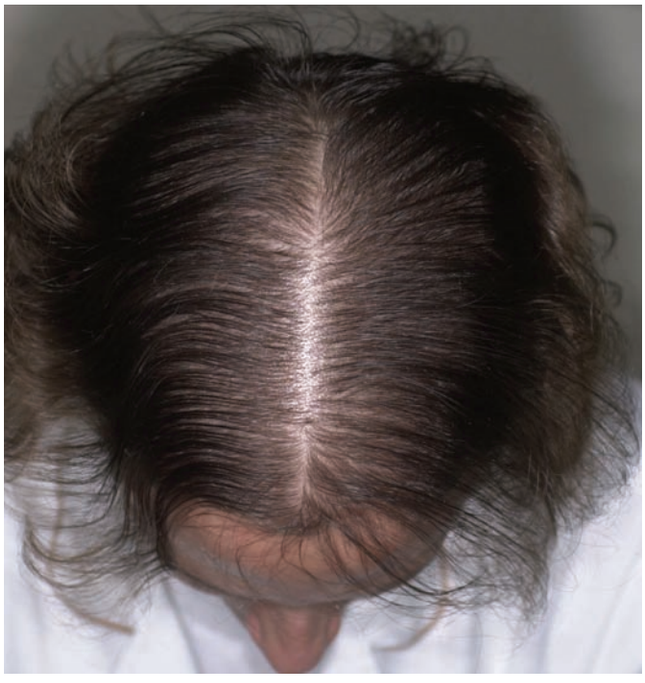


Figure 2.12
Acute telogen effluvium.

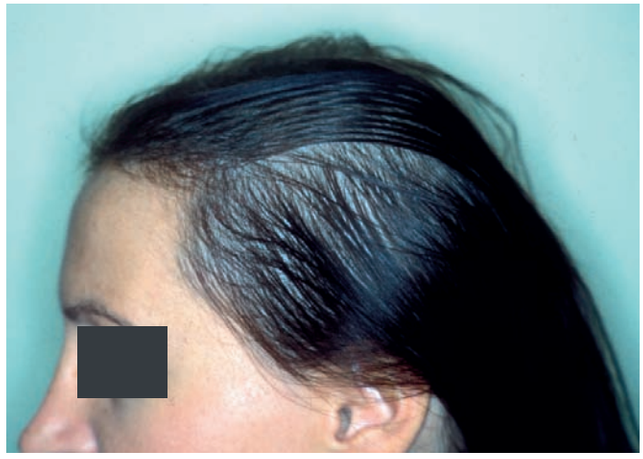


Figure 2.13
Alopecia areata incognita.

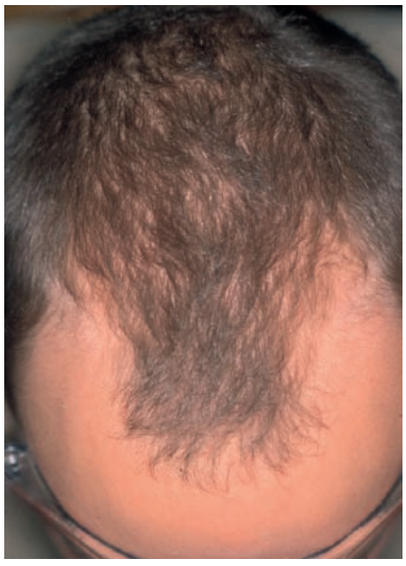
Non-patchy hair thinning

Diffuse hair thinning

- Normal pull test (< than two roots for pull site) androgenetic alopecia: in women hair thinning often affects parietal and occipital scalp (Figure 2.11) even though thinning is more marked on the top.
- Abnormal pull test (> than two roots for pull site):
  – telogen roots (acute telogen effluvium) (Figure 2.12)
  – telogen roots (alopecia areata incognita) (Figure 2.13)
  – telogen and dystrophic roots (anagen effluvium)

Patterned hair thinning (androgenetic alopecia) (Figure 2.14)

Always make a central part and compare hair density at the top with hair density in the occipital area (Figures 2.15, 2.16).



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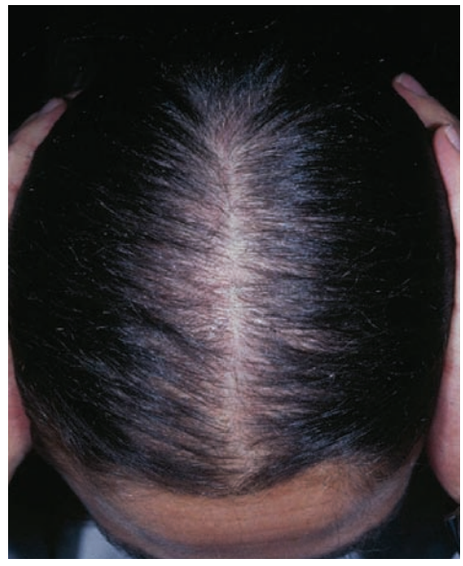
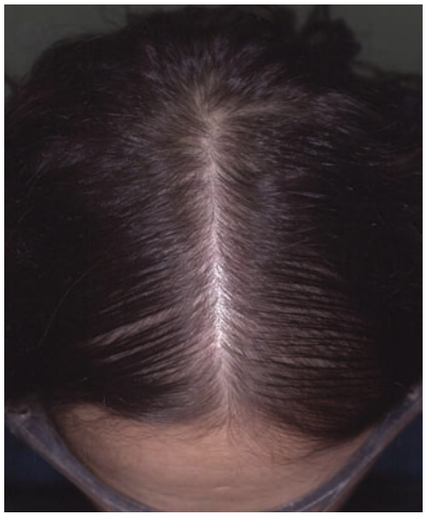


b

Figure 2.14
Patterned hair loss due to AGA in a man (a) and a woman (b).



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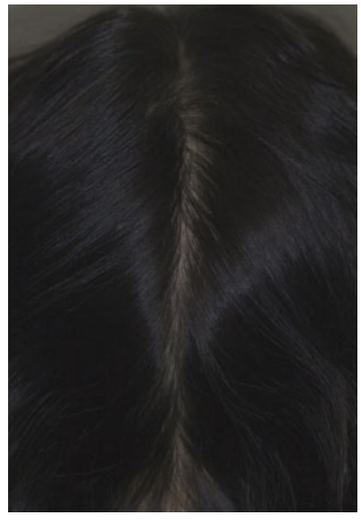


2.16

Figures 2.15, 2.16
Comparison of hair density (anterior vs posterior) by central partline.

Figure 2.17
Patchy alopecia due to alopecia areata.

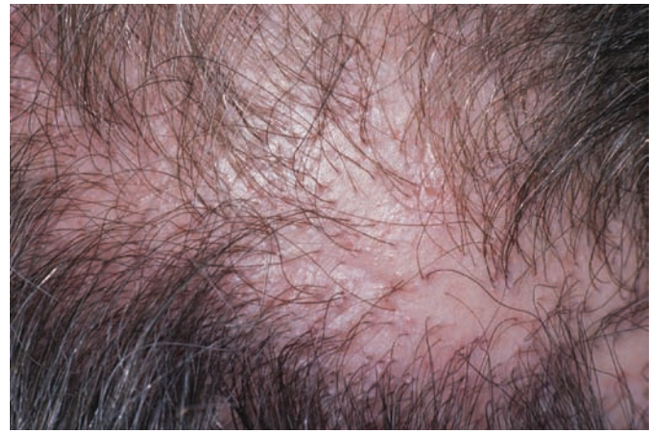


Figure 2.18
Patchy alopecia and erythema in DLE.

Figure 2.19
Anagen roots with epithelial sheaths at the pull test in cicatricial alopecia.

Patchy hair thinning

1. Normal scalp devoid of terminal hairs with/without vellus hair (alopecia areata) (Figure 2.17)
2. Atrophic scalp/presence of erythema and hyperkeratosis surrounding the alopecic area (Figure 2.18) (lichen plano-pilaris, discoid lupus erythematosus). Pull test may reveal anagen roots with thickened sheaths (Figure 2.19).
3. Atrophic scalp/presence of inflammatory papules and pustules/presence of tufted hairs (Figure 2.20) (folliculitis decalvans). Pull test may reveal anagen roots with thickened sheaths.
4. Normal scalp with short terminal hairs of different length:
   – negative pull test/no telogen hairs at the trichogram (trichotillomania)
   – beaded hair shafts at the microscope (monilethrix) (Figure 2.21).

How to perform a pull test

Grasp a tuft of 100–200 hairs 2 cm from the scalp emergency (Figure 2.22) and gently pull with the thumb and index fingers along the hair shafts toward the distal tip.

- Diffuse hair loss: the pull should be done in at least five scalp areas.

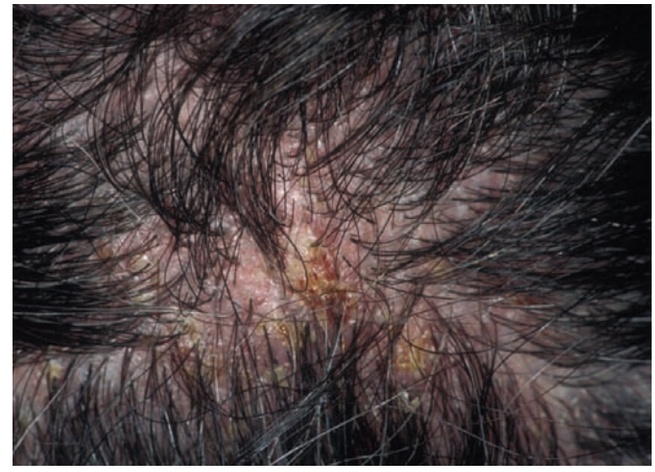


Figure 2.20
Patchy alopecia due to folliculitis decalvans.

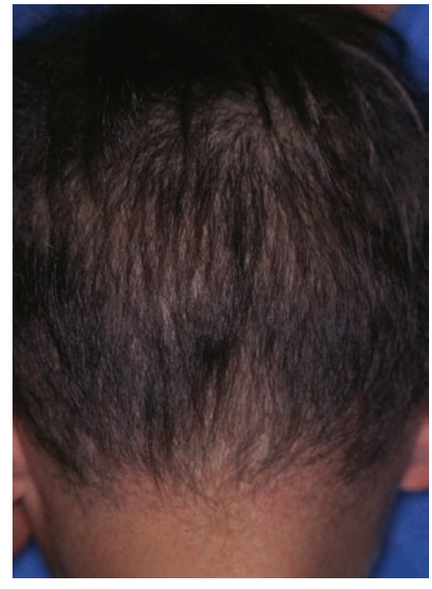


Figure 2.21
Patchy alopecia due to trichotillomania.

Figure 2.22
Pull test technique.

Figure 2.23
Telogen roots at the microscope.

- Patterned hair loss: the pull should be done both in the androgen- and non-androgen-dependent scalp.
- Patchy hair loss: the pull should be done at the patch margins and in the apparently unaffected scalp.

The number of hair that is extracted with the pull is reduced by shampooing, brushing and combing. We prefer to perform the pull test the same day or 1 day after shampooing.

- Positive pull test: > 10 hairs
- Negative pull test: < 5 hairs

Examination of the pulled hair at the microscope

Telogen root (Figure 2.23)

1. Telogen roots without epithelial sac: club-shaped, completely keratinized, represent telogen hair at the end of the telogen phase (Figures 2.4b, 2.6).

Figure 2.24
Anagen roots at the microscope.

Figure 2.25
Anagen roots devoid of sheaths typical of LAH.

2. Telogen roots with epithelial sac, with or without remnant of pigment, represent telogen hair shed before the end of telogen phase (Figure 2.5).

Both these roots are seen in all types of hair loss. The presence of short-tipped telogen hairs is typical for androgenetic alopecia.

Anagen root

1. Anagen roots with thickened sheaths. They are typically seen in cicatricial alopecia (Figure 2.24).
2. Anagen roots devoid of sheaths (loose anagen hair, LAH). They are typical of the loose anagen hair syndrome (Figure 2.25).

Figure 2.26
Dystrophic anagen hair roots.

Figure 2.27
Trichogram in a LAH.

3. Dystrophic anagen hair root. These are actually hairs that are broken at the level of the keratogenous zone and present a fractured proximal end. The hairs correspond to the distal part of the exclamation mark, hairs and are diagnostic for alopecia areata. They are also typical for anagen effluvium (Figure 2.26).

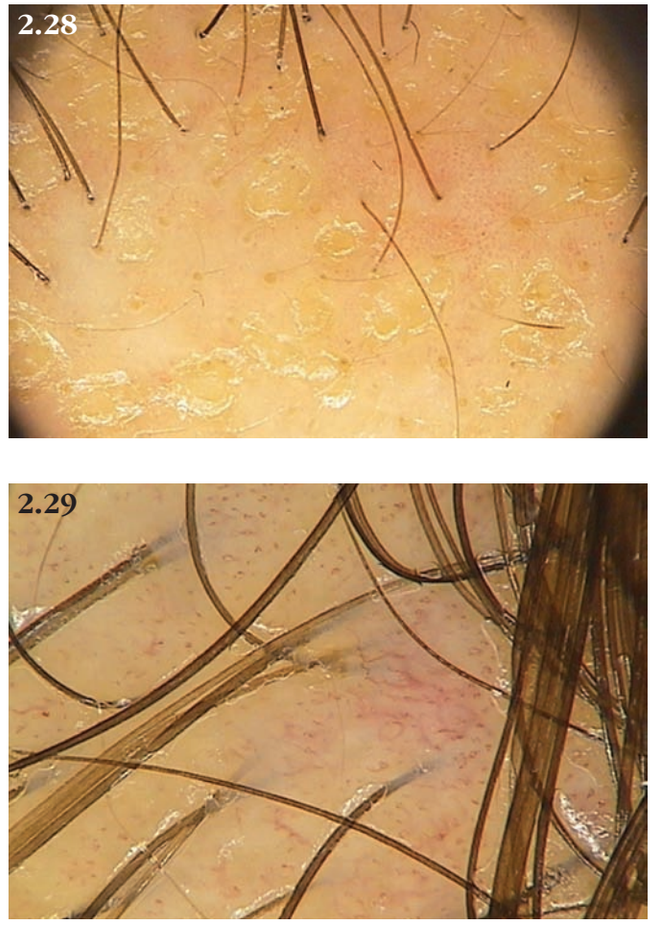
Trichogram

Trichogram is important if you suspect a loose anagen hair syndrome. A prevalence of anagen hair devoid of sheath is diagnostic for this syndrome (Figure 2.27). In all the other hair disorders the trichogram can only give information on anagen to telogen ratio, but is not diagnostic.

Videodermoscopy

Videodermoscopy is useful for close examination of the scalp of the follicular ostia and the hair shafts. Magnification ranges from 20× to 80×.

- Scalp scaling and dandruff: dermoscopy permits to distinguish psoriasis and sebopsoriasis from seborrheic dermatitis. In psoriasis and sebopsoriasis dermoscopy shows tightly coiled capillary loops (Figures 2.28, 2.29).
- Patchy alopecia: alopecia areata can be distinguished from other causes of patchy alopecia though



Figures 2.28, 2.29
Dermoscopy of the scalp in psoriasis: tightly coiled capillary loops.

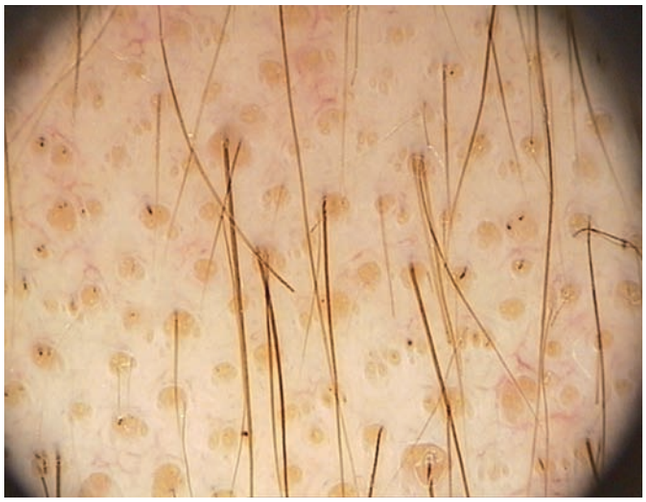


Figure 2.30
Monomorphous yellow dots in alopecia areata.

Figure 2.31
Hair diameter variability and peripilar signs in AGA.

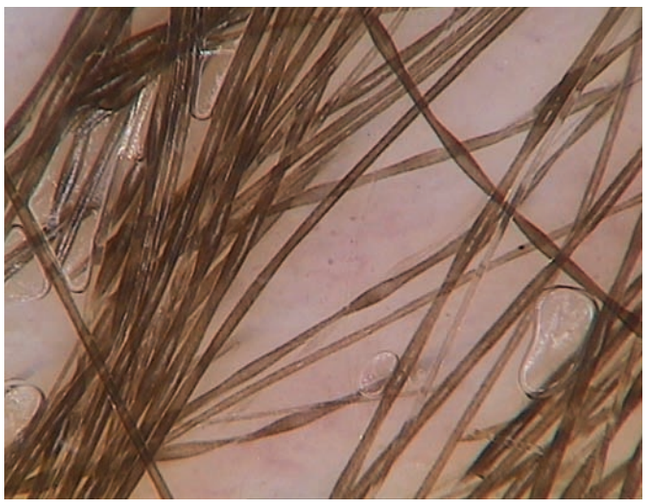


Figure 2.32
Monilethrix observed *in vivo* by dermoscopy.

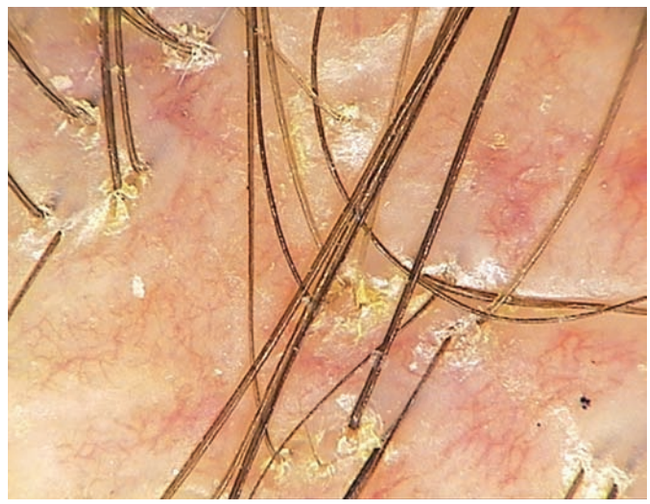


Figure 2.33
Dermoscopy in LPP.

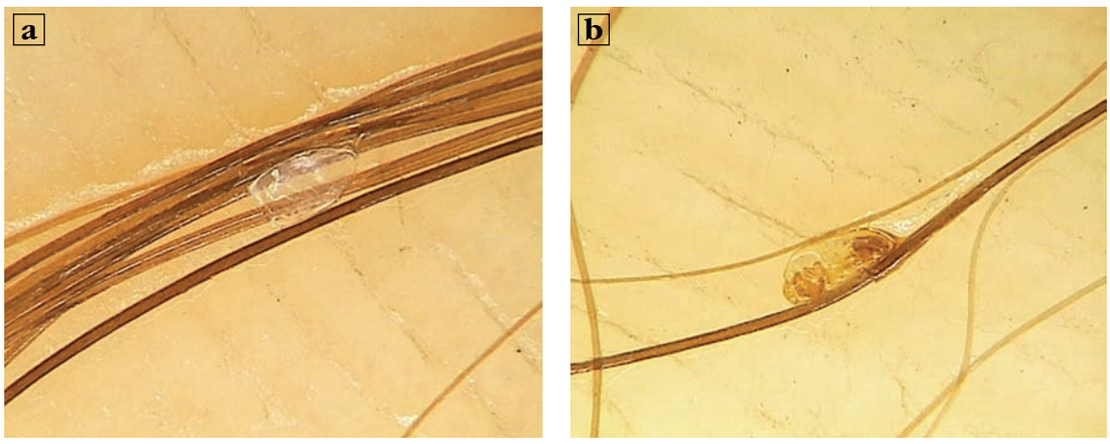
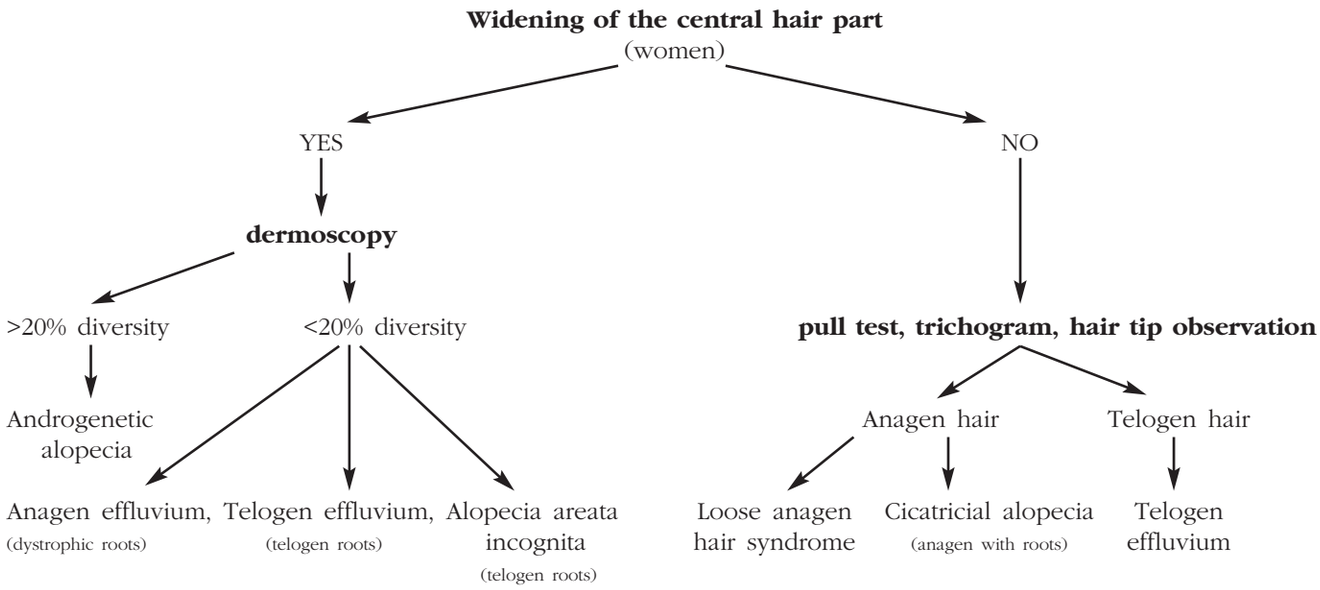


Figure 2.34
Nits at dermoscopy: (a) empty nit and (b) viable nit.

dermoscopy, which shows numerous monomorphous yellow dots (Figure 2.30). These dots disappear with hair regrowth.

- Androgenetic alopecia: more than 20% variability in the hair shaft diameter is typical of androgenetic alopecia and diagnostic in the early phases. Peripilar signs, which appear as brown halos around the follicular ostia are a sign of perifollicular inflammation which is often associated with androgenetic alopecia (Figure 2.31).
- Inherited and acquired hair shaft disorders: the hair shaft abnormalities are easily recognized in vivo at high magnification (Figure 2.32).
- Scarring alopecia: in lichen plano-pilaris (Figure 2.33) and discoid lupus erythematosus, dermoscopy shows scalp atrophy due to loss of follicular ostia and keratotic plugs around the remaining hairs. In folliculitis decalvans tufted folliculitis is often evident.
- Nits: dermoscopy permits to distinguish empty (Figure 2.34a) from viable nits (Figure 2.34b).

Evaluation of hair loss algorithm



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3 Disorders of hair pigmentation

Hair graying

Hair graying is a consequence of a reduced activity of hair bulb melanocytes together with a defective transfer of melanin from melanocytes to keratinocytes. In white hair melanogenic melanocytes are completely absent. Gray hair results from the admixture of pigmented and white hairs (Figure 3.1) as well as from gradual loss of pigment along the individual hair shaft (Figure 3.2). Hair graying is significantly correlated with chronological aging and is considered one of the most obvious manifestations of the aging process.

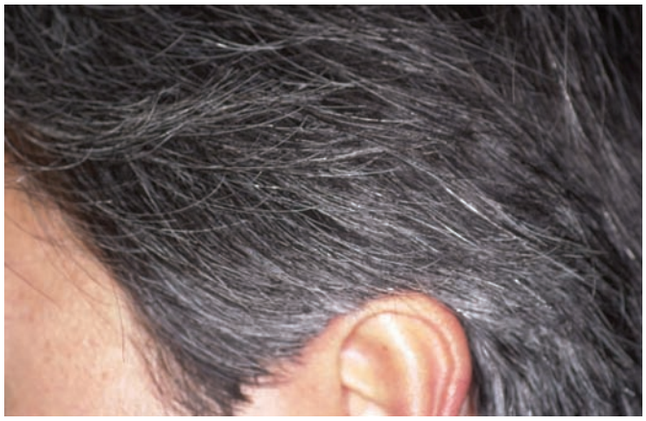


Figure 3.1
Hair graying resulting from mixture of white and pigmented hair.

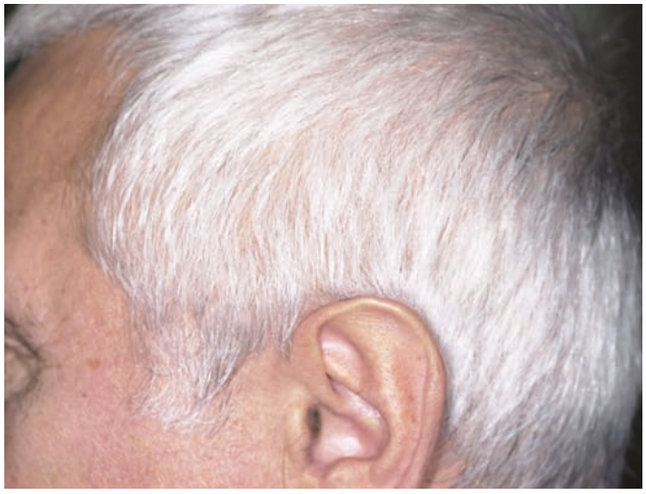


Figure 3.2
Diffuse hair graying.

Age of onset of hair graying:
- Caucasians mid 30s
- Asians late 30s
- Africans mid 40s.

Body areas listed according with onset of hair graying:
- Temples
- Vertex
- Remaining scalp
- Beard
- Body hair.

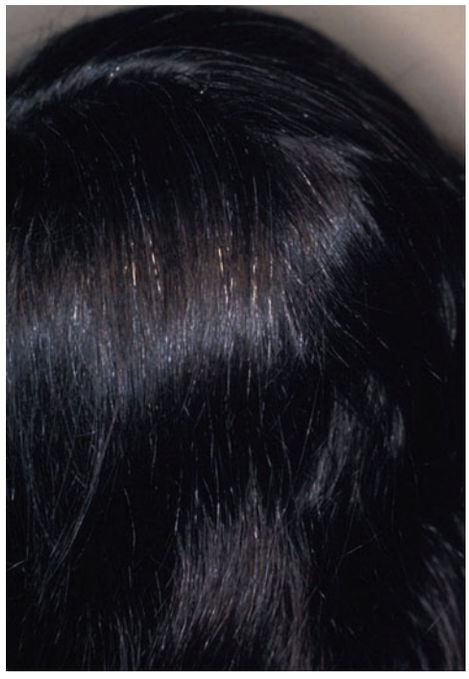
Percentage of gray hair by age
- 30 yrs = 30%
- 50 yrs = 50%.

Premature canities

Significant graying before the age of 20 in whites and before the age of 30 in blacks is termed premature canities (Figures 3.3, 3.4). Acute graying is usually a symptom of alopecia areata (see page 39). Causes of premature canities are reported in Table 3.1.

Table 3.1 Causes of premature canities

Alopecia areata (Figures 3.16, 3.17)
Ataxia teleangectasia
Autoimmune diseases
Böök syndrome
Cardiovascular diseases
Celiac disorder
Cigarette smoking
Cri-du-chat syndrome (chromosome 5p)
Drugs
Dyskeratosis congenita
Osteopenia/osteoporosis
Progeria/Werner syndrome
Rothmund–Thomson syndrome
Vitiligo
Vogt–Koyanagi–Harada syndrome
Waardenburg syndrome



3.3

3.4

Figures 3.3, 3.4
Premature canities in an 11-year-old girl.

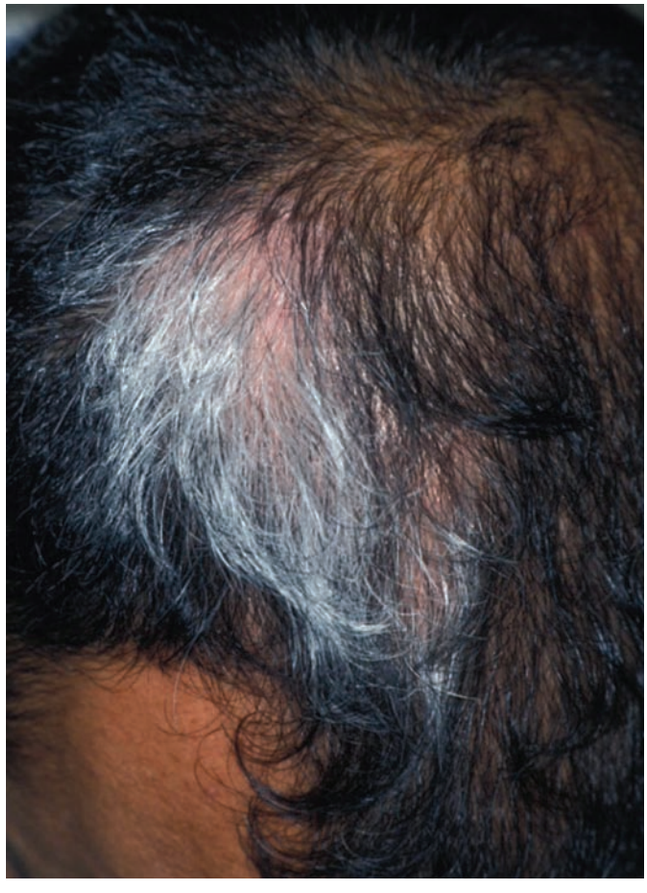


Figure 3.5
Scalp vitiligo: note scalp depigmentation.

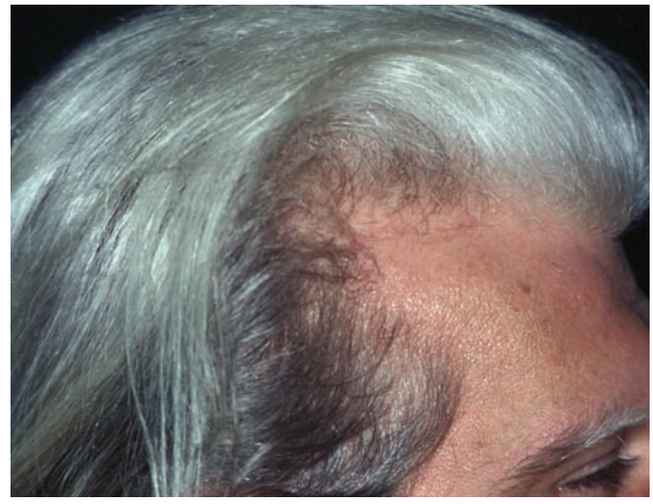


Figure 3.6
Piebaldism: triangular white forelock associated with white medial third of the eyebrows.

Poliosis

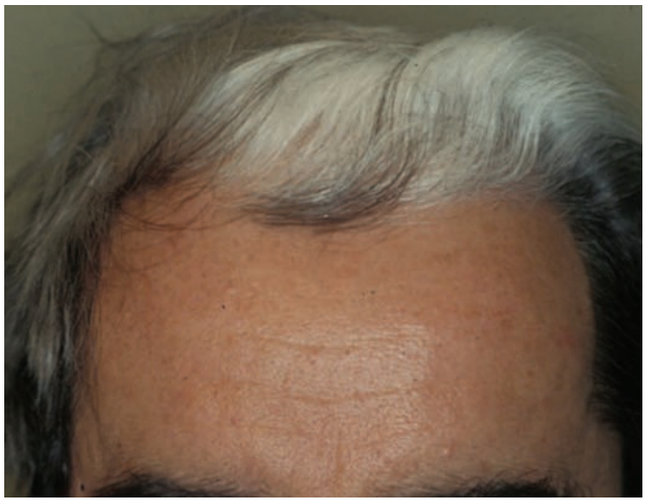
Poliosis describes the presence of a tuft or a localized patch of white hairs in the scalp or eyebrows or eyelashes. Poliosis is seen in vitiligo (Figure 3.5), piebaldism (Figures 3.6–3.8), and Waardenburg syndrome (Figure 3.9, Table 3.2).

Albinism

The albinisms are genetic disorders characterized by decrease or absence of melanin in the skin, hair and eyes in the presence of normal number, structure and distribution of melanocytes. Hair color varies from white to light brown, depending on the genetic defect (Table 3.3).

Piebaldism

Piebaldism is an autosomal dominant disorder due to mutations in the human KIT gene characterized by congenital poliosis and skin hypopigmentation. The distribution of the hair and skin hypopigmentation is typical. A triangular white forelock, which may extend



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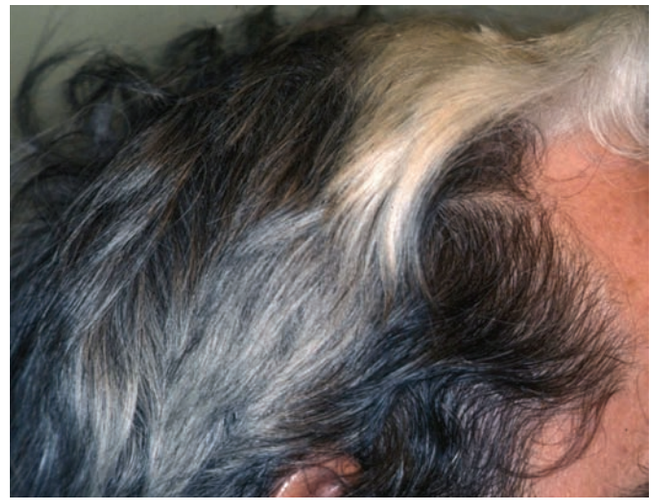

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Figures 3.7, 3.8
Piebaldism. White forelock.

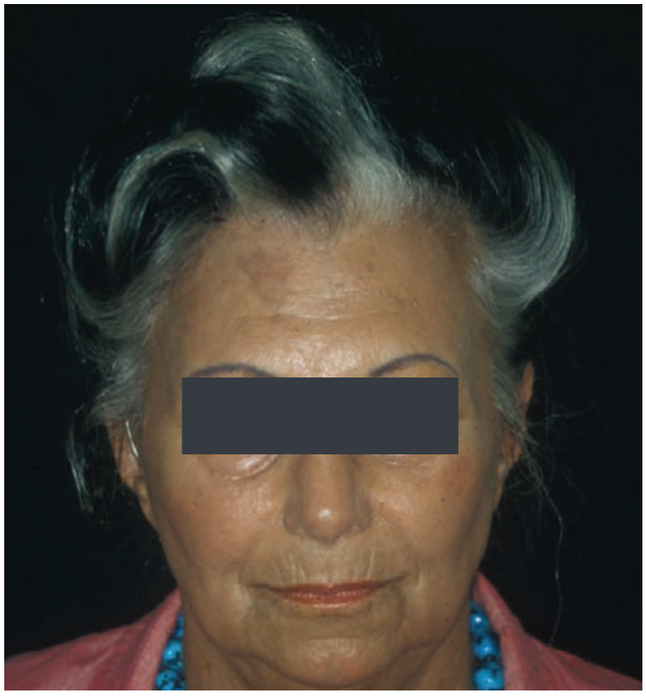


Figure 3.9
Waardenburg syndrome.

Table 3.2 Poliosis

Alopecia areata (Figure 3.18)
Herpes zoster
LED/LES
Tuberous sclerosis
Vitiligo
Vogt–Koyanagi–Harada syndrome
Waardenburg syndrome

Table 3.3 Hair color in albinisms

Oculocutaneous albinism 1A	White
Oculocutaneous albinism 1B	Pale yellow
Oculocutaneous albinism 2	Lighter than non-affected relatives
Oculocutaneous albinism 3	Reddish
Chediak–Higashi syndrome (MIM 214500)	Silvery
Hermansky–Pudlak syndrome (MIM 203300)	White to brown

to the medial third of the eyebrows is present in 90% of patients (Figures 3.6–3.8). The skin hypopigmentation is strikingly symmetrical and involves the anterior trunk and limbs, but not hands and feet.

Waardenburg syndrome

Waardenburg syndrome is an autosomal dominant disorder characterized by poliosis, deafness, dystopia canthorum and heterochromia iridis. The white forelock is often associated with premature canities (Figure 3.9).

Hair lightening

Patients with phenylketonuria and Menkes' disease have a hair color that is characteristically lighter than that of not affected relatives. Acquired hair lightening may be caused by drugs (see page 68), nutritional disorders and hair weathering (Table 3.4).

Table 3.4 Causes of hair lightening

Drugs	
Fatty acid deficiency	
Hemodialysis	
Histidinemia	
Homocystinuria	
Kwashiorkor	
Malabsorption	
Menkes' syndrome	
Phenylketonuria	
Weathering	Sun exposure
	Hypochlorous acid in swimming pool water

Hair darkening

Anecdotal reports indicate that outer root sheath melanocytes of white hair may be induced to migrate to the bulb area and then be activated to produce melanin resulting in repigmentation of white hair. The most common causes of hair darkening are alopecia areata, drugs and inflammatory diseases (Figure 3.10, Table 3.5).

Heterochromia

A certain degree of heterochromia in the hair color is common in fair-haired individuals and the color of beard and moustaches is usually lighter than that of the scalp hair. The presence of scalp hair heterochromia with spiral or horizontal band pattern along Blaschko lines can be a sign of scalp pigmentary mosaicism (Figures 3.11–3.14).

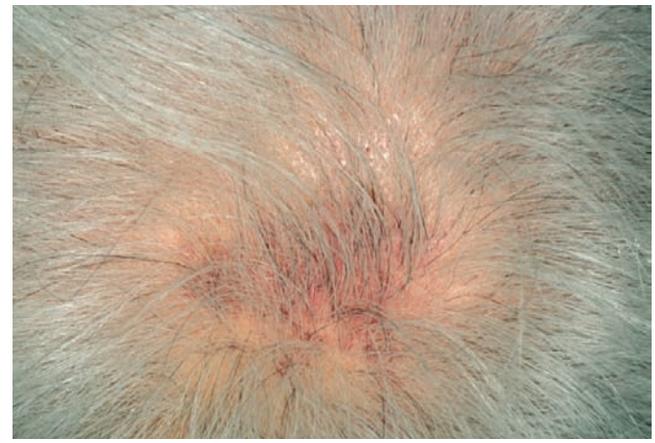


Figure 3.10
Localized hair darkening after acute pustular inflammation.

Table 3.5 Causes of hair darkening

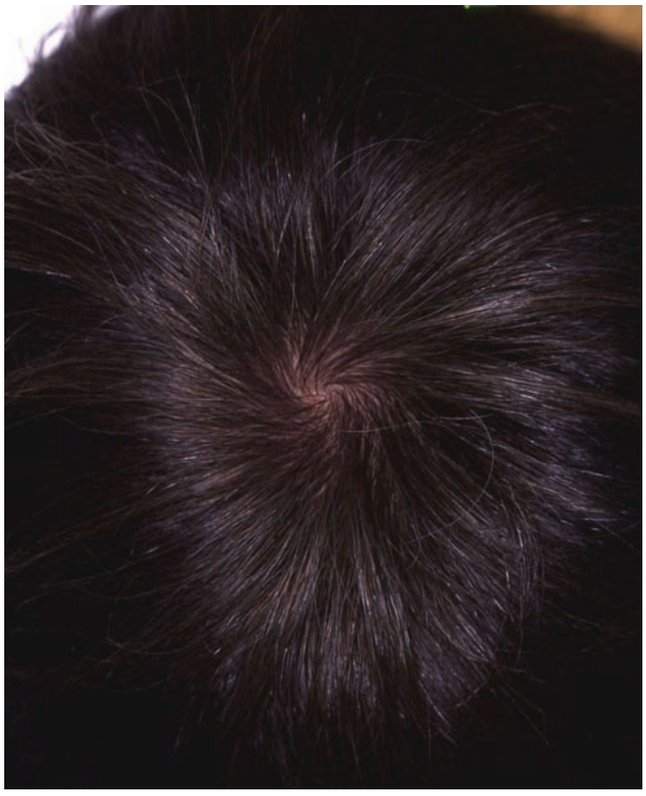
Addison's disease	Inflammatory diseases
Alopecia areata	Porphyria
Drugs (see page 68)	

Hair discoloration

Abnormal hair color can develop in severely damaged hair that can be penetrated by exogenous substances (Table 3.6). Metals are the most common causes of hair discoloration. Green hair is an unusual condition due to deposition of copper from exogenous source (Figure 3.15). The condition is seen in subjects with natural or bleached blond hairs or in subjects with white or gray hair.

Management of acquired hair discoloration (Table 3.7)

- Identify the cause of exogenous pigmentation
- Cut the damaged discolored portion of the hair
- Utilize shampoos containing chelating agents (sodium EDTA, D-penicillamine)
- Green hair can be discolored by immersion in 3% hydrogen peroxide solution.



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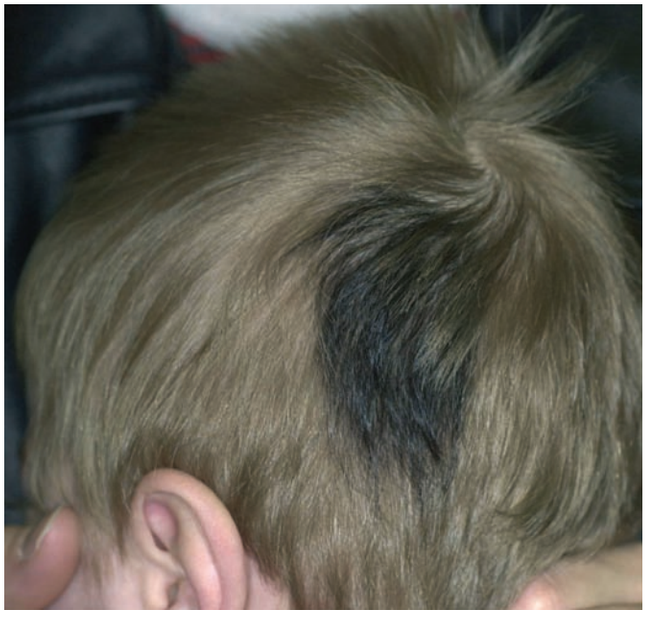

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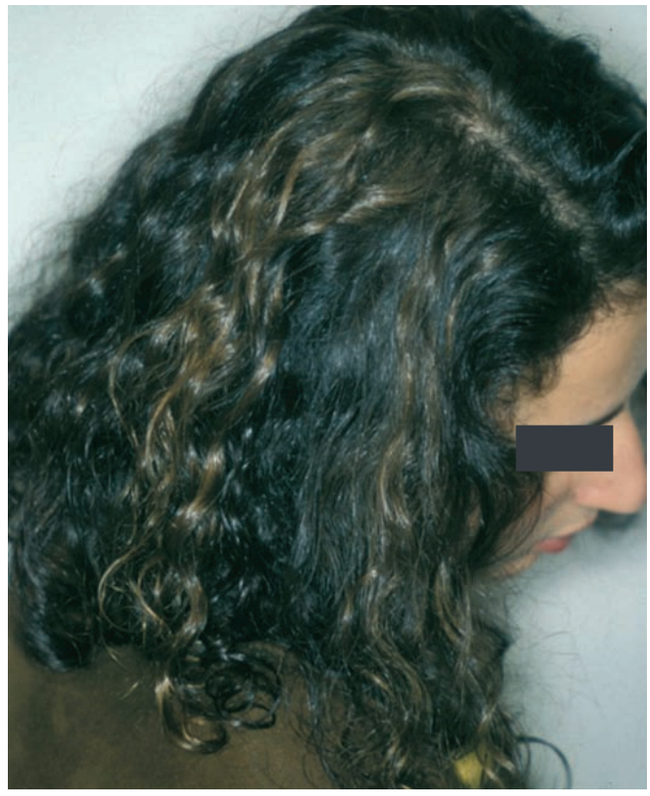

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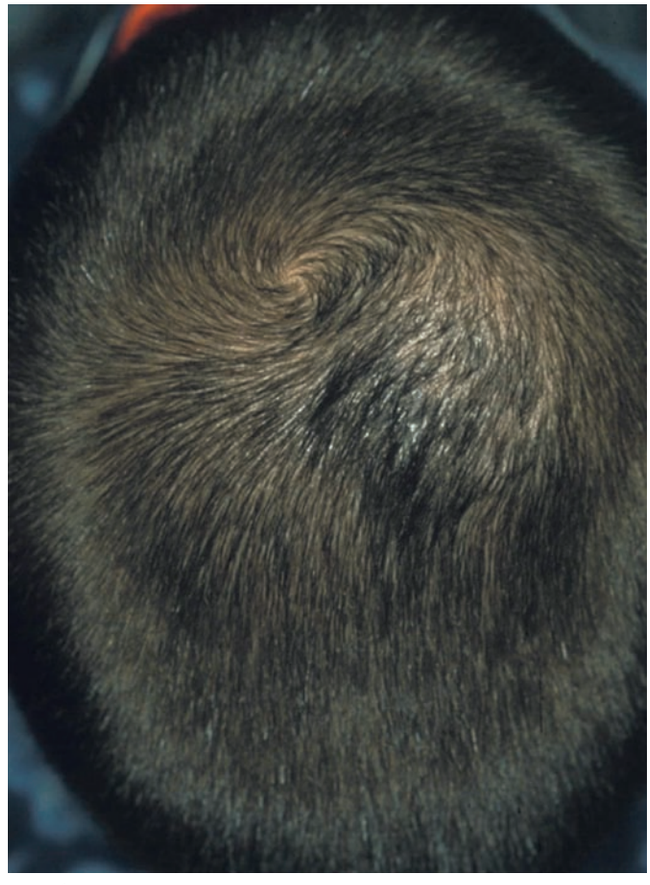


3.14

Figures 3.11–3.14
Scalp mosaicism.

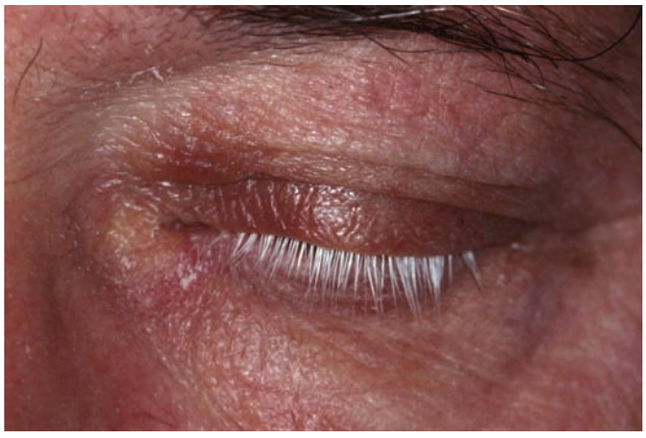
Figure 3.15
Greenish discoloration of the hair due to deposition of copper in severely weathered long hair.

Table 3.6 Causes of hair discoloration

Green hair	Chlorine Chromium Cobalt Copper (occupational exposure, cosmetics, tap water, swimming pool water) Minoxidil (topical) Nickel Selenium sulfide shampoo Tar shampoo Yellow mercury oxide
Blue hair	Cobalt Indigo
Red hair	Chinoform Trinitrotoluene (TNT)
Yellow hair	Anthralin Cigarette smoking Minoxidil (topical) Picric acid Resorcin

Table 3.7 Changes in the hair color: clues for diagnosis

Clinical features			*Clues for diagnosis*
White	Diffuse whitening	Albinism	Presence since infancy, ocular and skin depigmentation
		Alopecia areata	Diffuse alopecia due to loss of pigmented hairs
	Tufts of white hair (poliosis)	Alopecia areata	Previous history of patchy hair loss
		Vitiligo	Vitiligo of the corresponding scalp
		Piebaldism	Presence since childhood, triangular white forelock
		Waardenburg syndrome	Family members affected, deafness, dystopia canthorum, heterochromia iridis
Lightening	Diffuse lightening	Drugs	
		Nutritional disorders	
	Tufts of light hair	Hair weathering	
Darkening	Diffuse darkening	Hair regrowth (chemotherapy, alopecia areata)	



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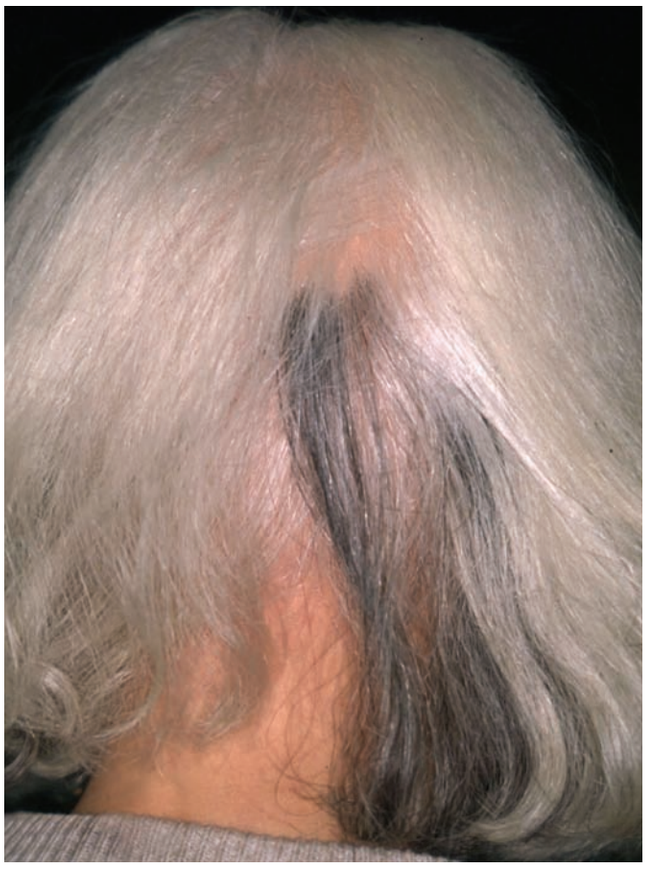

3.17

Figures 3.16, 3.17
Hair regrowth in alopecia areata may consist in white hair.

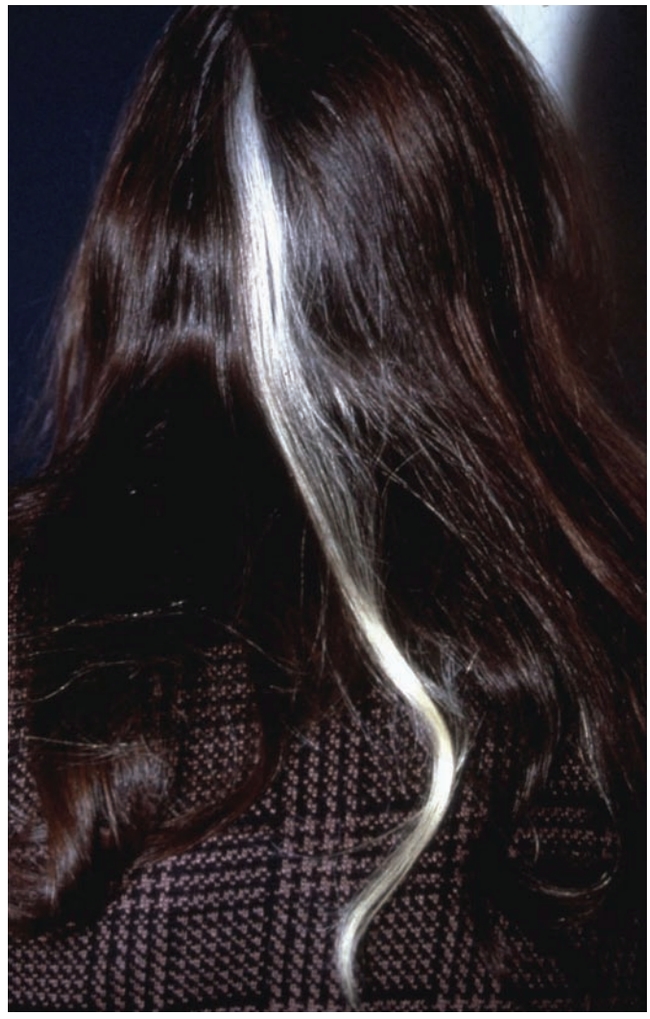

Figure 3.18
A tuft of white hair may persist forever after regrowth in alopecia areata.

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4 Hair shaft disorders

Hair shaft disorders can be congenital or acquired, the latter most commonly caused by mechanical, physical or chemical traumas.

Congenital hair shaft disorders

Congenital hair shaft disorders are usually evident from birth or the first months of life. The abnormalities are usually limited to the scalp, but may involve other terminal hair and rarely body hair. Congenital hair shaft disorders may be classified into two main groups:

1. Hair shaft disorders causing increased fragility and therefore hair breakage with patchy or diffuse alopecia.
2. Hair shaft disorders without hair fragility. These may interfere with hair texture and luster but do not produce alopecia.

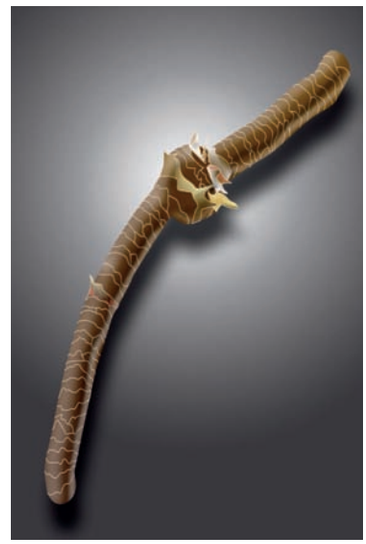
Congenital hair shaft disorders associated with fragility

Trichorrhexis invaginata (bamboo hair)

Diagnostic clue
The hair shaft shows multiple knots along its length (Figures 4.1–4.3). The knots consists of a proximal cup-shaped portion and a distal ball-shaped portion resembling the ball and cup joint of bamboo. Hair breakage occurs at the nodes.

Clinical features
Hair of patients with trichorrhexis invaginata is dry, dull, fragile and short. Hair breakage mostly affects scalp areas exposed to friction. Alopecia may be severe in some cases. The hair shaft disorder frequently affects eyelashes, eyebrows, as well as secondary sexual hair. The eyebrows usually show multiple bamboo nodes, and may present the abnormality, even when the scalp hair, which improves with age, appears normal.

Associated findings
Trichorrhexis invaginata may be isolated, but most commonly is part of Netherton disease (MIM 256500) (Figures 4.4, 4.5), a rare autosomal recessive genodermatosis which combines ichthyosis, bamboo hair and atopic dermatitis (Table 4.1).



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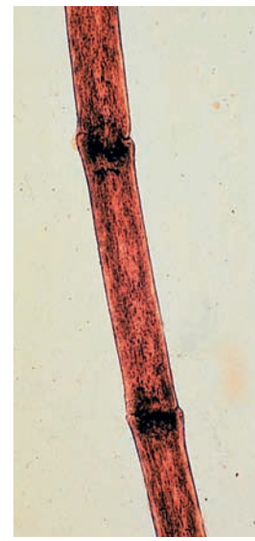

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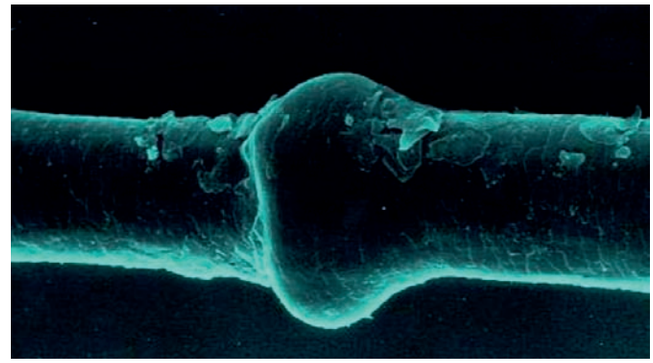
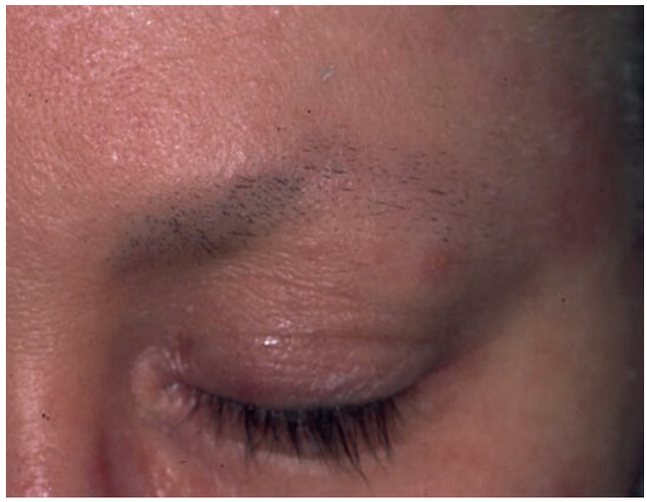


4.3

Figures 4.1–4.3
Trichorrexis invaginata.

Table 4.1 Netherton's syndrome

Autosomal recessive
More frequent in females
Ichthyosis
Linearis circumflexa
Lamellar
Vulgaris
X-linked
Trichorrhexis invaginata
Atopy (75% of cases)



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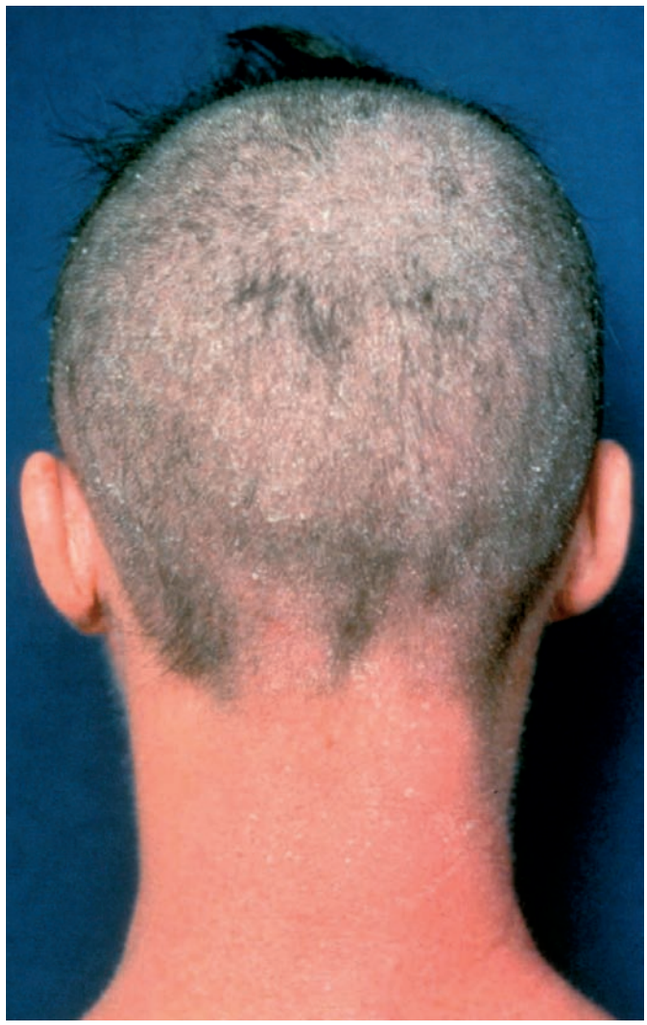


4.5

Figures 4.4, 4.5

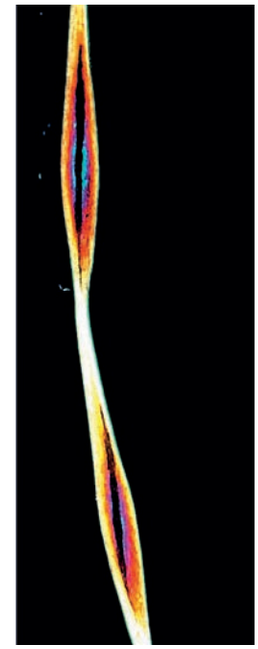
Netherton disease: trichorrexis invaginata, ichthyosis and atopic dermatitis.

4.6

Figures 4.6–4.8

Monilethrix: the hair shaft has a beaded appearence. Medulla is absent in the internodes.

4.7a



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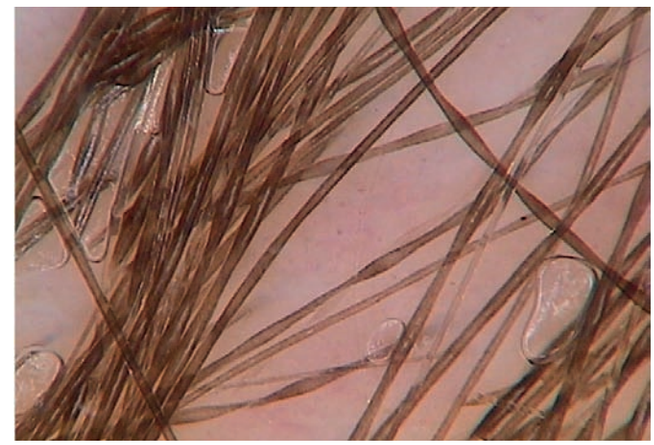

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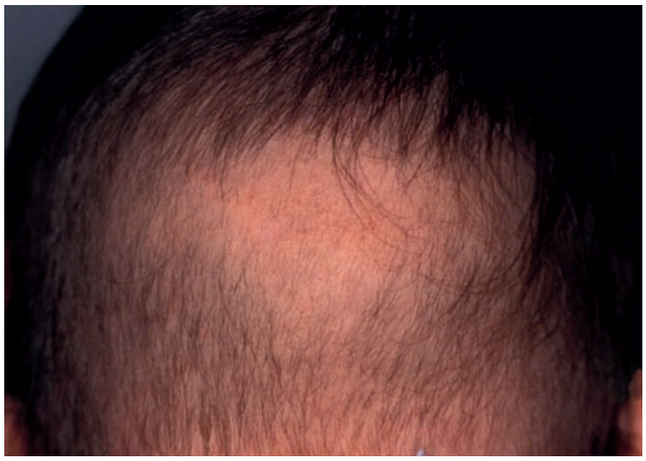

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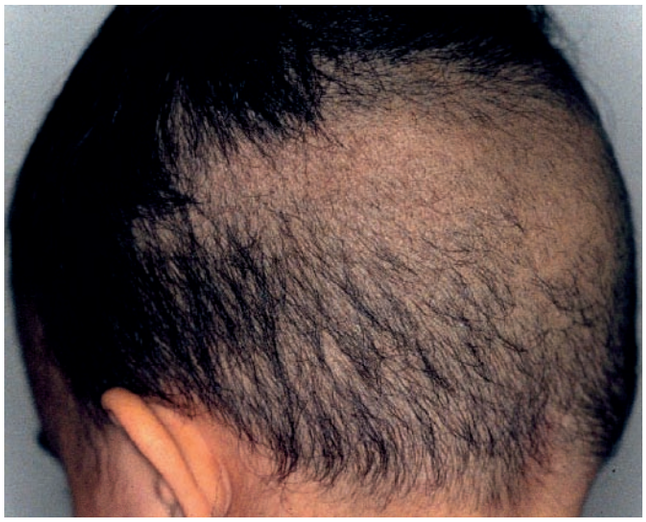

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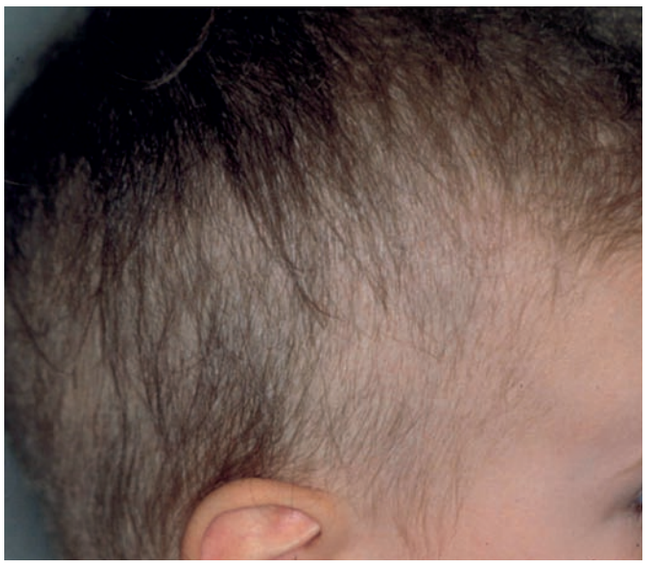


4.11

Figures 4.9–4.11
Monilethrix: alopecia is more severe in the areas of friction.

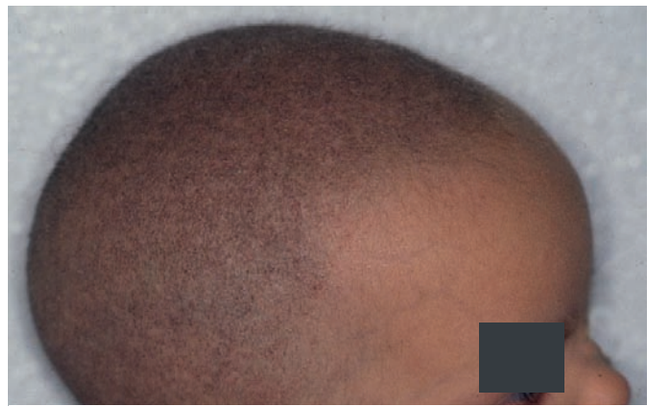
Ichthyosis, which may be present at birth as congenital erythrodermia, is in most cases ichthyosis linearis circumflexa, characterized by migratory serpiginous circinate plaques of scaly erythema with a double-edged scale at the advancing borders.

Genetics
Netherton's syndrome has been mapped to chromosome 5q32 and the gene has been identified as SPINK5 which encodes the serine-protease inhibitor LETKI.

Prenatal diagnosis of Netherton's syndrome is possible by molecular analysis of SPINK5.

Monilethrix (MIM158000, 177750, 252700)

Diagnostic clue
The hair shaft has a beaded appearance due to the presence of elliptical nodes (Figures 4.6–4.8), which have the diameter of normal hair and are medullated, regularly



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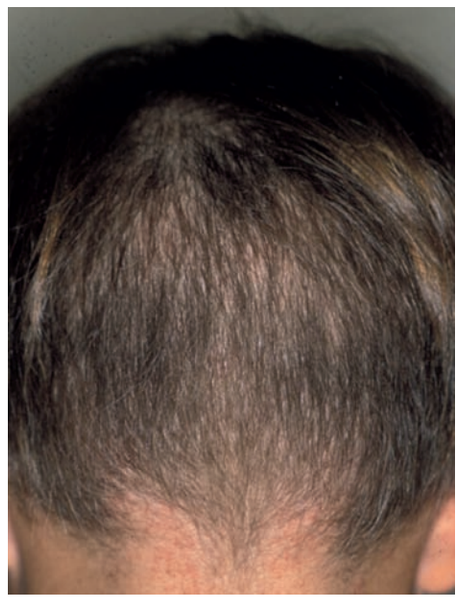


4.13

Figures 4.12, 4.13
Monilethrix in two siblings.

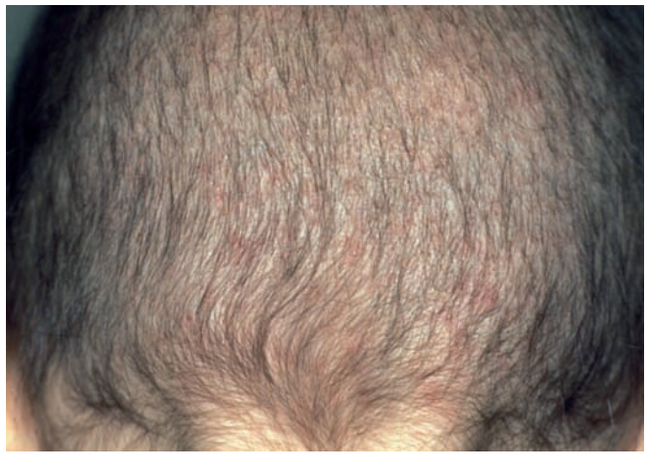


Figure 4.14
Monilethrix: note hyperkeratotic papule of the neck.

separated by internodes which are narrow, devoid of medulla and are the site of fracture.

Clinical features
The hair is dull, fragile and breaks easily, especially in the nape and occipital areas (Figures 4.9–4.11). The severity of the disease may considerably vary even among members of the same family (Figures 4.12, 4.13). Follicular keratosis of the affected scalp and keratosis pilaris are typical (Figure 4.14).

Monilethrix does not affect lanugo hair and usually becomes evident with growing of mature hair. At birth the scalp hair is either normal or absent; in the latter case the scalp has a shaved appearance. Scraping of the scalp may show the typical beads resulting from broken monilethrix hairs. Hair fragility improves with age.

Genetics
Monilethrix is inherited as an autosomal dominant condition with variable expression. Several mutations in the human basic hair keratins hHb1 and hHb6 have been reported. Linkage to the type II keratin cluster is on 12q13.

Pili torti

Diagnostic clue
Pili torti are flattened and present twisting of the hair shaft through 180° at irregular intervals (see Table 4.2 for differential diagnosis with monilethrix) (Figures 4.15, 4.16). A small number of these hairs can be frequently found both in normal scalp, in association with other hair shaft abnormalities and in several rare genetic syndromes (Table 4.3).

Syndromes associated with pili torti:

- Menkes syndrome (MIM 309400). Menkes kinky hair syndrome is an X-linked autosomal recessive

Table 4.2 Hair microscopy

Pili torti	Hair shaft shifts in and out of focus as the lens traverses its length
Moniletrix	The wide nodes and narrow internodes remain in the same plane of focus

4.15

4.16

Figures 4.15, 4.16
Pili torti.

Table 4.3 Conditions associated with pili torti

Bazex syndrome
Beare syndrome
Björnstad syndrome
Citrullinemia
Conradi–Hünnermann syndrome
Crandall's syndrome
Familial acnes conglobata
Hypohidrotic ectodermal dysplasia
Menkes syndrome
Rapp–Hodgkin syndrome
Salti–Salem syndrome
Trichothiodystrophy

defect of copper metabolism, which combines pili torti and progressive neurological disfunction. The hair is fine, silver or white and fragile and has a kinky or wiry appearance. The Menkes' gene has been mapped to Xq13.3.

- Björnstad syndrome (MIM 262000). This autosomal recessive syndrome associates pili torti with hearing problems varying from deafness to reduced hearing in defined frequencies. The hair is short, dry and fragile with severe alopecia in some cases. The syndrome has been mapped to the gene locus 2q34–36.

Trichorrhexis nodosa (Table 4.4)

Diagnostic clue
The hair shaft shows one or more knots (Figures 4.17–4.19) that easily break off producing two brush-like tips (Figure 4.20).

Clinical features
The hair shaft shows small whitish knots visible with the unaided eye (Figure 4.21). The hair is fragile and short due to breakage.

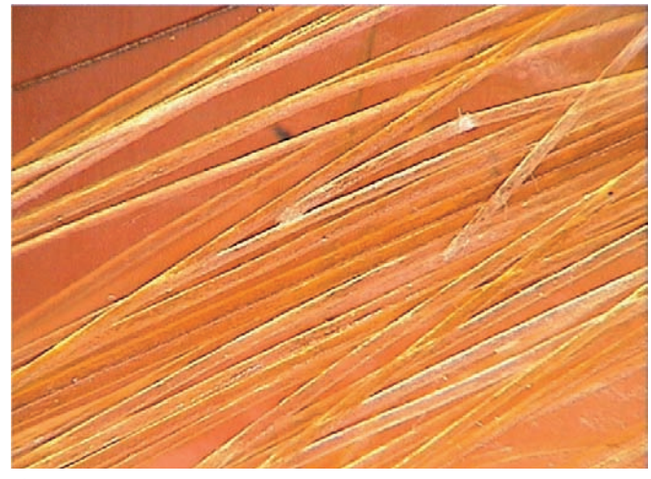
Acquired trichorrhexis nodosa is frequent, representing a typical sign of hair weathering (see page 33). Congenital trichorrhexis nodosa is very rare.

Table 4.4 Conditions associated with congenital trichorrhexis nodosa

Arginino-succinic aciduria
Citrullinemia
Defective teeth and nails
Follicular hyperkeratosis
Menkes syndrome

4.17

4.18



4.19

Figures 4.17–4.19
Trichorrexis nodosa.

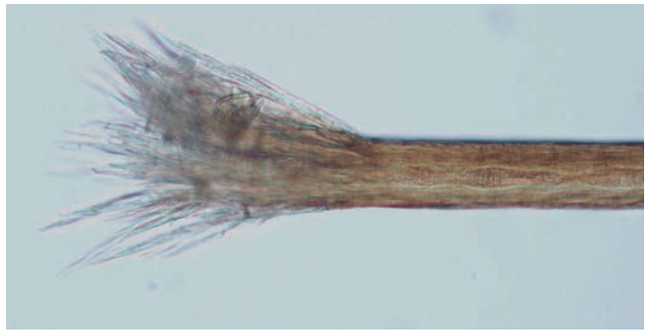


Figure 4.20
Trichorrexis nodosa: after breakage the shaft presents a bush like tip.

Figure 4.21
Multiple knots of trichorrexis nodosa along the hair shaft.

Trichothiodystrophy (MIM 601675)

Diagnostic clue
Trichothiodystrophy is due to a reduction in the sulfur and cystine content of hair. The condition is autosomal recessive. The affected hair has been assimilated to a tiger tail because of a typical alternating birefringence under polarized microscopy due to the presence of alternated transverse dark and bright bands along the shaft (Figure 4.22). Trichorrhexis nodosa, trichoschisis and pili torti are often found. The presence of some 'tiger tail hair' is not exclusive of trichothiodystrophy and may be found in normal individuals. It is important to analyze the proximal portion of the hair shaft to avoid changes due to weathering. Amino acid analysis may confirm the diagnosis revealing a 50% decrease in cystine and sulfur content.

Clinical features
The hair is brittle, sparse, dry, unruly and short (Figure 4.23). Eyebrows and eyelashes can also be affected. Brittle nails are often associated.

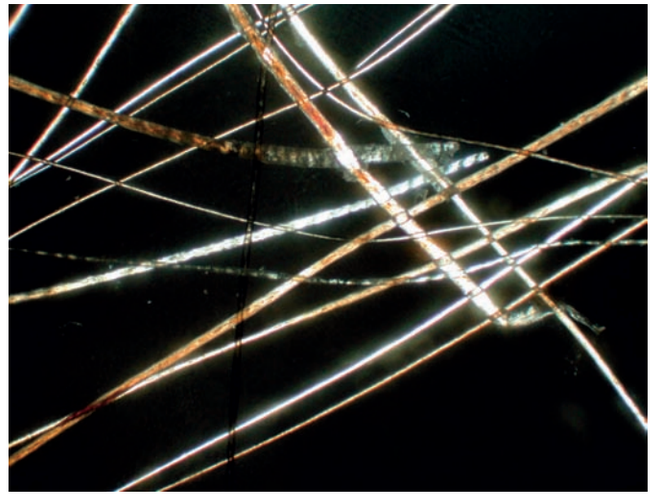


Figure 4.22
Trichothyodystrophy: typical tigertail aspect of the hair shaft at polarized microscopy.

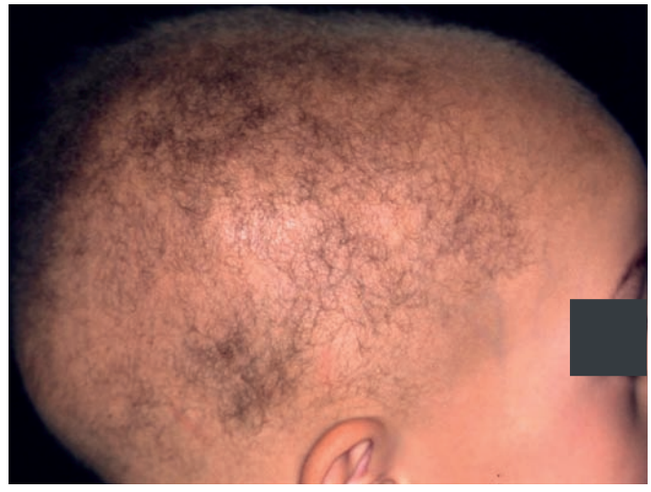


Figure 4.23
Trichothyodystrophy: severe alopecia with sparse, short, dry, unruly hair.

Table 4.5 Trichothiodystrophy

Congenital hair defect + mental retardation decreased fertility growth retardation	→ BIDS
+ ichthyosis	→ IBIDS
+ photosensitivity (40–50%)	→ PIBIDS
Marinesco–Sjögren syndrome	

Figure 4.24
Pili bifurcati.

Associated features
The presence of associated features permits to distinguish several syndromes (Table 4.5). Photosensitivity, with mutations of the XPD, XPB or TTDA gene, is present in about 50% of cases but there is no predisposition to skin cancers.

Pili bifurcati

Diagnostic clue
The hair shaft shows intermittent longitudinal splitting along its length (Figure 4.24). Each branch has its own cuticle. Pili bifurcati may occur in association with other hair abnormalities such as pili canaliculi and monilethrix.

Clinical features
The condition is rare and may produce a diffuse alopecia, more evident in some areas.

Treatment

Gentle handling of hair to minimize breakage is useful in all patients with hair shaft fragility.

Monilethrix
- Oral retinoids (etretinate) (D)
- Oral vitamin A + vitamin E (E)
- 2% topical minoxidil (E)
- Oral L-cystine 1 g/day (E).

Congenital hair shaft disorders without fragility

Diffuse partial woolly hair (DPWH)

Diagnostic clue
This is a rare disturbance of scalp hair characterized by the concomitant presence of two distinct hair populations (Figures 4.25, 4.26):

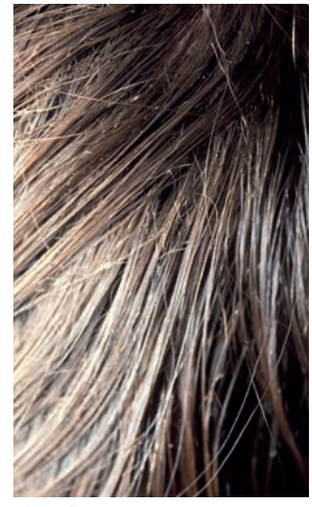
- straight, long, pigmented hairs
- short, fine, hypopigmented and curly hairs (approximately 20–30%).

At SEM the curly shafts are thin (30–40 μm), have a flattened section and show single torsions and canalicular formations.

DPWH is probably a consequence of follicle miniaturization and therefore a variety of mild AGA.

A variant of DPWH affecting the distal part of the hair shaft of prepubertal girls has been described as acquired partial curling of hair (APCH). This condition appears to be reversible.

4.25



4.26

Figures 4.25, 4.26
Diffuse partial woolly hair.

Figure 4.27
Pili annulati: alternating light and dark bands.

Clinical features
The condition may be inherited as an autosomal dominant trait and most commonly affects females. Patients complain of hair loss, reduced hair growth and hair roughness. Close examination reveals the two populations of hair.

Pili annulati

Diagnostic clue
The hair shaft presents alternating light and dark bands (Figure 4.27) that are visible with the unaided eye.

Clinical features
Pili annulati is an autosomal dominant condition with variable expression.

The hair has a shiny, spangled appearance (Figures 4.28, 4.29). The abnormality is more evident on blond or white hair. Pili annulati usually do not produce hair fragility, but affected hair is more susceptible to weathering due to the presence of air-filled cavities of the cortex in correspondence with dark bands (Figures 4.30, 4.31).

Axillary, beard and pubic hair can also be affected.

Although pili annulati have been occasionally reported in patients with alopecia areata, the association is coincidental.

4.28

4.29

Figures 4.28, 4.29
Pili annulati.

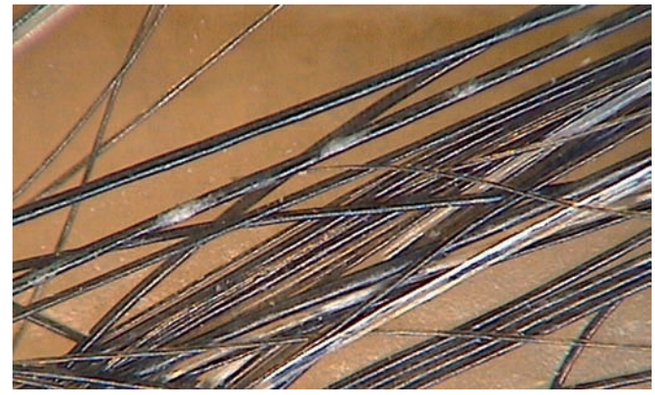


Figure 4.30
Pili annulati: dermoscopy shows the air-filled cavities along the hair shaft.

Figure 4.31
Pili annulati: dermatoscopic aspect, note trichorrexis nodosa due to hair fragility.

Woolly hair (Table 4.6)

Diagnostic clue
Woolly hair is twisted, does not group in locks, does not lie flat on the scalp and resembles African hair (Figure 4.32).

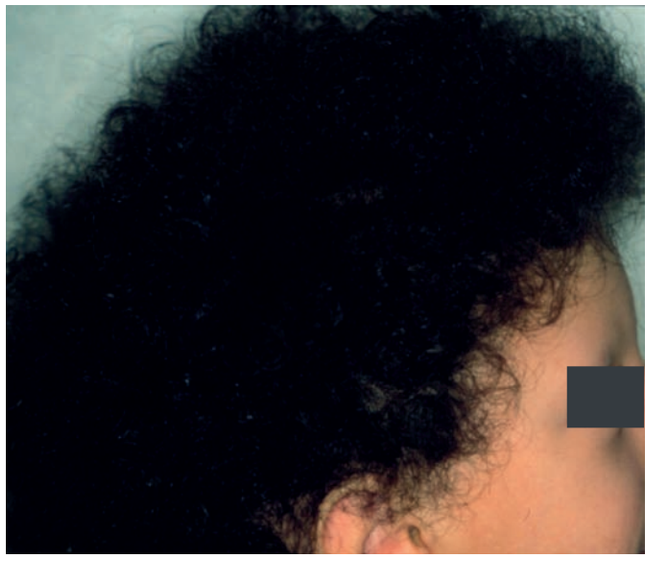
Table 4.6 Conditions associated with woolly hair

Acral keratoderma – caries (HWH)
Palmoplantar keratoderma – cardiac abnormalities (HWH)
Loose anagen hair (FWH)
Congenital ichthyosis, follicular atrophoderma (FWH)

Figure 4.32
Woolly hair.

Three types of woolly hair can be distinguished according to clinical features and inheritance:

- Hereditary woolly hair (HWH)
- Familial woolly hair (FWH)
- Woolly hair nevus (WHN).



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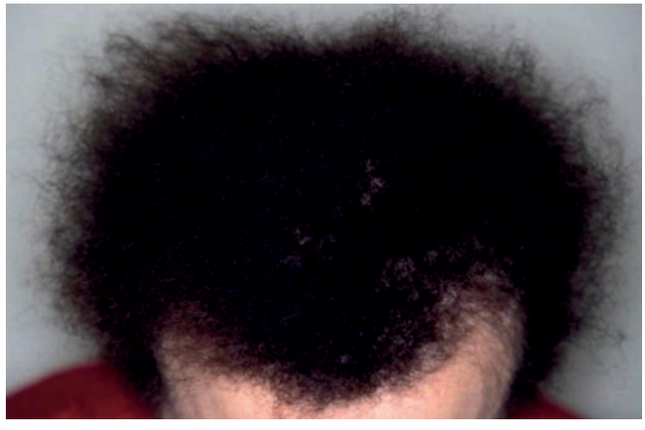

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4.35

Figures 4.33–4.35
Hereditary woolly hair.

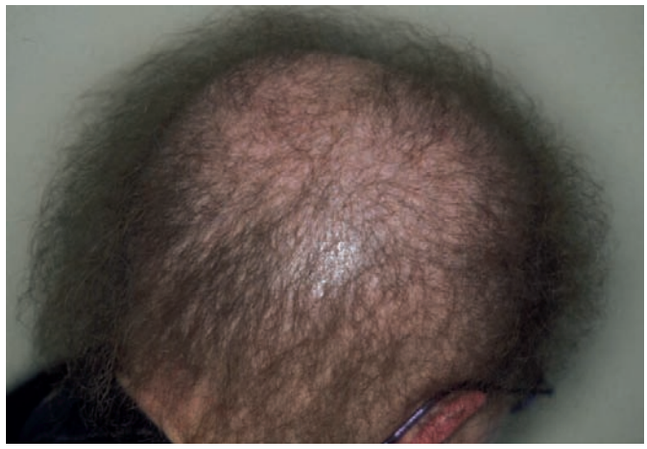


Figure 4.36
Familial woolly hair.

Clinical features
Hereditary woolly hair (HWH) is autosomal dominant inherited and not associated with hair fragility (Figures 4.33–4.35). The hair color is variable and the manageability improves with age. Hereditary woolly hair may be associated with palmoplantar keratoderma and cardiac abnormalities in some families (Naxos' syndrome, MIM 601214), with mutations in desmoplakin and plakoglobin genes.

Familial woolly hair (FWH) is autosomal recessively inherited and associated with hair thinning (Figure 4.36). The hair is sparse, thin and short. Severe alopecia is common.

Woolly hair nevus (WHN) is present at birth without a family history (Figure 4.37). An isolated patch of woolly hair is present among normal hair. The condition is frequently (50% of cases) associated with epidermal nevus of the neck or arm.

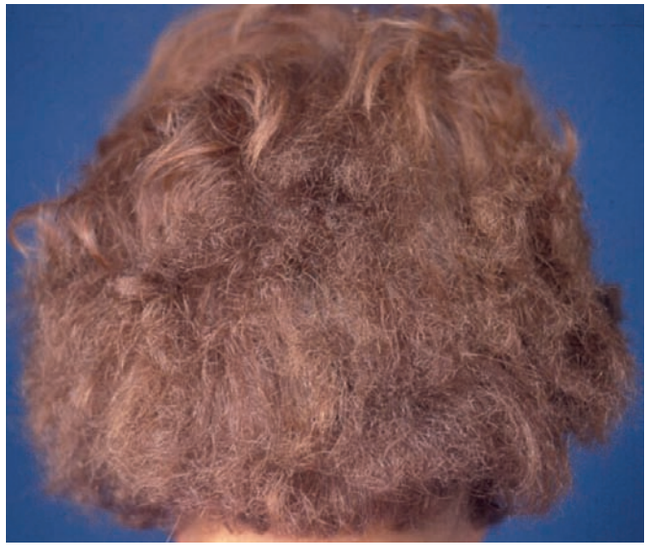


Figure 4.37
Woolly hair nevus.

Uncombable hair (pili trianguli et canaliculi)

Diagnostic clue
Uncombable hairs have a triangular or kidney section and a flattened surface that reflects light (Figure 4.38).

The diagnosis can be made by scalp dermoscopy or SEM which show the typical triangular or reniform-shape with longitudinal grooving and flattening in more than 50% of hairs (Figure 4.39).

A small number of uncombable hair can be detected in the general population.

Clinical features
Uncombable hair inheritance is variable. The hair is usually blond, dry and unruly and totally resists all efforts of styling. The overall appearance resembles synthetic doll-hair (Figures 4.40, 4.41). Eyebrows and eyelashes are normal. The condition usually appears during the first years of life and considerably improves with age, when the hair becomes long (Table 4.7).

Table 4.7 Conditions associated with uncombable hair

Ectodermal dysplasia
Retinal dystrophy
Juvenile cataract
Digit abnormalities
Dental abnormalities
Phalangoepiphyseal dysplasia
Mental retardation

Treatment

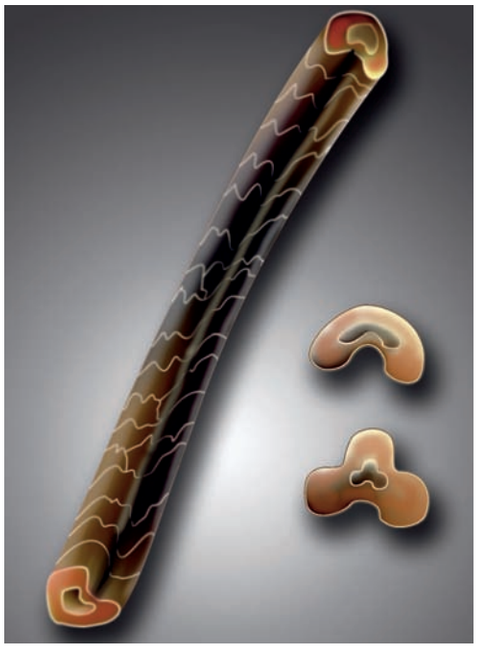
Uncombable hair (pili trianguli et canaliculi)
- Biotin 0.3 mg three times/day (E).

Acquired hair shaft disorders

Acquired hair shaft disorders associated with fragility

Hair weathering

The environment causes daily repetitive injuries to the hair shaft resulting in hair damage that can not be repaired and is more evident in the distal part of long



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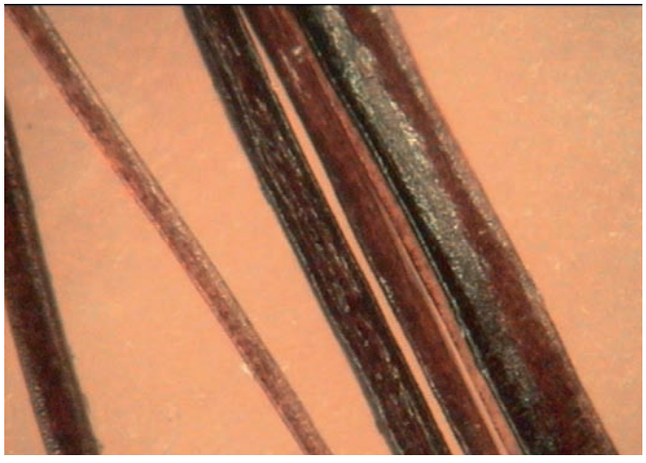

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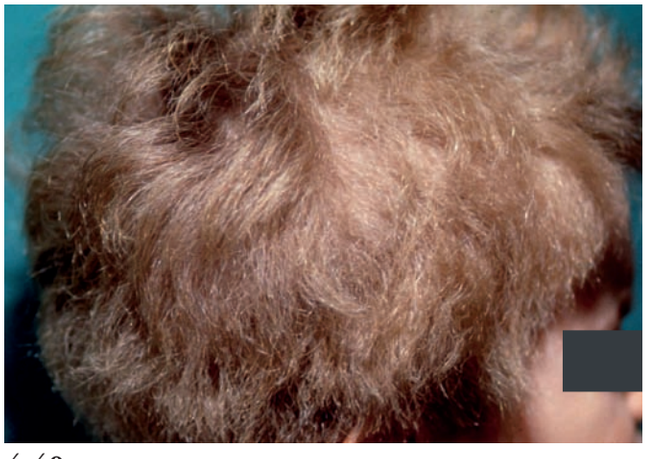


4.40

Figures 4.38–4.40
Uncombable hair.

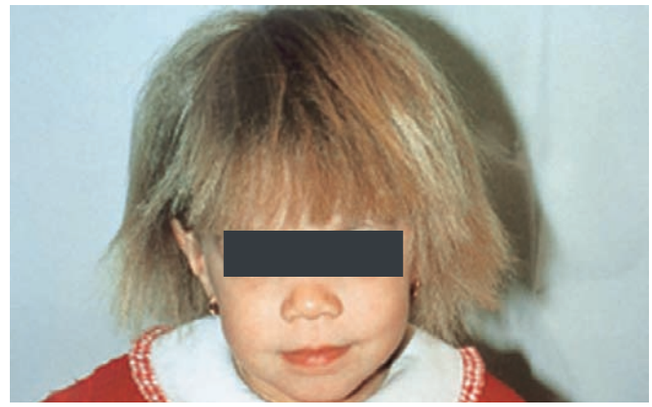


Figure 4.41
Uncombable hair.

hair shafts (Table 4.8). The resistance of hair of different individuals to mechanical, physical and chemical injuries is variable and probably inherited. Hair damage first involves the cuticle with uplifting and loss of the cuticular cells resulting in exposure and damage of the cortex (Figures 4.42–4.44). Hair cortex damage results in breakage of amino acids with formation of negative charges on the hair shaft surface.

Table 4.8 Causes of hair weathering

Environmental	UV Salt water/chlorinate water Wind Pollution
Physical	Hair dryer Hot combers Hot straighteners
Mechanical	Brushing Hair styling
Chemical	Dyeing/bleaching Perming

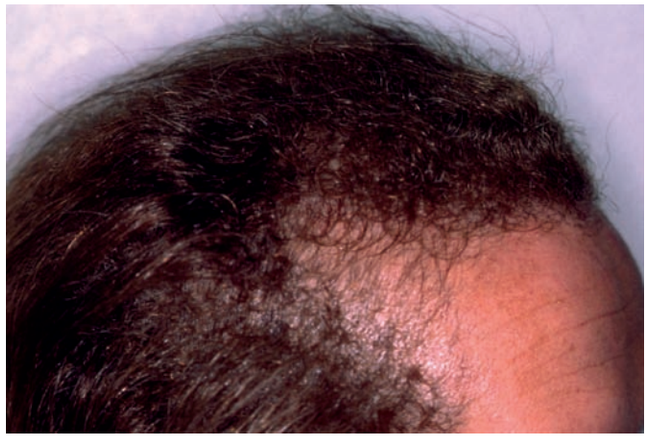
Diagnostic clue
Hair weathering produces hair breakage, trichorrhexis nodosa and trichoptilosis.

Clinical features
Damaged hair is dry, dull, frizzy, opaque, difficult to comb and more porous.

Trichorrhexis nodosa

Diagnostic clue
See congenital trichorrhexis nodosa (page 27).

4.42



4.43

4.44

Figures 4.42–4.44
Hair weathering.

Clinical features
The hair presents single or multiple white knots along the distal part of the hair shaft. Fracturing occurs in correspondence of the knots. Presence of trichorrhexis nodosa in the distal part of some hair shafts is common in subjects with long hair as a consequence of weathering. This common feature should be distinguished from generalized trichorrhexis nodosa, which involves a substantial percentage of scalp hair and probably represents an intrinsic weakness of the hair shaft to weathering. In generalized trichorrhexis nodosa the abnormality involves the distal shaft even though the hair is not necessarily long. Circumscribed patches of trichorrhexis nodosa are a consequence of rubbing or scratching (see page 102) and may affect also the beard or other terminal hair (see page 155). Proximal trichorrhexis nodosa is a typical consequence of aggressive hair styling in blacks.

Trichoptilosis

Diagnostic clue
The hair shaft shows a longitudinal splitting, 2–3 cm long, of its distal end. Central trichoptilosis can also occur. The bifurcated hair shaft is not surrounded by cuticle (Figure 4.45).

Clinical features
Trichoptilosis is the most common sign of weathering in long hair. The distal part of the shaft is frizzy and dry.

Trichoschisis

Diagnostic clue
The hair shaft shows a sharp transverse fracture, which is better seen under polarizing microscopy (Figure 4.46).

Figure 4.45
Trichoptilosis.

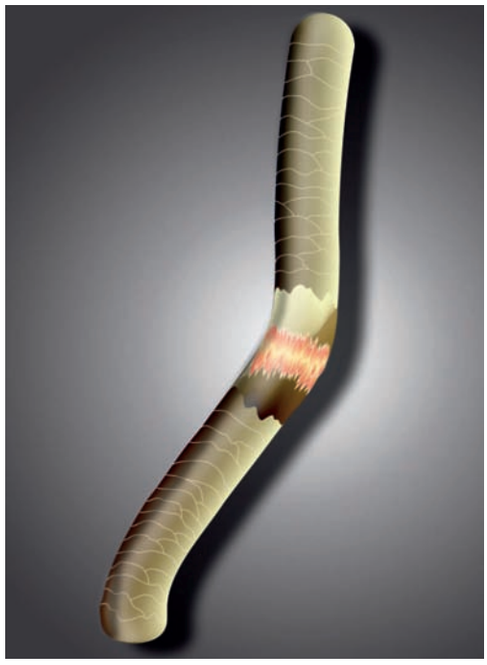


Figure 4.46
Trichoschisis.

Treatment

Hair weathering
- Utilize shampoos for dry/damaged hair
- Regular use of conditioner
- Regular cutting of distal tips
- Reduce mechanical/physical/chemical injuries
- Oral L-cystine 1 g/day (E).

Acquired hair shaft disorders without fragility

Acquired progressive kinking of the hair (APKH)

Diagnostic clue
Hair shaft examination of affected hair shows:

- irregular twisting and periodic reduction in the diameter at optical microscopy
- longitudinal grooves along the hair shaft at SEM.

Clinical features
The term APKH includes rather different conditions characterized by acquired curly, frizzy and lusterless hair (Table 4.9). These include:

1. Kinking of the hair over periauricular areas of the scalp (whisker hair). Whisker hair is short and curly and resembles the hair of the beard. This condition is strongly associated with severe AGA.
2. Acquired progressive kinking of androgen-dependent hair associated with thinning (Figure 4.47). This variety of acquired hair kinking represents a modality of onset of AGA associated with poor prognosis.

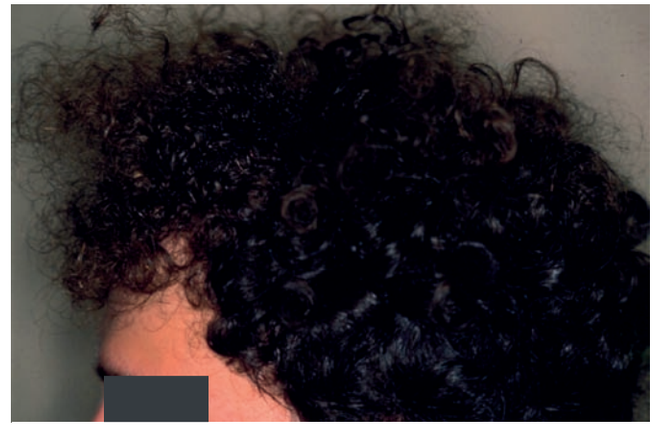


Figure 4.47
Acquired progressive kinking of the hair.

3. Rapidly progressive kinking of most or all the scalp hair without associated hair thinning (rare).
4. Acquired reversible hair kinking before or after puberty.
5. Acquired hair kinking involving a localized non-androgen-dependent area of the scalp.
6. Drug-induced hair kinking; may or not be reversible (see page 69).

Treatment

Acquired progressive kinking of the hair (APKH)
When localized to androgen-dependent scalp treat with finasteride 1 mg (E). Topical minoxidil is not effective (D).

Table 4.9 Differential diagnosis of kinky\curly hair

	Time of appearance	*Hair shaft*	*Involved area*	*Hair loss*	*Course*	*Clue for diagnosis*
DPWH	After puberty	Irregularly curled oval shape	Whole scalp ~ 20–30% of scalp hair	Yes	Progression	Short curly hairs intermingled with normal hair
APKH	After puberty in most cases	Irregularly curled oval shape	Androgen-dependent scalp	Yes	Progression	Curly hair in the frontotemporal scalp
			Non-androgen-dependent scalp	No	May be reversible	Localized tuft of curly hair
APCH	Prepubertal	Irregularly curled oval shape	Whole scalp, distal shaft	No	May be reversible	Abnormalities limited to the distal shaft
Woolly hair	At birth	Irregularly curled oval shape	Whole scalp	No AD	Stable	Negroid appearance of the hair
				Yes AR	Progression	Hypopigmented sparse negroid hair
Woolly hair nevus	At birth	Irregularly curled oval shape	Occipital scalp	No	Stable	Congenital localized tuft of curly hair
Matting	Adults	Weathering	Occipital scalp	No	May be reversible	Hair tangling in areas of friction
Uncombable hair	3–12 years	Kidney-shaped section	Whole scalp	No	Improvement	Spun glass silvery unruly hair

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5 Alopecia areata

Alopecia areata (AA) is a common form of non-cicatricial alopecia, characterized by patchy hair loss in the absence of inflammatory signs.

A genetic susceptibility to AA has been established and other family members are affected by the disease in about 30% of cases (Figure 5.1).

The disease affects both sexes at any age, but severe forms are more frequent in males and often start during childhood. Although AA frequently resolves spontaneously, relapses occur in a high percentage of patients.

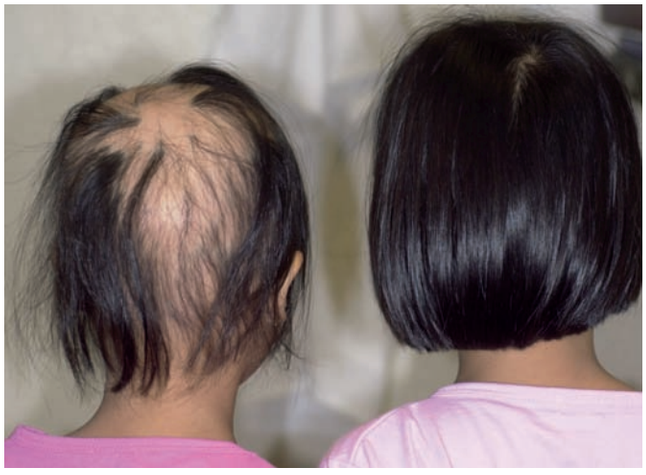
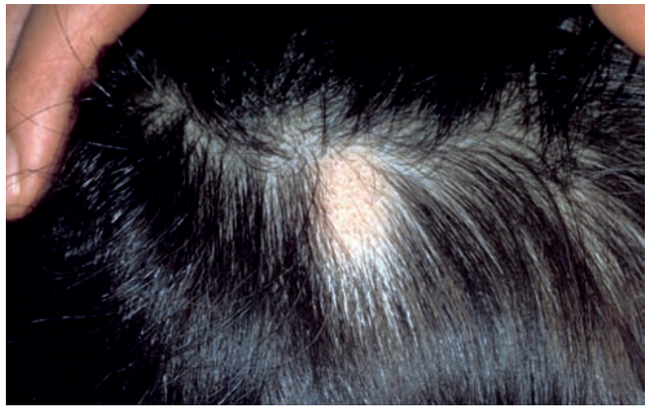


Figure 5.1
In monozigotic twins AA may present with different severity.

Clinical features

The disease usually starts abruptly with one or multiple patches of hair loss that usually enlarge in a centrifugal way (Figures 5.2–5.4). Diffuse shedding may or may not be present in the surrounding scalp. Exclamation point hairs are often present along the advancing border of the patch. The scalp is usually normal without signs of inflammation.

AA may involve any hairy body area (Figures 5.5–5.7), but is more common on the scalp and on the beard area. Rarely it may exclusively affect the eyelashes and/or the eyebrows (Figures 5.8–5.11). Severity of AA may be evaluated according to the percentage of scalp involvement. Severe forms affect all scalp (AT, alopecia totalis) (Figure 5.12) or all body hair (AU, alopecia universalis). Involvement of the scalp margins (ophiasis) (Figures



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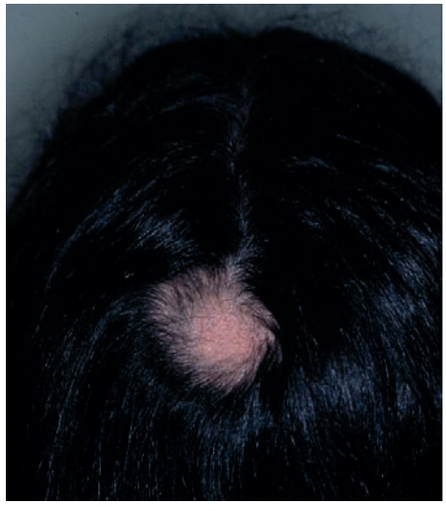

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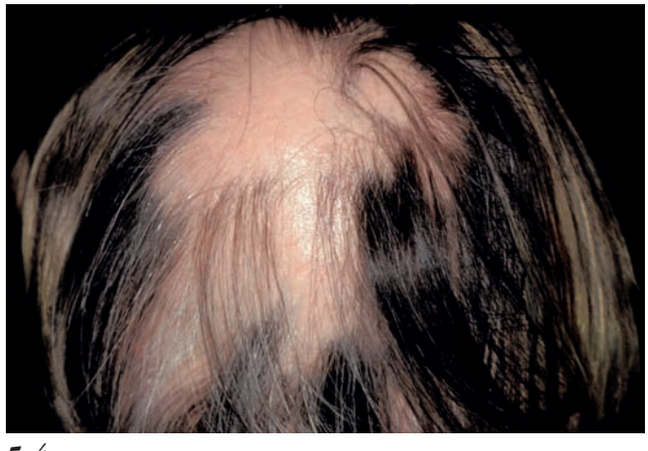
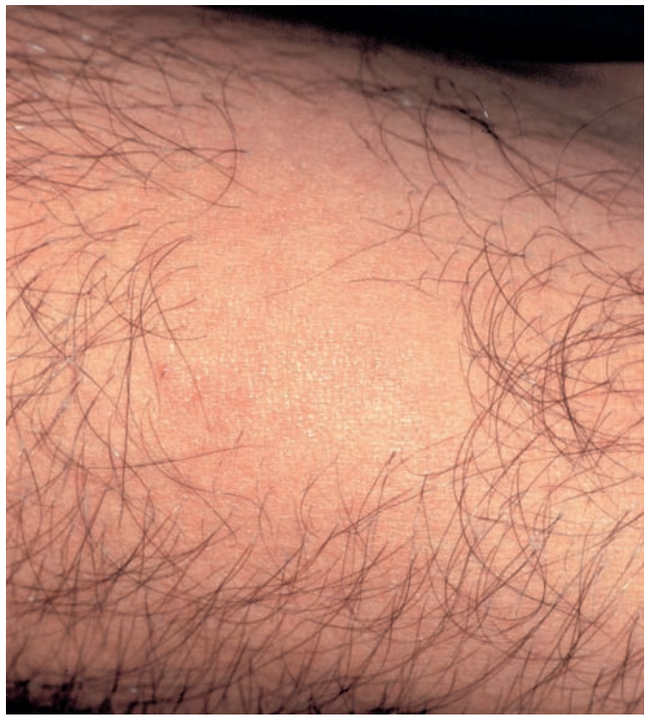


5.4

Figures 5.2–5.4
Alopecia areata: patches that enlarges in a centrifugal way.



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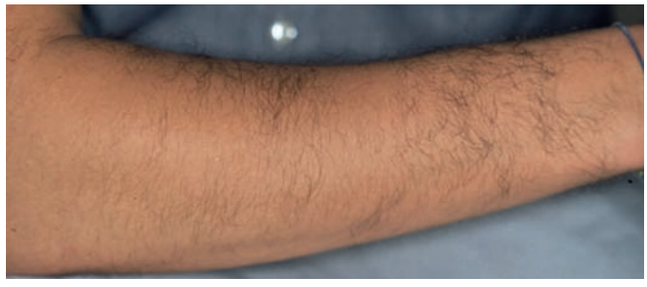


5.6

Figures 5.5, 5.6
A forearm involvement in AA.

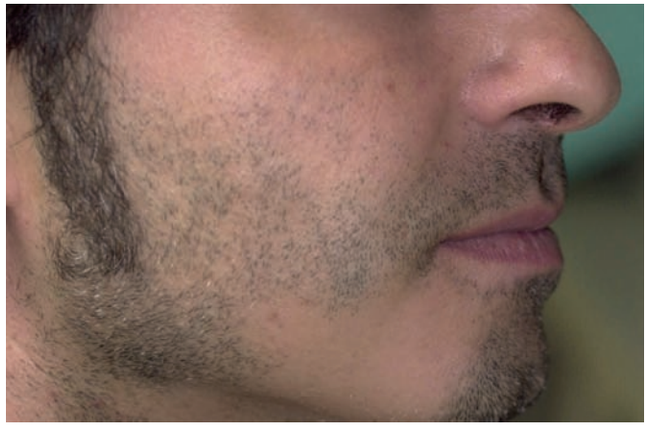
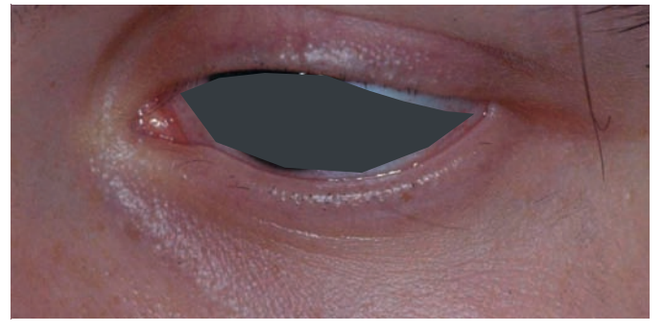


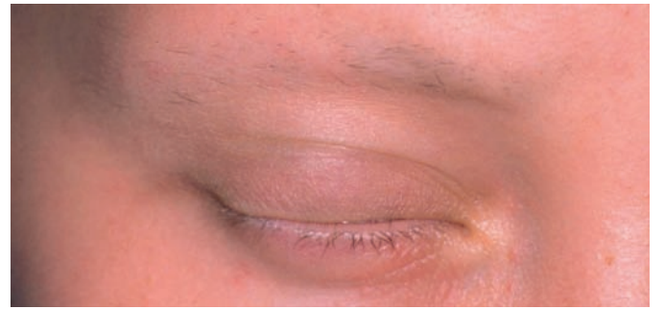
Figure 5.7
AA of the beard.

5.8



5.9

Figures 5.8, 5.9
AA of the eyelashes.



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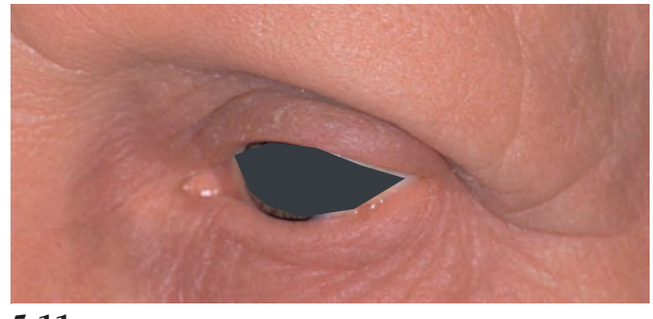

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Figures 5.10, 5.11
AA of the eyelashes and eyebrows.

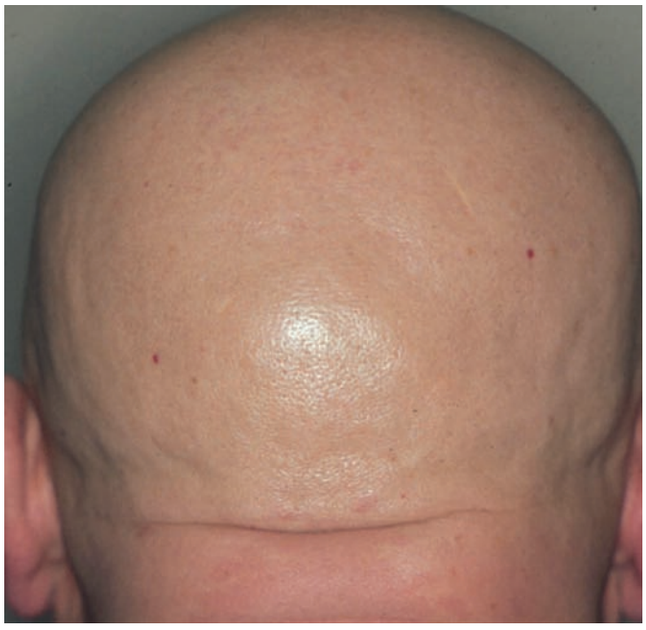
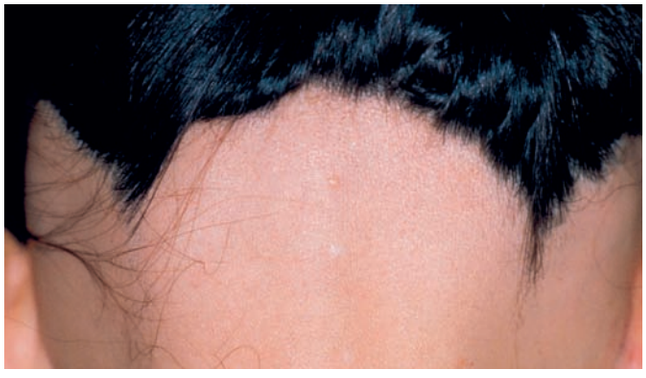


Figure 5.12
Alopecia totalis.



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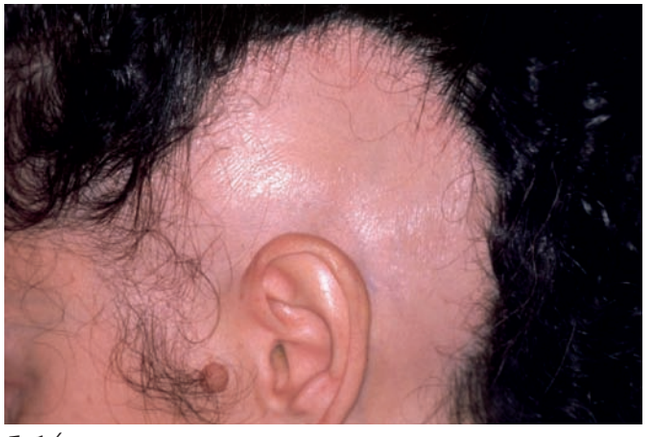

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Figures 5.13, 5.14
Ophiasis.

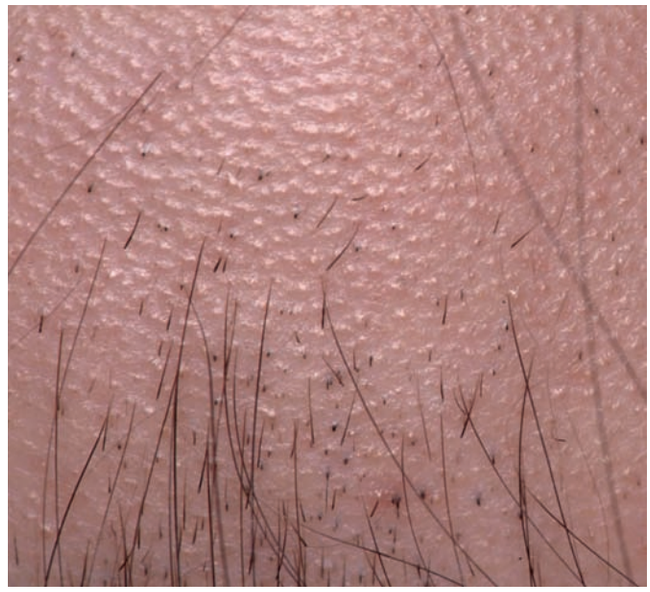
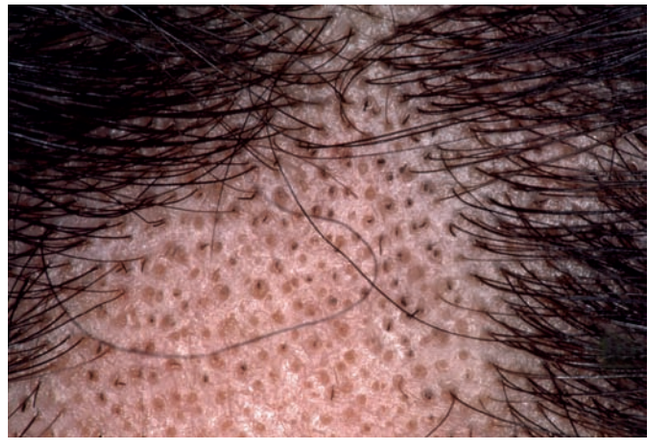


Figure 5.15
Exclamation point hairs.

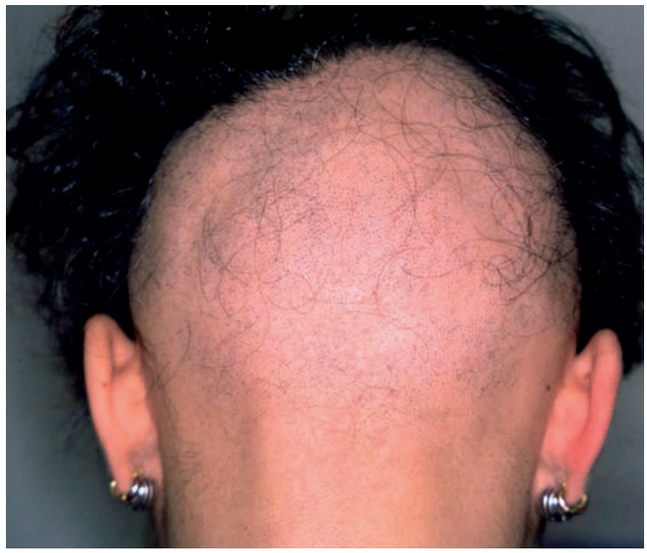


Figures 5.16
Dystrophic and cadaverized hairs.

5.13, 5.14) is associated with a poor prognosis. The affected scalp may be completely devoid of hair or may be covered by vellus, unpigmented, short hair. Acute AA is characterized by the presence of exclamation point hairs and cadaverized hairs (Figures 5.15–5.20).

Uncommon presentations include:

- Sudden whitening of the hair: results from selective involvement of pigmented hair. Patients complain that their hair has turned white in a few days (Figures 5.21, 5.22). Hair thinning is more or less evident. The prognosis is usually good. White hairs are also typically seen in the regrowing phase of AA (Figure 5.23).



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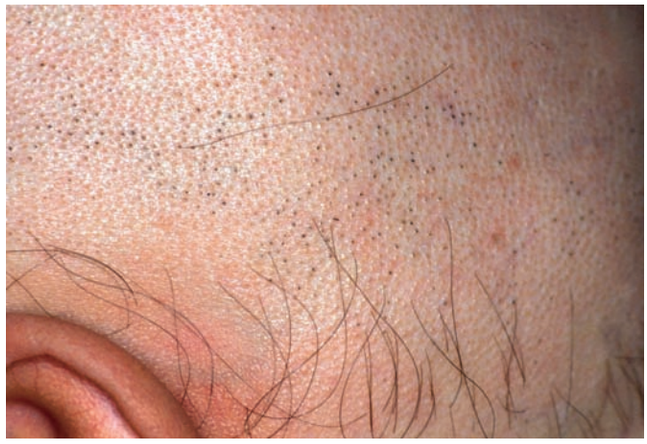

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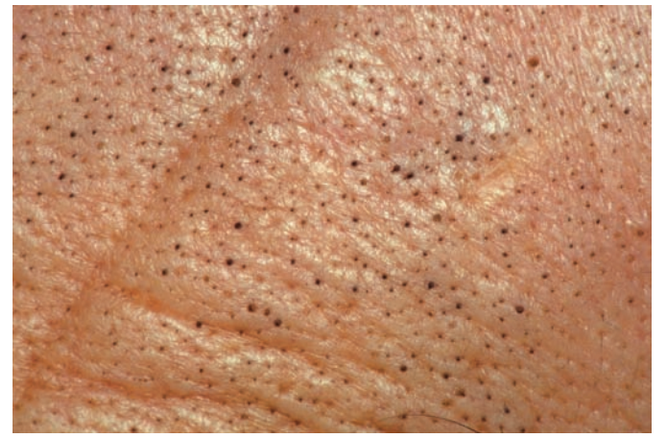

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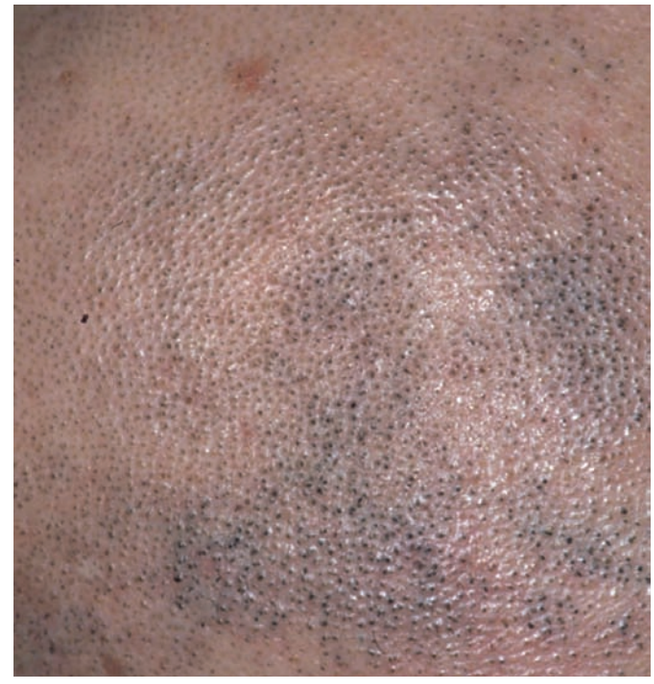
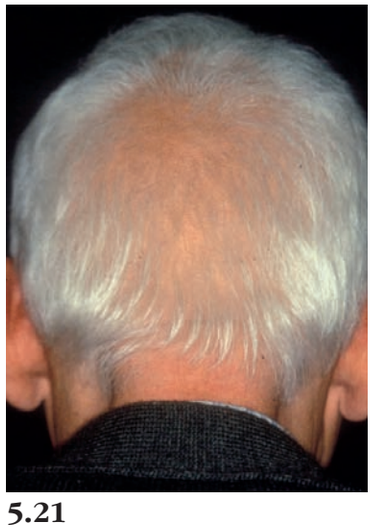


5.20

Figures 5.17–5.20
Cadaverized hairs in acute AA.



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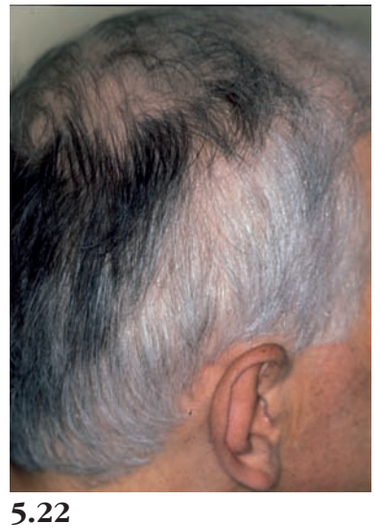

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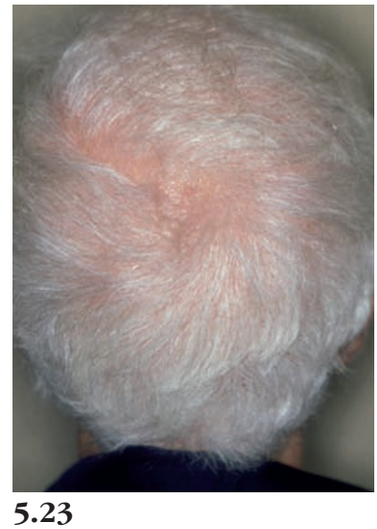
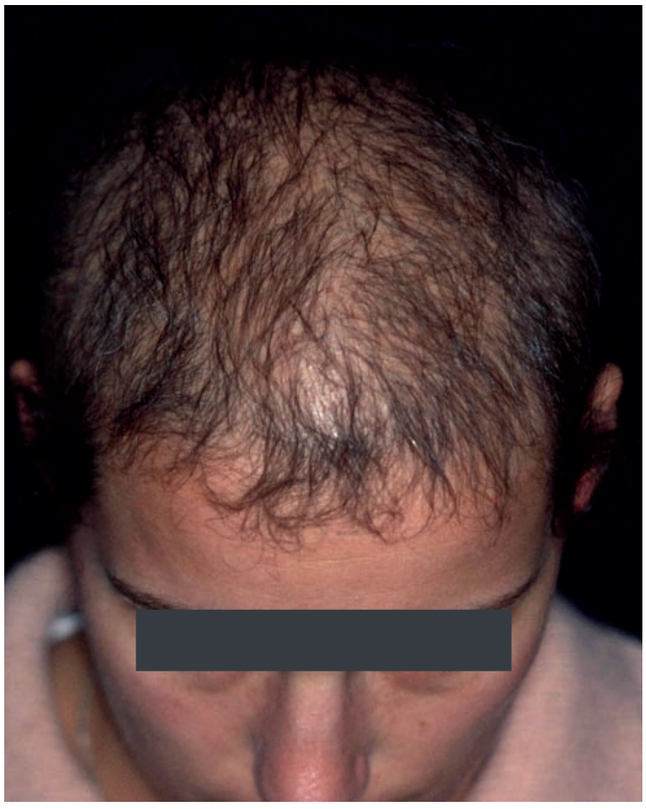


5.23

Figures 5.21, 5.22
Sudden whitening.

Figure 5.23
White hair regrowth in a patient with AA.



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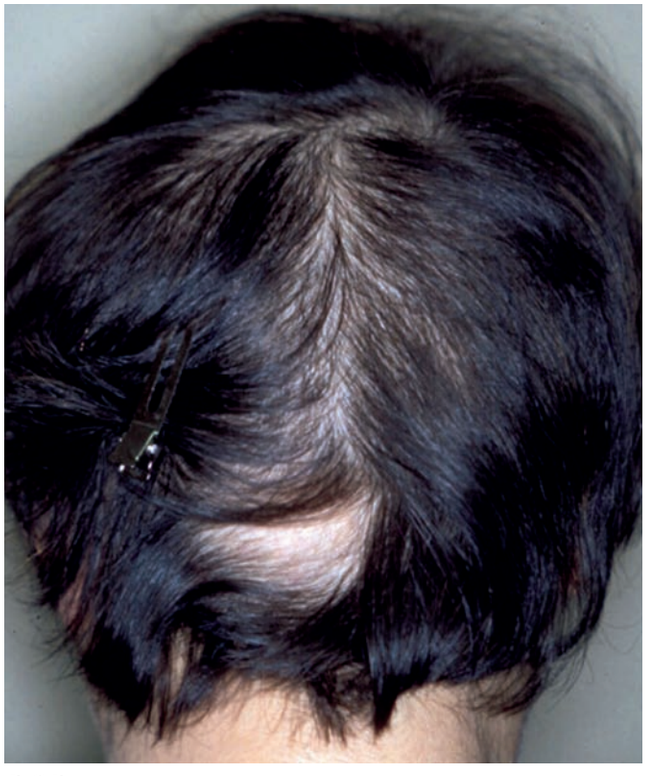

5.25

Figures 5.24, 5.25
AA incognita.

- Alopecia areata incognita: hair thinning is diffuse and typical patches may not be evident (Figures 5.24, 5.25). Differential diagnosis with TE requires pathology.

Associated diseases

- *Nail abnormalities* are common, especially in children. These include:
  - superficial pitting with a geometric pattern (Figure 5.26)
  - twenty nail dystrophy (trachyonychia). The nails are rough due to excessive longitudinal ridging (Figure 5.27).

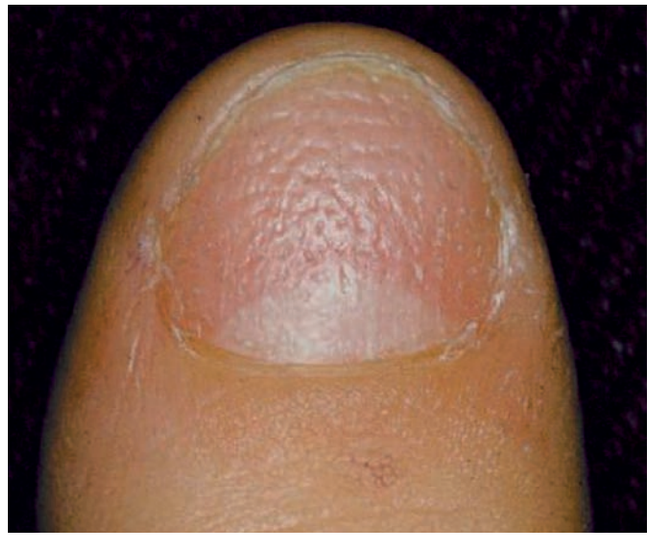

Figure 5.26
Superficial geometric pitting in AA.

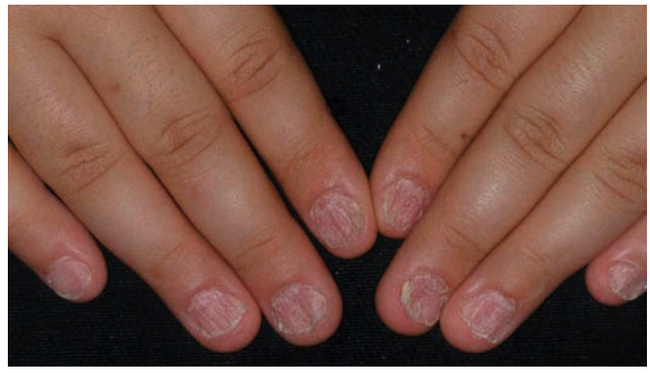

Figure 5.27
Twenty nail dystrophy (TND).

- *Thyroid autoimmune disease.* Clinical or subclinical thyroiditis can be detected in up to 30% of patients with alopecia areata.
- *Celiac disease.* The association is uncommon and diet does not influence the course of AA.
- *Vitiligo.*
- *Down's syndrome.* Prognosis of AA is usually unfavorable.

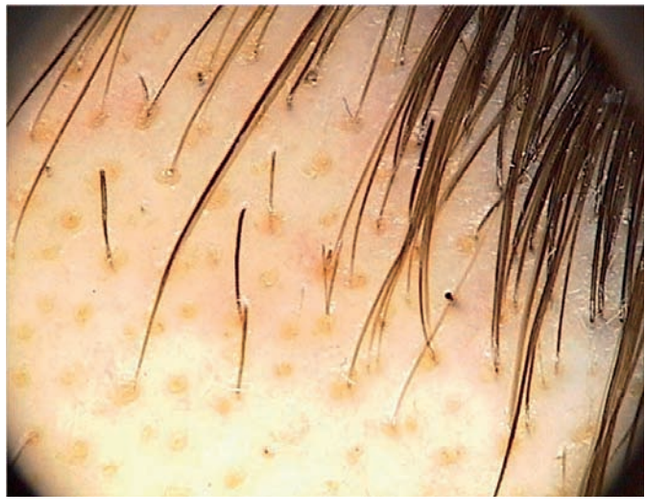

Figure 5.28
Dermoscopy: exclamation point hairs.

Figure 5.29
Broken tip.

Diagnosis

The pull test is useful to establish if the disease is slowly or rapidly progressing. In the latter case tufts of telogen and dystrophic hairs are easily extracted. Microscopic examination shows telogen hair and hair shafts with a broken, frayed proximal end (Figure 5.29).

Dermatoscopic examination of the scalp reveals:

- Exclamation point hairs (Figure 5.28). These are 3 mm long telogen hairs with a pigmented distal broken tip.
- Short pigmented hair with a distal broken tip.
- Twisted hairs broken at the scalp level.
- Peripilar monomorphous yellow dots. These are round or polycyclic in shape, but essentially uniform in color and multiple in number (Figure 5.30).
- The presence of yellow dots can be a clue for the diagnosis of AA in AA incognita (Figure 5.31).

Figure 5.30
Yellow dots in AA.

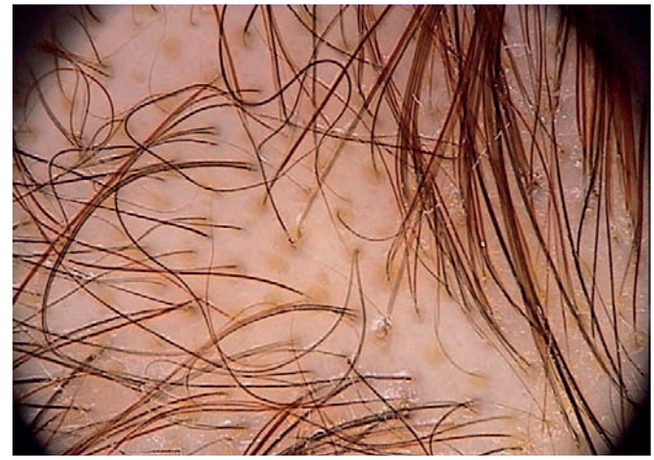

Figure 5.31
Yellow dots in AA incognita.

Table 5.1 Therapeutic strategies for alopecia areata in adults

	Stable	*Active*	*Children*
Single/multiple patches	Intralesional steroids Anthralin Topical immunotherapy	Systemic steroids (pulse) Topical steroids under occlusion	Anthralin Topical immunotherapy
AT/AU	Topical immunotherapy Phototherapy Topical steroids under occlusion		Topical immunotherapy

Treatment (Table 5.1)

- Systemic steroids: High-dose pulse corticosteroid therapy is effective in acute ACM (≅ 60% of cases), but not in ophiasis alopecia (B) or longstanding AT/AU.
  - Methylprednisolone i.v. 500 mg/day for 3 days a month for 3 months (10 mg/kg/day in children)
  - Oral prednisone 300 mg/month (5 mg/kg in children)
  - Oral desametasone 40 mg/month (5 mg/day for 2 consecutive days every week).
- Intralesional steroids: They are effective in localized forms and eyebrows (B). Triamcinolone acetonide should be diluted in saline solution (5–10 mg/ml) and injected within the patch. Maximum dosage should not exceed 20 mg for session. Treatment should be repeated monthly.
- Topical steroids: 25% of patients with longstanding AT/AU respond to treatment with high-potency topical steroids under occlusion (A), but relapses are common. Without occlusion, they are probably only a placebo treatment. Clobetasone propionate 0.05% ointment should be applied under occlusion at night time. Daily dosage should not exceed 2.5 g. Scalp folliculitis is a frequent side effect.
- Topical immunotherapy with DPCP or SADBE (A): Sensitization is obtained utilizing 2% in acetone squaric acid debutylesther or diphenylcyclopropenone under closed patch test for 48 hours. Treatment is performed with weekly application of the allergen diluted in acetone at a concentration able to induce a mild scalp contact dermatitis. This concentration can considerably vary among the different patients and even in the same patient during the treatment period. Hair regrowth is obtained in about 20% of patients with severe AT/AU. Topical immunotherapy is scarcely effective in acute, rapidly expanding disease.
- Phototherapy (B):
  - Narrowband UVB and UVA photochemotherapy are an option for patients with AT/AU. Since presence of hair limits UV penetration, maintenance of hair regrowth is difficult.
  - Topical PUVA (PUVA turban): 0.0001% 8-MOP solution is applied using a soaked cotton towel at 37°C for 20 minutes, followed by UVA irradiation 3–4 times a week, with cumulative UVA dose ranging between 60.9 to 178.2 J/cm^2.
- Topical anthralin (B): 1% anthralin is applied daily for 2 hours. The drug induces mild erythema and irritation of the scalp. This treatment is a good option for children and mild forms of ACM.
- Systemic CyA (D): 5 mg/kg/day.
- Non-effective treatments:
  - Imiquimod.
  - Topical tacrolimus.
  - Extracorporeal photophoresis.
- Cosmetic aids:
  - Tattooing of the eyebrows is a valid cosmetic aid in AT or AU.
  - Wigs.

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6 Androgenetic alopecia

Androgenetic alopecia (AGA) is the most common form of hair loss, affecting up to 80% of men and 50% of women in the course of their life. The disease typically manifests after puberty and is evident by the age of 30. Onset in childhood is rare, but up to 15% of adolescents show signs of the disease.

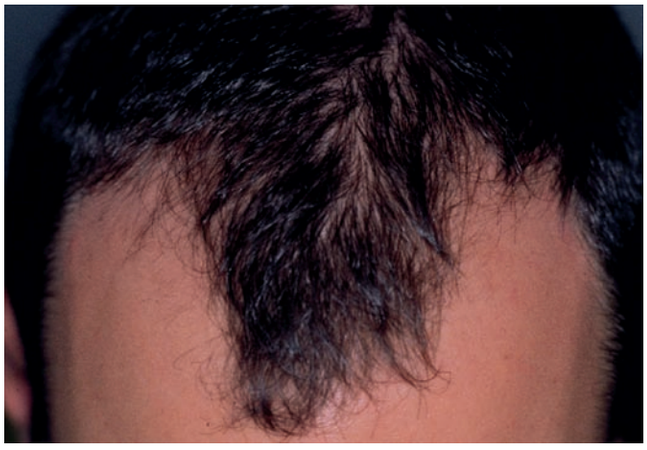
Clinical features

Clinical manifestations of AGA are different in the two sexes (MPHL, male pattern hair loss; FPHL, female pattern hair loss) (Table 6.1).

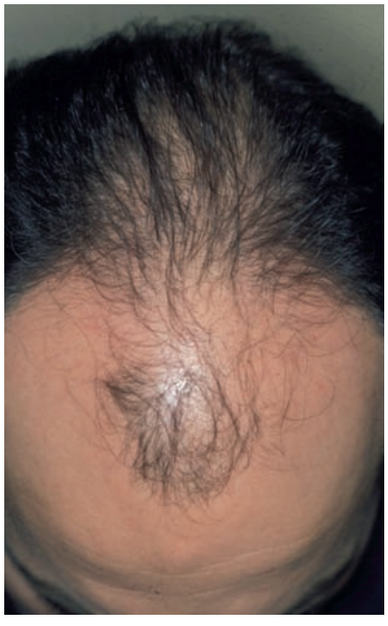
Table 6.1 AGA: clinical features

Men	**Women**
Hamilton–Norwood type	Ludwig type
Ludwig type	Christmas tree pattern
Acquired progressive kinking of the hair	Hamilton type
Whisker hair	

- In most men AGA involves the fronto-temporal area and the vertex, following a pattern that corresponds to the Hamilton–Norwood scale (Figures 6.1–6.5).

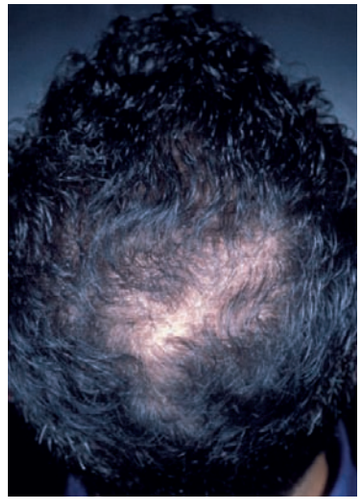


6.1



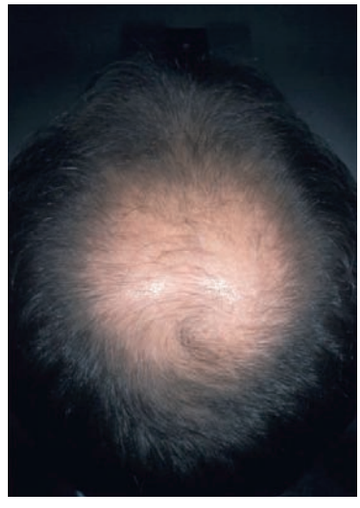
Figures 6.1, 6.2
Frontotemporal involvement in AGA in man.

6.2



Figures 6.3, 6.4
Vertex involvement in male AGA.

6.3



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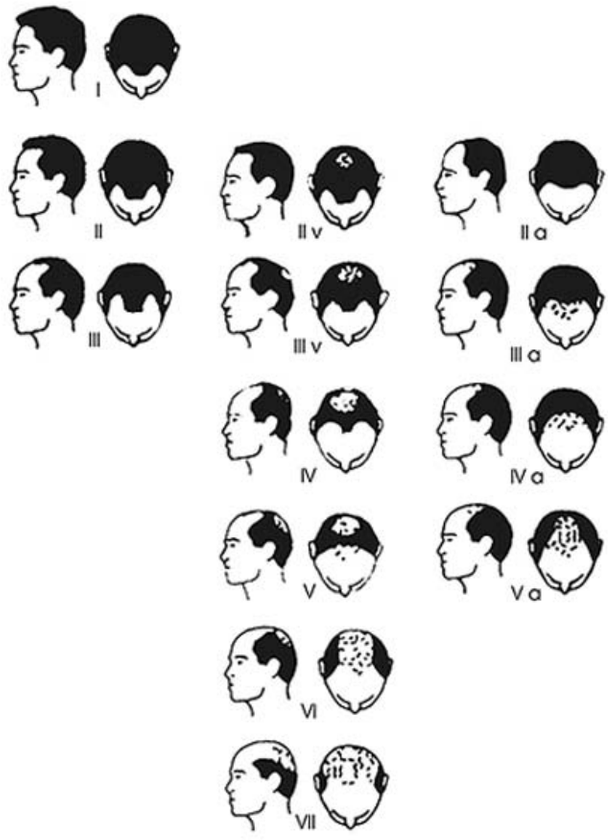
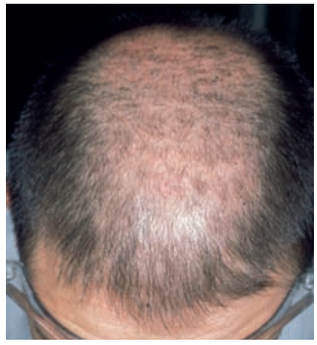


Figure 6.5
Hamilton–Norwood scale.

- Rarely, however, men develop diffuse thinning of the crown with maintenance of the frontal hairline with a pattern that resembles the Ludwig type observed in women (Figures 6.6, 6.7).



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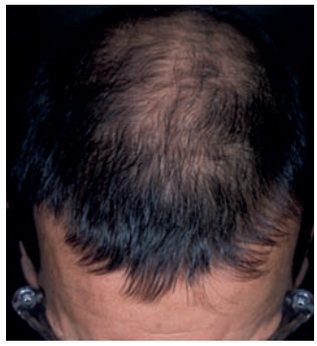
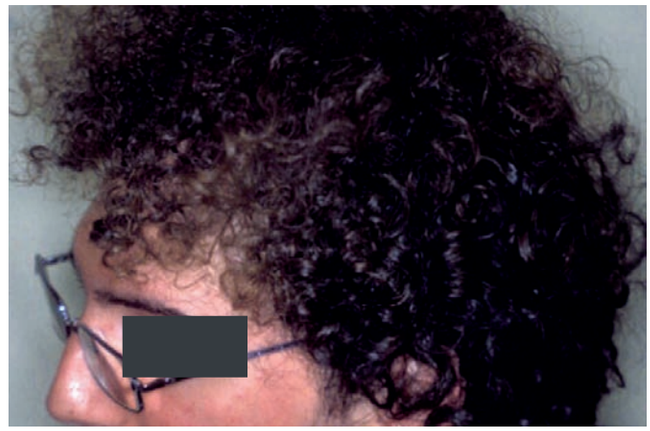


6.7

Figures 6.6, 6.7
AGA with female pattern in man.

- In acquired progressive kinking of the hair (APKH), kinking and lightening of the hair of the androgen-dependent areas precede the development of evident thinning. Kinking may involve the peri-auricular areas where terminal hair become short and curling, resembling beard hair (whisker hair) (Figures 6.8–6.10).



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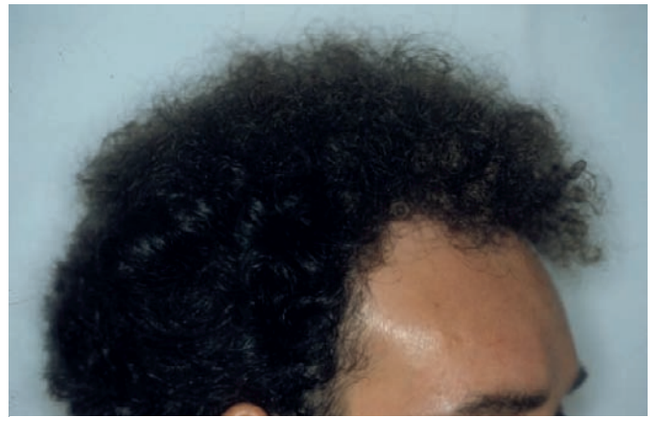

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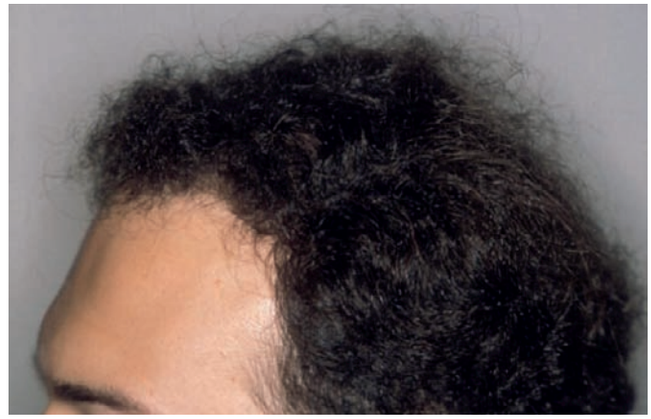

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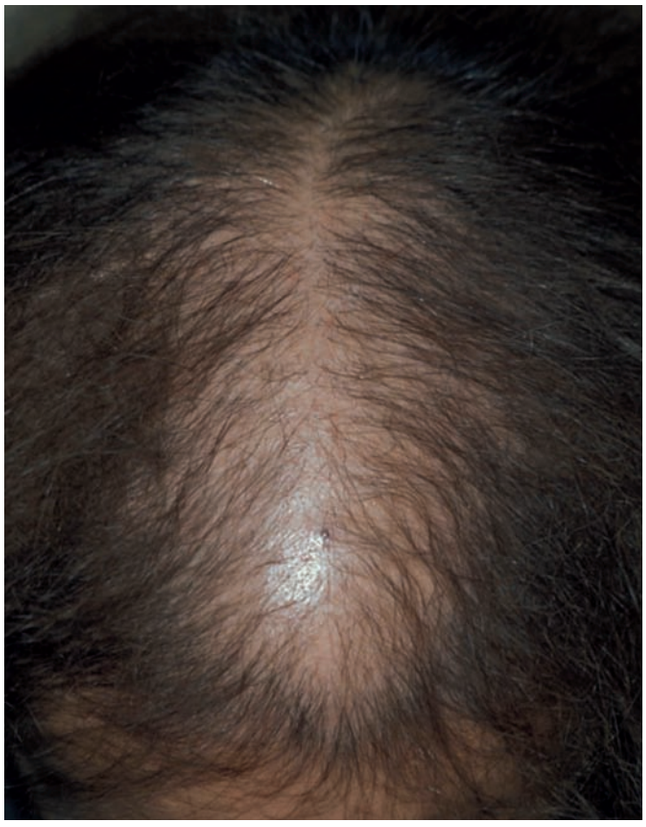
Figures 6.8–6.10
APKH.



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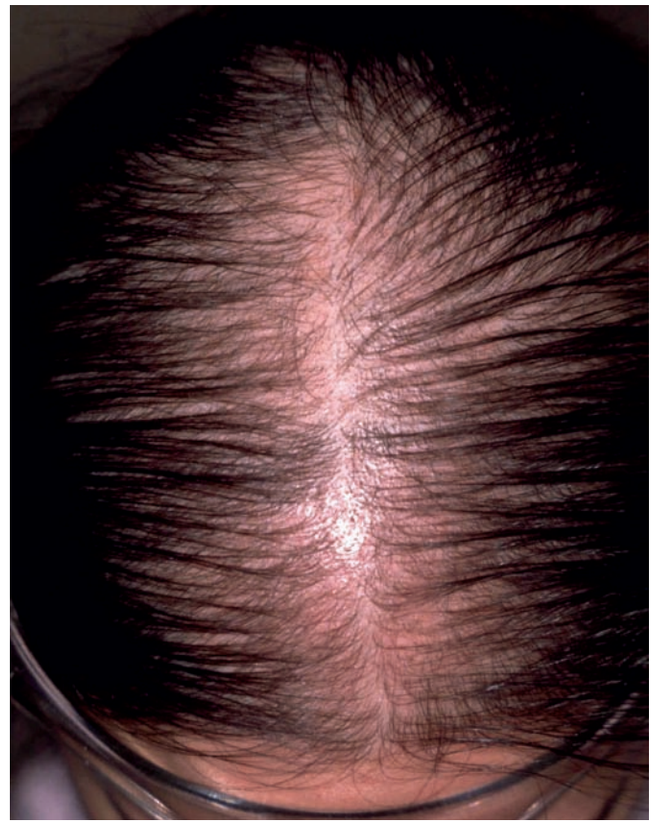


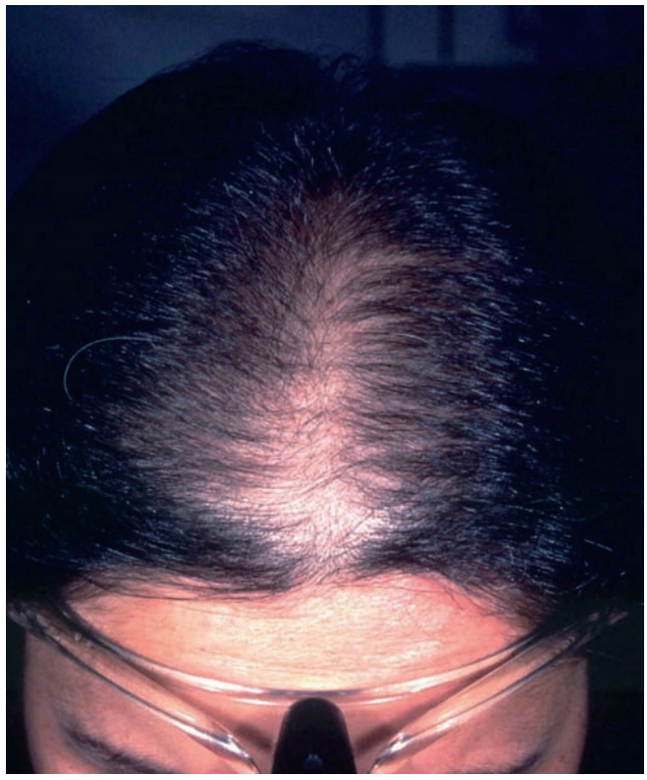
6.12

Figures 6.11, 6.12
AGA in female with a Ludwig pattern.

In women three different patterns can be observed:

- Diffuse thinning of the crown region with maintenance of the frontal hairline (Ludwig type) (Figures 6.11–6.13).
- Thinning and widening of the central part of the scalp with breach of frontal hairline (Christmas tree pattern) (Figures 6.14–6.17).
- Thinning associated with bitemporal recession (Hamilton type) (Figures 6.18, 6.19).

Figure 6.13
Ludwig scale.



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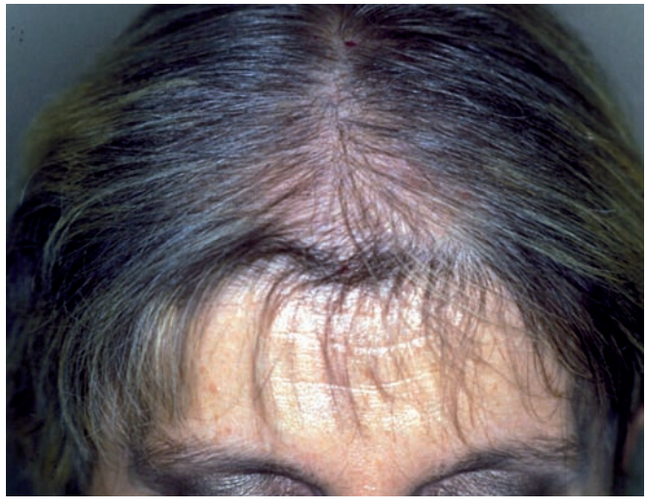

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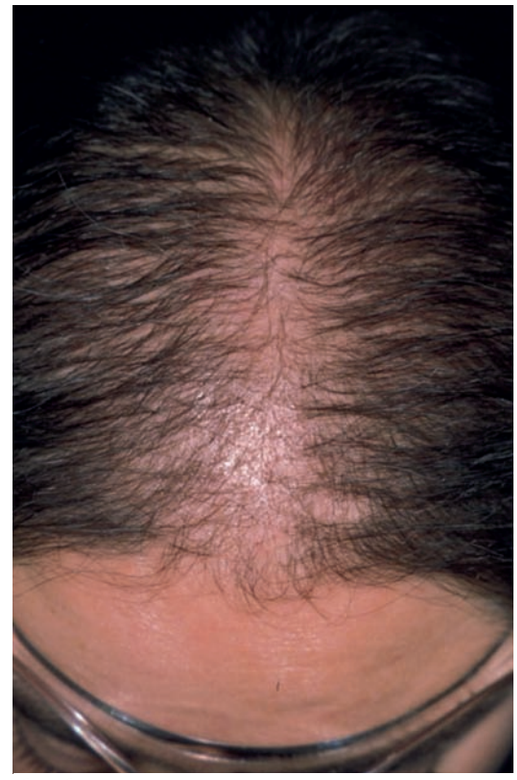

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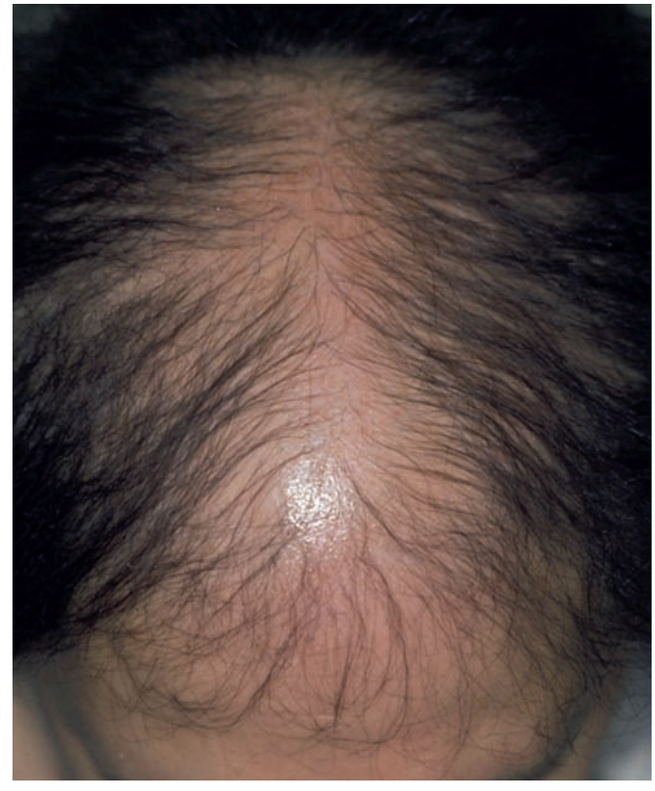
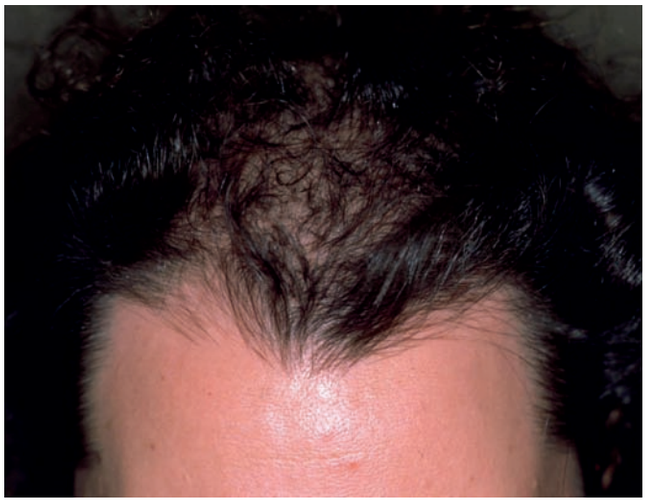


6.17

Figures 6.14–6.17
Christmas tree pattern in female AGA.



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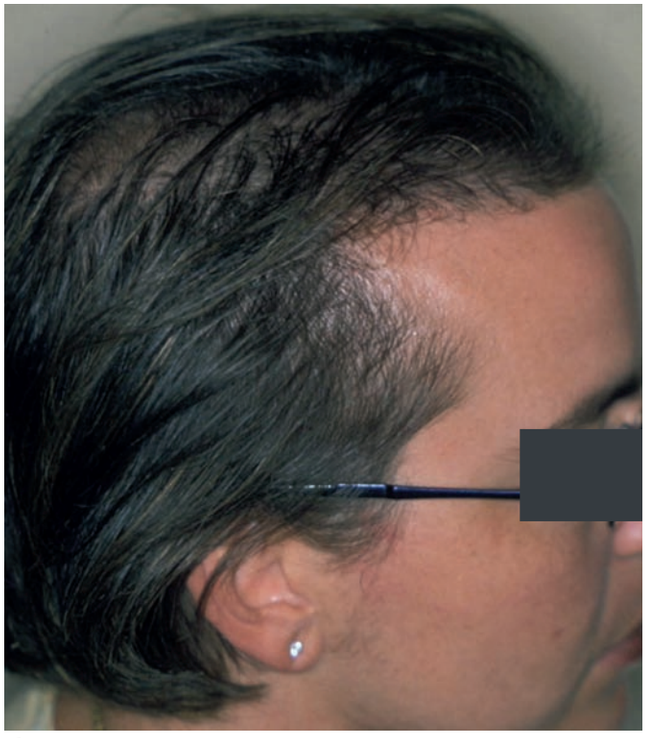


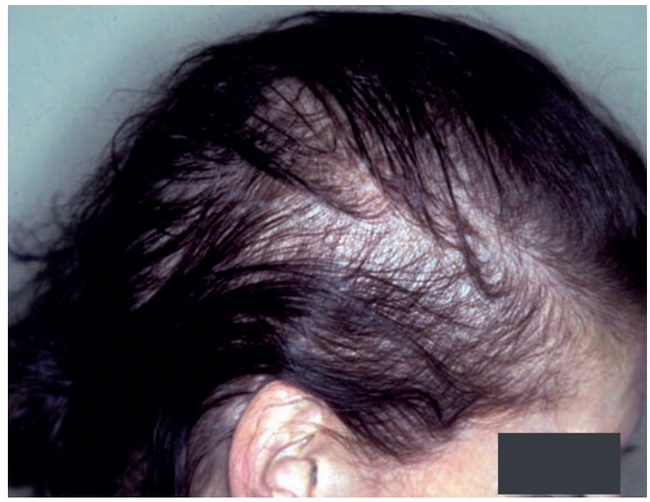
6.19

Figures 6.18, 6.19
AGA with Hamilton pattern in a female.

Involvement of the parietal and occipital scalp with diffuse alopecia is not rare (Figures 6.20–6.22).

Hair thinning in AGA results from four associated different factors.

1. Hair miniaturization: reduction in the hair shaft diameter, length and pigmentation (Figures 6.23–6.25). This is a very early sign that can be better



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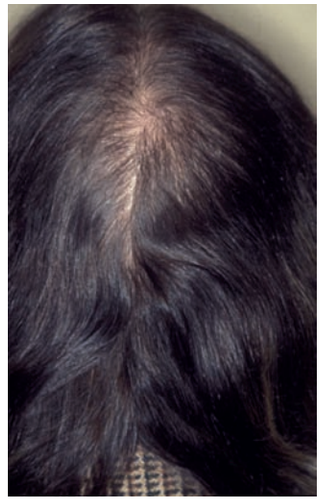

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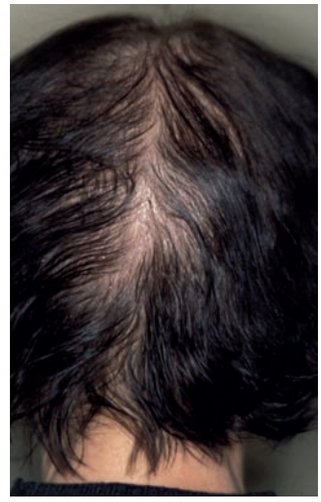

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Figures 6.20–6.22
AGA in female with involvement of parietal and occipital scalp.

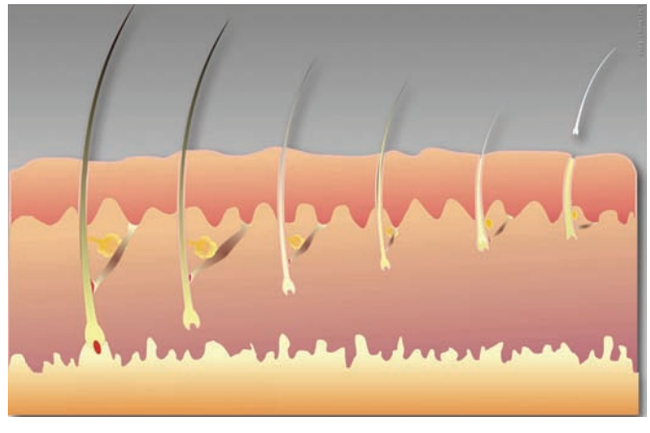
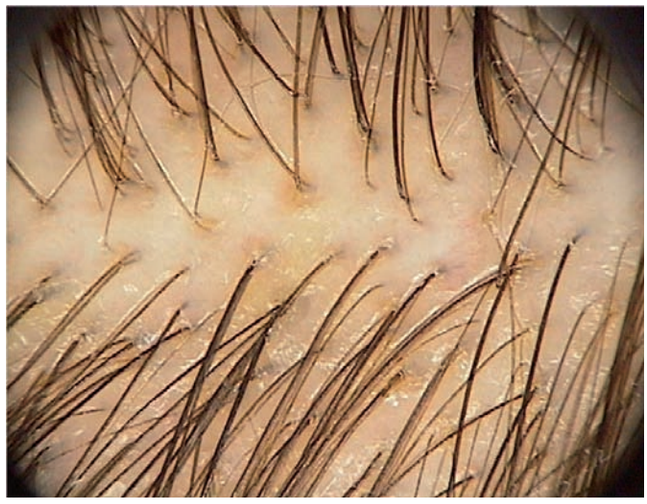


Figure 6.23
Alteration of the hair cycle in AGA.



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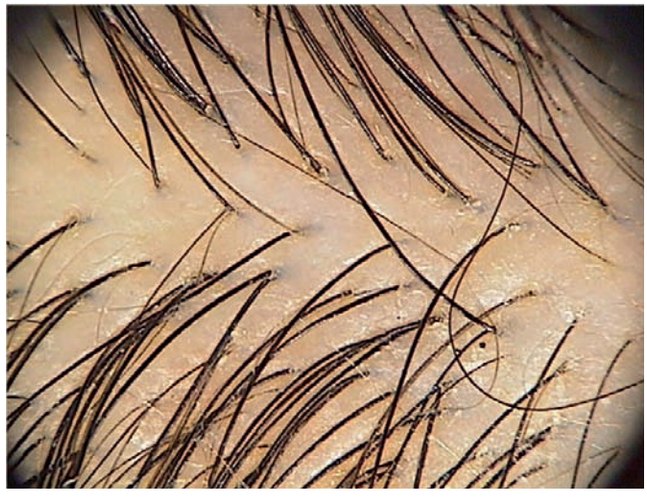

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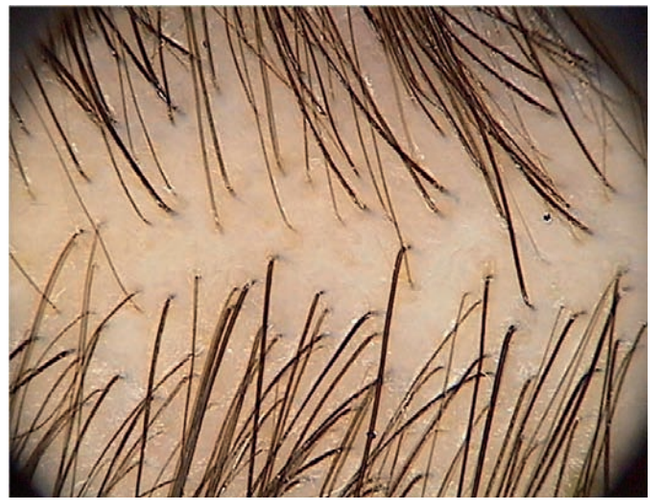
Figures 6.24, 6.25

AGA: scalp dermoscopy shows various degrees of hair diameter diversity.



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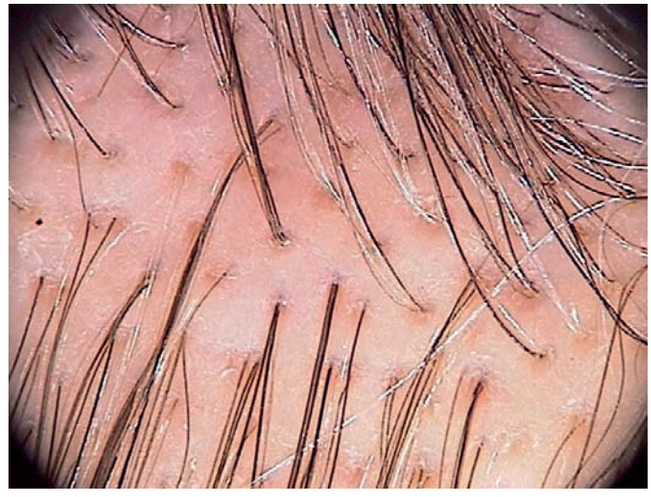


6.27

Figures 6.26, 6.27

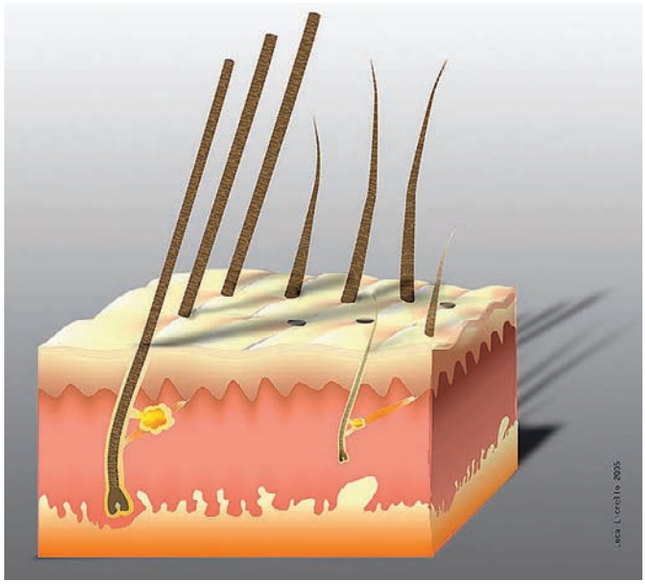
Peripilar signs at scalp dermoscopy in AGA.

detected using scalp dermoscopy, which shows hair diameter diversity (more than 20%) in the affected area. Peripilar signs may also be observed (Figures 6.26, 6.27).

2. Empty follicle phenomenon: some follicles are empty due to delayed anagen re-entry after shedding of telogen hair (Figures 6.28, 6.29). This feature is more evident in females and produces 1–3 mm areas of complete hair loss.
3. Increased telogen shedding: patients often complain of increased hair loss that is due to the fact that miniaturized follicles have short anagen phase and therefore the percentage of telogen hair is increased in scalp regions affected by the disease. Sometimes, however, an episode of telogen effluvium precipitates or worsens AGA.
4. A true loss of hair follicles: fibrosis may replace hair follicles in some cases.

Associated diseases

- In premenopausal women, FPHL may be a cutaneous sign of hyperandrogenism, together with hirsutism and acne (Figure 6.30). Polycystic ovarian syndrome (PCOS) is found in about 30% of women with FPHL.



6.28

6.29

Figures 6.28, 6.29
Empty follicles in AGA.

- AGA is frequently associated with psychological disturbances including anxiety and depression, particularly in patients seeking treatment.
- Several studies have correlated AGA with an increased frequency of coronary artery disease (CAD).

Diagnosis

Diagnosis of androgenetic alopecia is usually simple (Table 6.2).

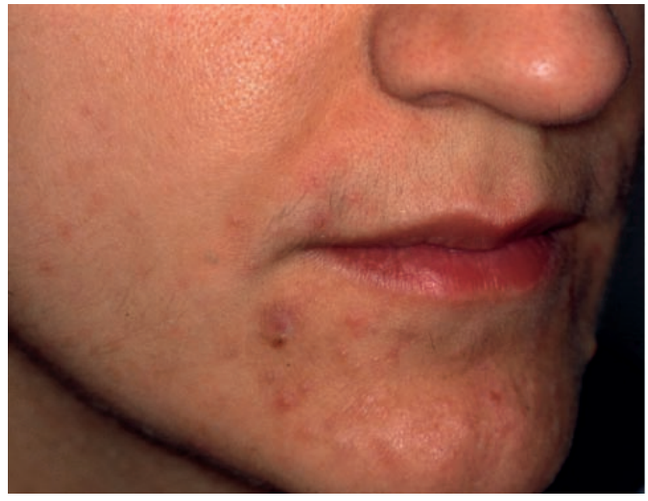


Figure 6.30
Cutaneous signs of hyperandrogenism may be associated with AGA in females.

Table 6.2 Diagnostic features of AGA

Variation in the hair shaft diameter
Patterned distribution
Positive pull test with extraction of short telogen hair in the affected area

In men, clinical examination typically reveals hair thinning in the frontotemporal and vertex regions (Figure 6.31).

In women, comparing the density of hair in the crown region with that of the occipital region (Figures 6.32,

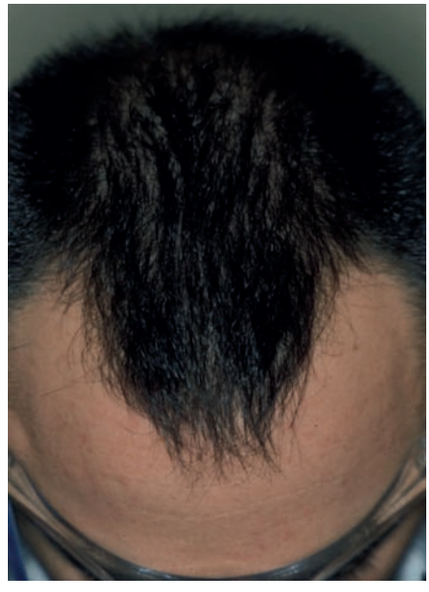
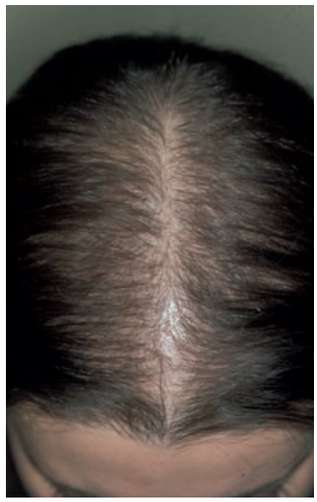


Figure 6.31
AGA in man.



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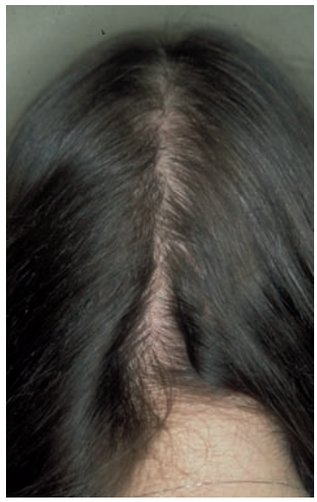


6.33

Figures 6.32, 6.33

Comparison of hair density of the crown region with that of the occiptal region for diagnosing AGA in females.

6.33) can be very helpful to differentiate mild forms of FPHL from diffuse forms of hair loss.

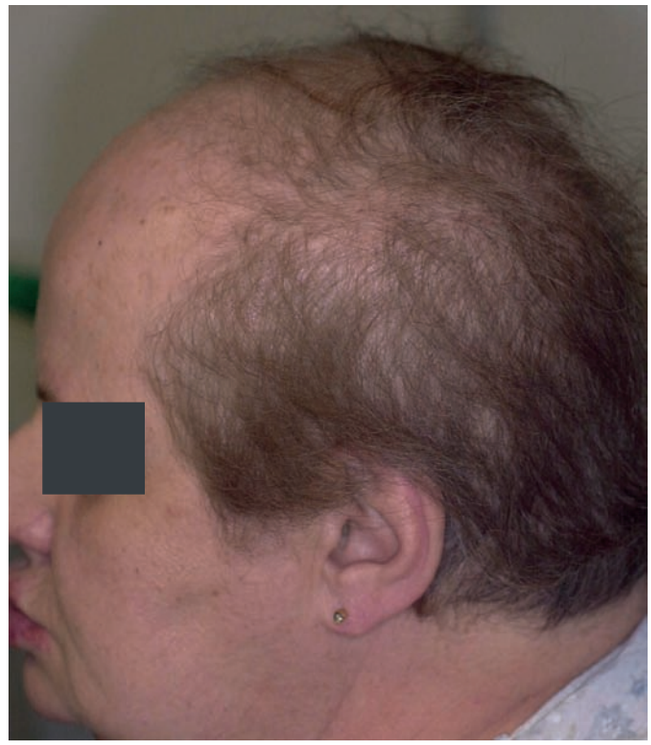
- Variation in the hair shaft diameter is typical of AGA and is an important feature for early diagnosis of the disease. This can be better visualized using scalp dermoscopy.
- Pull test may be negative or reveal an increased telogen shedding (Table 6.3). The presence of telogen hairs, shorter than 3 cm, representing the telogen phase of miniaturized follicles, is diagnostic for AGA.

Laboratory investigations are not necessary, except for female patients with signs of hyperandrogenism (Table 6.4, Figures 6.34, 6.35).

Table 6.3 Interpretation of pull test results in patients with AGA

	Slow progressive	*Stable*	*Progressive (look for causes of associated TE)*
Crown region	–	+	+
Occipital/parietal region	–	–	+

–, negative; +, positive.



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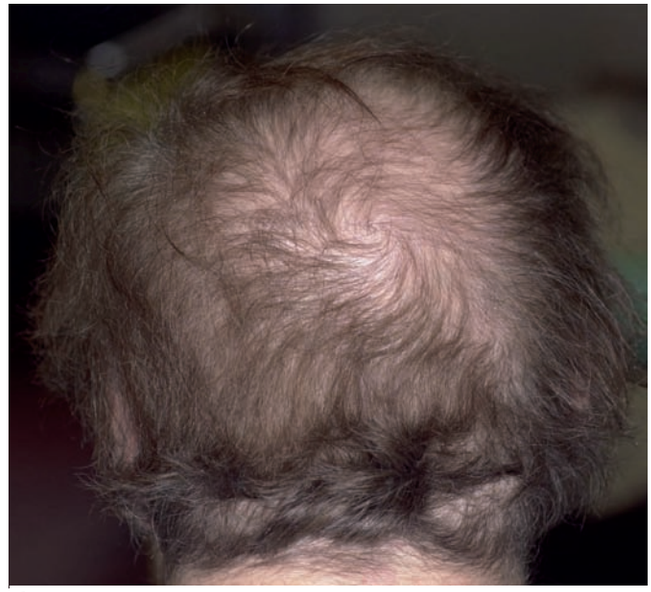

6.35

Figures 6.34, 6.35

Severe Hamilton type AGA and other signs of hyperandrogenism in a woman with adrenal tumor.

Outcome

AGA is a progressive disease that, if left untreated, tends to worsen with time (Figure 6.36) and eventually leading

Table 6.4 Laboratory investigations suggested for women with signs of hyperandrogenism

FSH	17OH-progesterone
LH	Prolactine
Testosterone	SHBG
DHEA	Cortisol
DHEA-S	Thyroid function
Androstenedione	Ovary scan

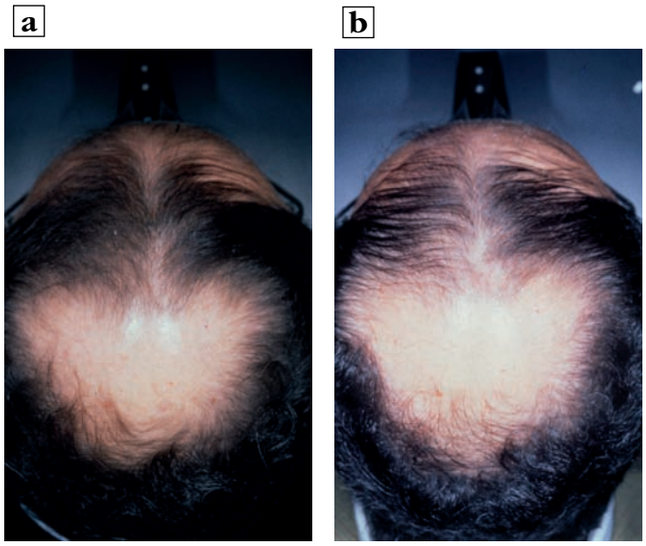

Figure 6.36a,b
Progression of AGA in 13 years.

to total baldness of the top of the scalp (Figure 6.37). Progression of the disease can be slow or very fast, especially in patients with a strong family history or in females with hormonal disturbances. Rapid progression of AGA is also observed after any pathological condition that induces telogen effluvium.

Treatment

Patients with AGA should be instructed about the importance of avoiding factors known to induce telogen effluvium that may accelerate progression of the disease (Table 6.5).

If used correctly, available medical treatments for AGA arrest progression of the disease and reverse miniaturization in most cases. Advanced stages (Hamilton IV, Ludwig III) are unlikely to benefit from medical treatment.

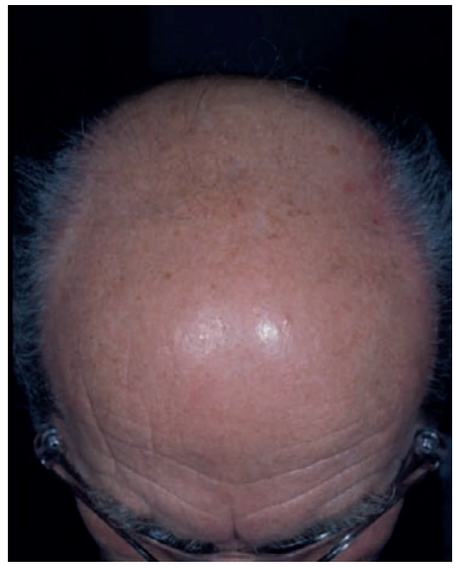

Figure 6.37
Severe AGA in a man: total baldness of the top of the scalp.

Table 6.5 AGA: To be avoided

Sun exposure (wear a hat)
Sun bed tanning
Smoking
Restrictive diets
Drug inducing hair loss (see page 61)

Treatments depend on the sex of the patient, his/her attitude to oral or topical agents and severity of AGA.

Men

- First choice: oral finasteride 1 mg/daily (A). The drug prevents progression of the disease in 99% of patients and produces clinical improvement in 66% after 2 years of continuous treatment. It is effective both in the fronto-temporal region and in the vertex Finasteride acts by inhibiting type II 5x reductase, the enzyme that converts testosterone into dyhydrotestosterone, the hormone responsible for hair follicle miniaturization (Figure 6.38). Side effects are observed in less than 2% of patients (and in a slightly smaller percentage of patients receiving placebo) and include decreased libido, erectile dysfunction and diminished ejaculate volume. All these problems are mild and promptly resolve after discontinuation of the drug.
- Second choice: 5% topical minoxidil 1 ml bid (A). Forty to 60% of patients show visible improvement after 2 years of treatment. The major side effects of minoxidil treatment are scalp irritation and allergic

contact dermatitis, which precludes further use of the drug.
- Although the finasteride–minoxidil association has not been widely tested in humans, results on animal models suggest that association of the two treatments leads to better results than one treatment regimen alone.

Treatments for AGA should be continued for at least 1 year before assessing any effectiveness, even though the first signs of improvement can already appear after 4–6 months of therapy. Regular drug use is mandatory for any efficacy, and, if effective, should be prolonged. Minoxidil interruption produces an acute telogen effluvium that starts 3–4 months after interruption. The occurrence of hair loss is not prevented by concomitant finasteride treatment. Interruption of finasteride is also followed by gradual hair loss with return to the pretreatment status 1 year after interruption of treatment.

Discuss with the patient the possibility of surgical treatment with mini- and micro-grafts and scalp reduction.

Finasteride and/or minoxidil should in any case be prescribed as an adjuvant treatment before and after surgery.

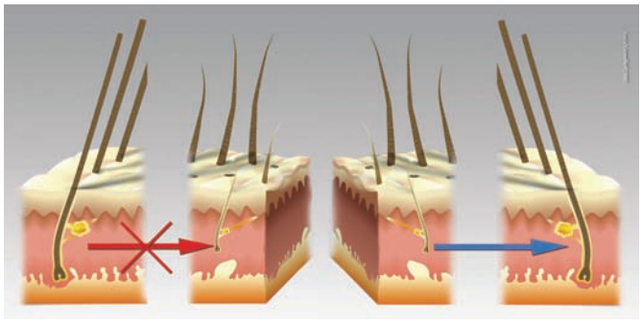

Figure 6.38

Treatment with finasteride prevents hair follicle miniaturization and transforms miniaturized follicles into terminal ones.

Women

- Two percent topical minoxidil 1 ml b.i.d. (A) is very effective in most women with evident clinical improvement in up to 80% of cases. Side effects include scalp irritation, contact dermatitis and reversible hypertrichosis, which is most commonly caused by local contamination. Advise the patient not to apply minoxidil before going to bed, thus avoiding contamination through the pillow. Five percent topical minoxidil once a day can be an option for women who complain that the lotion impairs their hair styling. It is not advisable to prescribe 5% topical minoxidil twice a day in women, because diffuse reversible hypertrichosis may occur.
- Oral finasteride (D) should only be prescribed in post-menopausal women or in women willing to use oral contraceptives. The drug is contraindicated in women that may become pregnant since it can block the development of the genitalia of the male fetus. Although a double blind study did not show any efficacy of finasteride in post-menopausal women, recent reports indicate that the drug may be effective in women, even in the absence of associated hyperandrogenism. Optimal dosages and modality of administration are still not established.
- Cyproterone acetate 25–50 mg/day (A) administered in the first 10 days of the menstrual cycle can be useful in women with biochemical hyperandrogenism. Contemporary administration of estrogen (ethynil estradiol or oral contraceptives) is mandatory. A controlled study comparing 2% topical minoxidil and 50 mg cyproterone acetate + Diane showed that minoxidil was more effective in women with low body mass index (BMI) and no signs of hyperandrogenism, whereas cyproterone acetate was more effective in women with biochemical hyperandrogenism.
- Evaluate the possibility of hair transplantation. Hair transplantation in women is often complicated by two factors: (1) hair thinning is often diffuse to the parietal and occipital regions with absence of a good donor area; (2) hair transplantation often results in effluvium of the pre-existing hair in the recipient area, which can produce a significant temporary worsening of the hair thinning.
- Cosmetic improvement can be obtained using topical preparations in powder or sprays that bind electrostatically to the hair and temporarily increase its thickness and color.

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7 Telogen effluvium

Introduction

Telogen effluvium (TE) describes diffuse loss of telogen hair. Most cases of TE are probably subclinical and epidemiological data on this condition are lacking.

Five types of telogen effluvium can be distinguished depending on the hair cycle abnormality (see Table 7.1):

1. Premature termination of anagen. This is the most common type and is caused by noxae that interrupt the mitotic activity of the hair matrix cells and induce telogen entry. These include systemic diseases, drugs, stress, weight loss, UV exposure, iron deficiency, cigarette smoking and inflammatory scalp disorders. The mechanism by which these different factors cause hair loss is not always clear and involves decreased vascular supply, increased free radical production, cytokine release and toxic effect.
2. Synchronization of the hair cycle due to excessive prolongation of anagen. It develops when the condition that prolongs the anagen duration subsides and synchronized follicles simultaneously enter telogen. This type of TE typically occurs post partum (2–3 months after delivery), after interruption of the contraceptive pill and after discontinuation of topical minoxidil.
3. Synchronization of the hair cycle due to reduced anagen duration. When duration of anagen is reduced the hair cycle shortens and a large number of follicles enter telogen at the same time. It has been established that for every 50% reduction in duration of anagen there is a corresponding doubling in telogen follicles. This type of TE is typically seen in the androgen-dependent scalp of patients with AGA.
4. Immediate telogen release (premature teloptosis) in subjects with synchronized hair cycle. This is a consequence of premature detachment of the club hair from the follicle with shortening of the normal telogen phase. This type of telogen effluvium becomes clinically evident only when the number of telogen follicles is abnormally increased due to hair cycle synchronization.
5. Delayed telogen release. Results from prolonged retention of club hair which remains anchored to the follicle longer than normal. This phenomenon is responsible for molting of mammals and may possibly also occur in humans suffering from jet lag. Delayed teloptosis can occur in keratinization disorders such as scalp psoriasis.

Most commonly telogen effluvium results from simultaneous passage of a large number of follicles from the anagen to the telogen phase (type 1). Hair loss appears after about 3 months, corresponding to the duration of telogen. Most club hairs are in fact retained within the follicle during telogen to be shed when the follicle produces a new anagen hair. Types 2–5 of TE only occur when the hair follicles have synchronized the hair cycle and the number of telogen follicles in the scalp is higher than normal. The most common cause of synchronization of the hair cycle is androgenetic alopecia and patients with this condition have a high susceptibility to types 2–5 TE.

Clinical features

Hair density may be normal, slightly (Figure 7.1) or severely (Figures 7.2, 7.3) decreased. Hair thinning is usually more evident on the top of the scalp due to the frequent association between TE and AGA. Diffuse hair thinning is uncommon.

Figure 7.1
Acute telogen effluvium.

Table 7.1 Telogen effluvium

Type	*Course*	*Delay*	*Causes*	*Clues*
1. Premature termination of anagen	A/C	2–3 months	Drugs (see page 63) Systemic diseases Acrodermatitis entheropathica Anesthesia Brucellosis Dermatomyositis Diabetes Fever Hemorrhages Hemodialysis Hepatic failure Hypo/hyperthyroidism Hypoparathyroidism Hypoprotidemia Iron deficiency Liver disorders Malaria Malnutrition/malabsorption Meningitis Pancreatitis Parenteral nutrition Pneumonia Pernicious anemia Renal failure Secondary syphilis Seizures Systemic lupus erythematosus Surgery Typhoid fever Tuberculosis Ulcerative colitis Virosis Inflammatory scalp disorders Contact dermatitis Psoriasis Seborrheic dermatitis Smoking Stress UV exposure Weight loss	Club telogen hair
2. Synchronization of the hair cycle due to excessive prolongation of anagen	A	2–3 months	Post-partum Interruption of contraceptive pill Interruption of topical minoxidil	Club telogen hair
3. Synchronization of the hair cycle due to reduced anagen duration	C		Androgenetic alopecia Short anagen syndrome	Short telogen hair
4. Immediate telogen release	A		Drugs stimulating hair regrowth Keratolytic agents Retinoids	Telogen hair with epithelial sac
5. Delayed telogen release	A		Molting Jet lag Scalp psoriasis	Club telogen hair

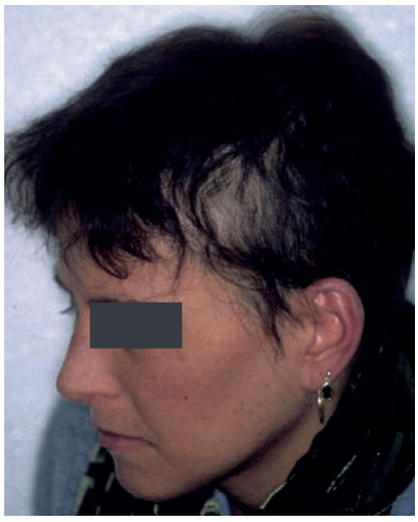


Figure 7.2
Acute telogen effluvium.

Figure 7.3
Chronic telogen effluvium.

Fifteen to 30% of patients with TE, especially women, complain of scalp paresthesia or pain (trichodynia). This may be exacerbated by combing. Trichodynia is not related to severity or prognosis of TE and may be caused by neuropeptide release.

Diagnosis

Diagnosis is based on the history and the results of the pull test. It does not require the evidence of alopecia and most patients may have a full head of hair.

- The diagnosis of TE can be made when the daily hair shedding is higher than 100. In most cases of acute TE, more than 200–300 hairs are shed daily.
- The patient usually can remember quite precisely when the increased hair shedding had started.

Figure 7.4
Dermoscopy of telogen effluvium.

- The pull test is positive with extraction of telogen hairs (often more than 10 hairs, even 1 day after shampooing). The trichogram shows a reduced anagen to telogen ratio (% of telogen >20%).
- Scalp dermoscopy shows a large number of short tip-pointed regrowing hair in the absence of hair diameter variability (Figure 7.4)
- Laboratory tests: exclude iron deficiency, thyroid diseases and autoimmune diseases.

Prognosis

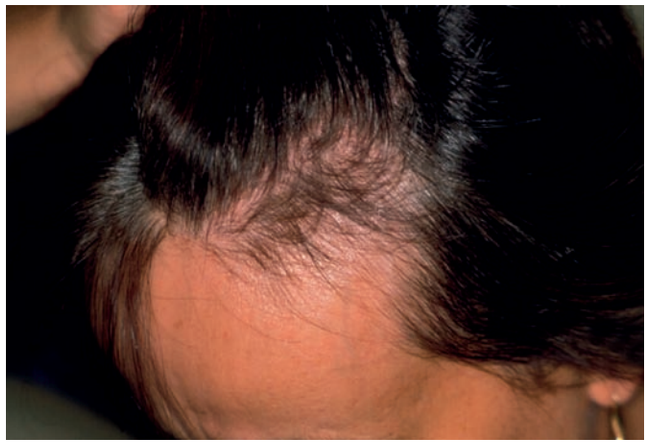
Prognosis depends on the cause and presence of subclinical or established androgenetic alopecia. Be aware that acute TE often unmasks androgenetic alopecia in predisposed subjects.

Treatment

- Removal of the cause
- Nutriceuticals containing amino acids and iron (E).

Chronic telogen effluvium

Chronic telogen effluvium (CTE) is characterized by increased hair shedding lasting for more than 6 months. The condition mostly affects middle-aged women and is idiopathic. The daily hair shedding is usually less important than in TE, but the patients are very distressed and refer that their hair volume has considerably decreased.



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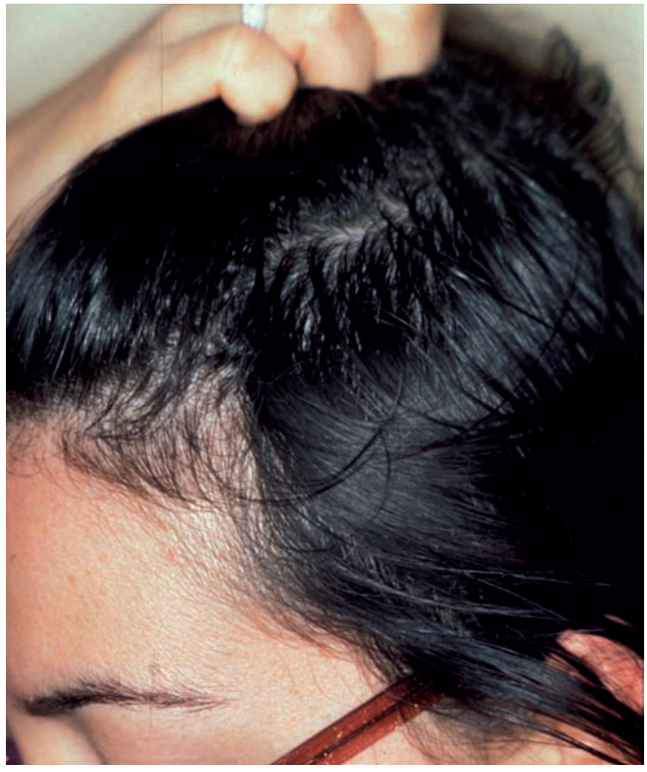

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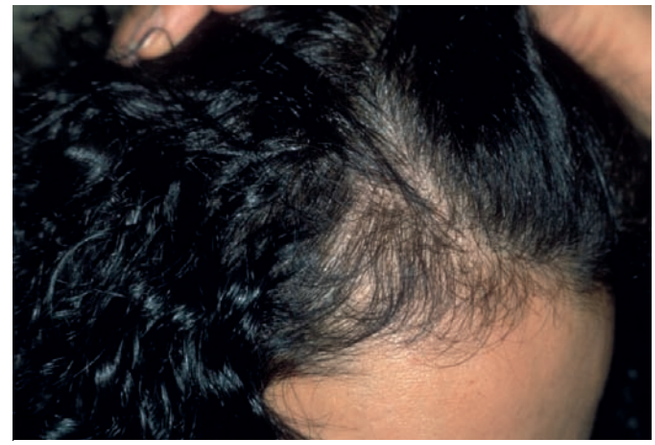

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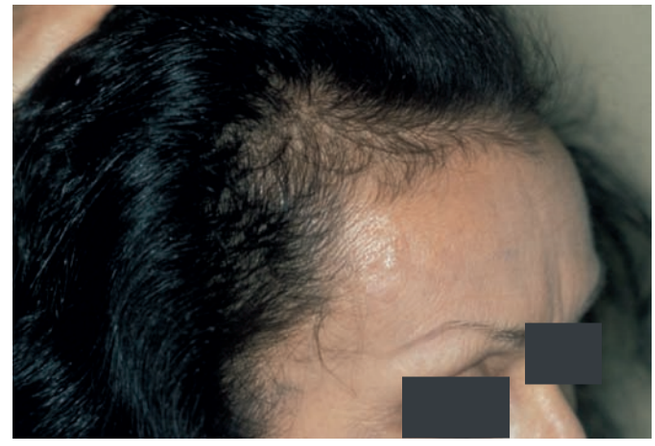

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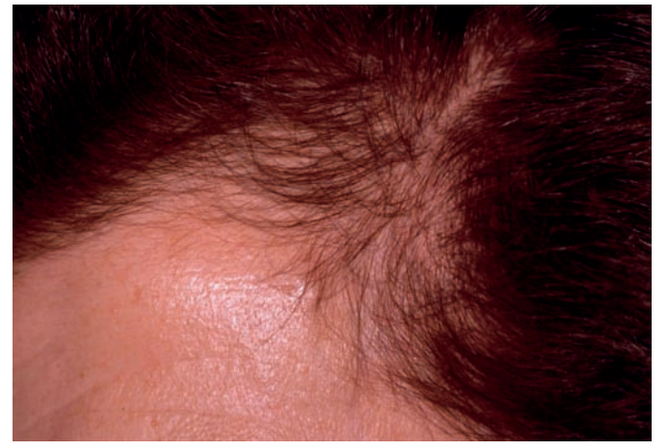


7.9

Figures 7.5–7.9

Chronic telogen effluvium: thinning is more marked on the temporal area.

The clinical examination usually shows a normal hair density. Typically there is a thinning of the temporal region where the normal hair has been replaced by 3–6 cm hair (Figures 7.5–7.9).

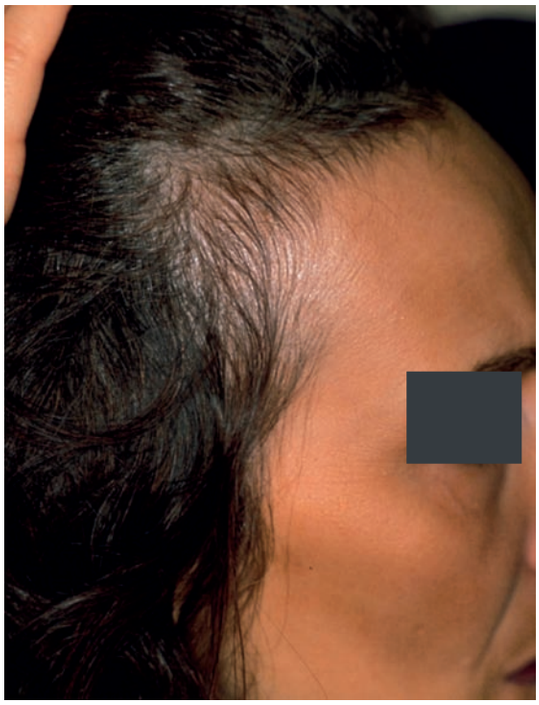
The fronto-temporal recession tends to worsen with time (Figures 7.10, 7.11)

Trichodynia may be a major complaint. CTE is usually not associated with signs of androgenetic alopecia.

Diagnosis

- Clinical: bitemporal hair thinning with presence of hair shorter than those of the surrounding scalp.
- Pull test: negative/positive.

Values of hair shedding are usually between 80–150 hairs daily.



7.10

7.11

Figures 7.10, 7.11
Worsening of the fronto-temporal recession.

Prognosis

The disease typically runs a chronic course but does not produce clinically evident hair thinning.

Treatment

- Systemic steroids (E): triamcinolone acetonide 1 mg/kg/month for 2–3 months
- Topical steroids (E)
- Topical minoxidil 2% (E).

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8 Drug-induced hair abnormalities

Drugs may induce hair loss, stimulate hair growth or, more rarely, induce changes in hair shape and color (Table 8.1).

Table 8.1 Effects of drugs on hair

Reversible hair loss
Permanent alopecia
Excessive hair growth
Changes in hair color
Changes in hair shape/texture

Drug-induced hair loss is usually completely reversible and is, in most cases, a consequence of a toxic effect of the drug on the hair matrix. Depending on the type of drug, dosage and patient susceptibility, hair loss presents as telogen effluvium, anagen effluvium or both.

Telogen effluvium is also commonly observed after discontinuation of drugs that prolong anagen such as topical minoxidil and oral contraceptives.

Although a large number of drugs have been occasionally reported to produce hair loss, only for a few drugs has the relation between drug intake and hair loss been proven (Table 8.2).

Reversible hair loss

Antineoplastic agents (Figure 8.1a,b)

Hair loss is the most common cutaneous side effect of antineoplastics. Hair loss is more frequent and severe in patients receiving combination chemotherapy than in those treated with a single drug.

In the majority of patients hair loss starts after the first or the second cycle of administration of chemotherapy and therefore 1–8 weeks after the start of treatment.

Other common effects of drug toxicity on hair formation are hair depigmentation and Pohl-Pinkus marks that consist of constrictions of the hair shaft.

In rare cases, hair regrowth is not complete and a permanent hair thinning persists (Figure 8.2).

Antipsychotic, antiepileptic and antianxiety agents

Lithium causes hair loss in up to 20% of long-term users. This may also be a consequence of lithium-induced

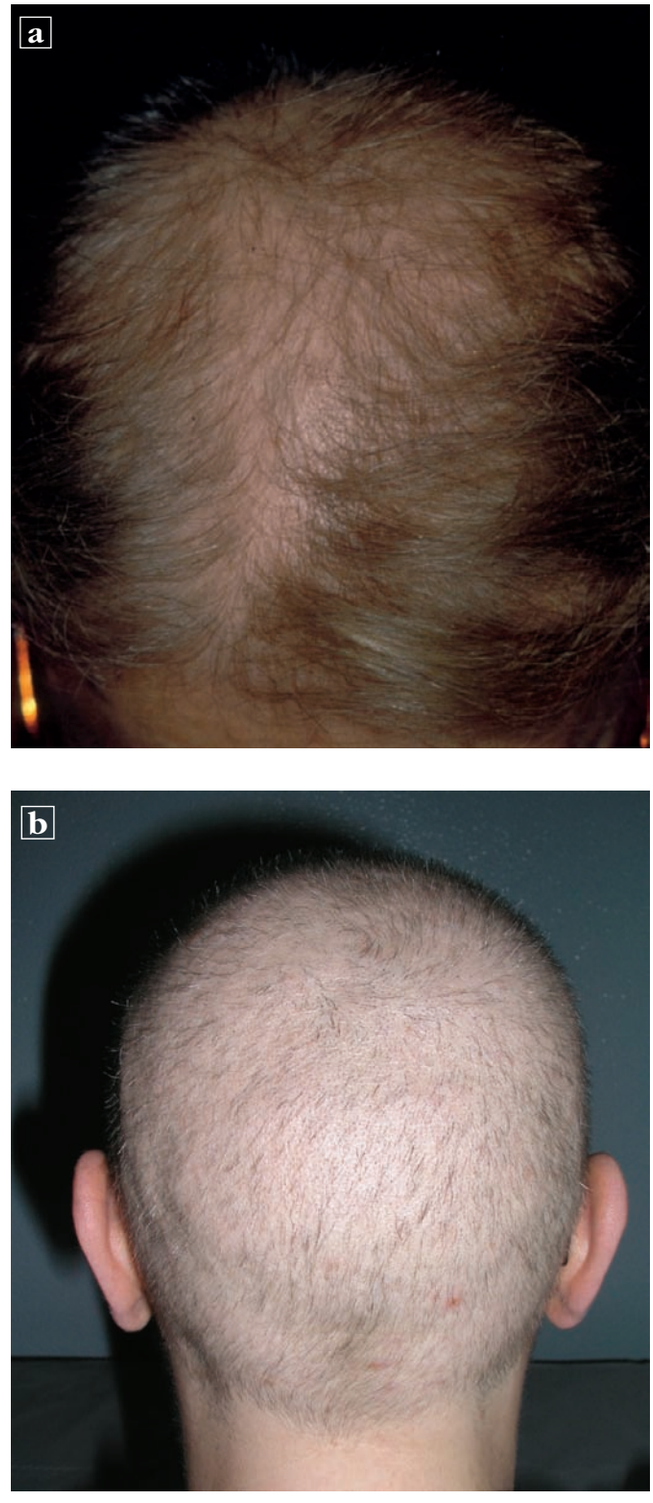


Figure 8.1a,b
Severe alopecia after chemotherapy.

hypothyroidism. Hair straightening has also been associated with lithium intake.

Valproic acid and divalproex frequently cause dose-dependent hair loss which may affect up to 30% of patients taking high dosages.

Hair loss is commonly observed in patients taking fluoxetine or paroxetine.

Barbiturates and benzodiazepines do not cause hair loss.

Table 8.2 Drugs reported to produce hair loss

ACE inhibitors (captopril, enalapril, moexipril, ramipril)
Allopurinol
Amfetamines*
Amiodarone
Analgesics/antinflammatories (ibuprofen, indometacin, naproxen)
Androgens°°°°*
Anticoagulants (coumarins, dextran, heparin/heparinoids)*
Antiepileptics (carbamazepine, hydantoines, troxidone, valproic acid, vigabatrin)*
Antipsychotics (flupenthixol decanoate, fluphenazine decanoate)
Antithyroid drugs (carbimazole, iodine, thiouracil)*
Appetite suppressants
Aromatase inhibitors (fadrozole, 4-OHA, vorozole)°°°°*
Benzimidazoles (albendazole, mebendazole)
β blockers (levobunolol, metoprolol, nadolol, propanolol, timolol)*
Bromocriptine
Buspirone
Butyrrophenones
Cantharidine
Cholestyramine
Chloramphenicol
Cidofovir
Cimetidine
Clonazepam
Clotrimazole
Colchicine
Contraceptives (oral)°°°
Danazol
Diclofenac
Dixyrazine
Dyazoxide
Ethambutol
Etionamide
Gentamicin
Glatiramer acetate
Glibenclamide
Gold salts
G-CSF (granulocyte-colony stimulating factor)
Haloperidol
Hypocholesterolemic drugs (clofibrate, fenofibrate)
Immunoglobulins
Indandiones
Indinavir*
Interferons*
Isonicotinic acid hydrazide°°
Leflunomide*
Levodopa
Lithium*
Maprotilene
Mesalazine
Methyldopa
Methysergide
Methyrapone
Minoxidil°
Nicotinic acid
Nitrofurantoin
Octreotide
Olanzapine
Pentosone polysulphate
Phenindione
Piroxicam
Potassium thiocyanate
Pyridostigmine
Radiation (<700 Gy)°°*
Retinol (vit A)*
Retinoids (acitretin, etretinate, isotretinoin)*
Risperidone
Salicylates
Serotonin uptake inhibitors (fluoxetin, paroxetin)*
Sulfasalazine
Tamoxifene
Terbinafine
Terfenadine
Thiamphenicol
Thrimetadione
Thyroxine
Tocopherol (vitamin E)
Trazodone
Triazoles (fluconazole, itraconazole)
Tricyclic antidepressants (amytriptiline, desipramine, doxepin, imipramine, maprotiline)
Triparanol
Vasopressin
Spironolactone

*Established by multiple reports or proved by rechallenge.
°May produce permanent alopecia.
°°May produce anagen effluvium.
°°°May produce telogen effluvium 3 months after discontinuation.
°°°°May produce androgenetic alopecia.

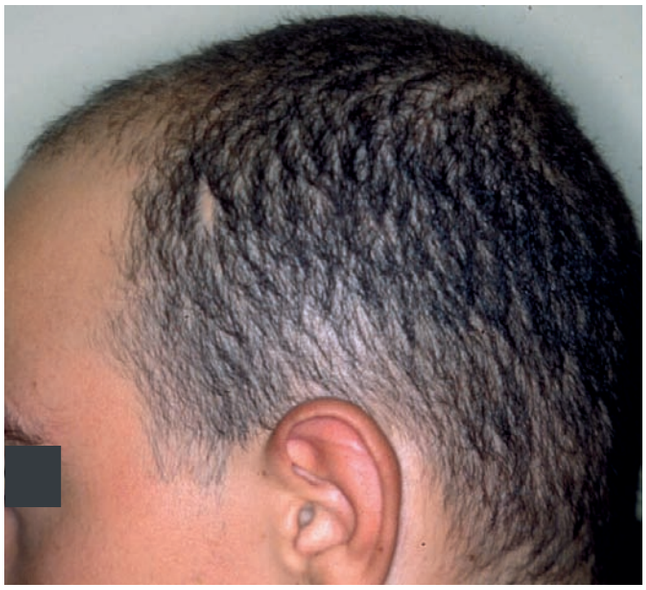


Figure 8.2
Diffuse hair thinning after chemotherapy.

Antiretrovirals

Severe telogen effluvium and patchy hair loss resembling alopecia areata are a common side effect of indinavir therapy, occurring in up to 10% of patients (Figure 8.3). Legs, pubic, thoracic and axillary hair are also often involved. Indinavir can also cause changes in hair texture and shape.

Hypertrichosis and darkening of the pubic hair have been reported in patients taking zidovudine (AZT).

Interferons (Table 8.3)

Hair loss occurs in up to 30% of patients and is not dose-related. Hair shedding is severe (Figure 8.4) and produces visible alopecia in most cases. Transient localized alopecia can also occur at the site of IFN-α injections.

Table 8.3 Interferons: hair side effects

Telogen effluvium
Alopecia at the injections sites
Hair graying
Eyelashes and eyebrows hypertrichosis
Hair straightening
Rough texture

Acquired hair straightening has been reported in patients treated with interferon-α plus ribavirin for HBV infection. Symptoms regressed after drug discontinuation and recurred after drug reintroduction.

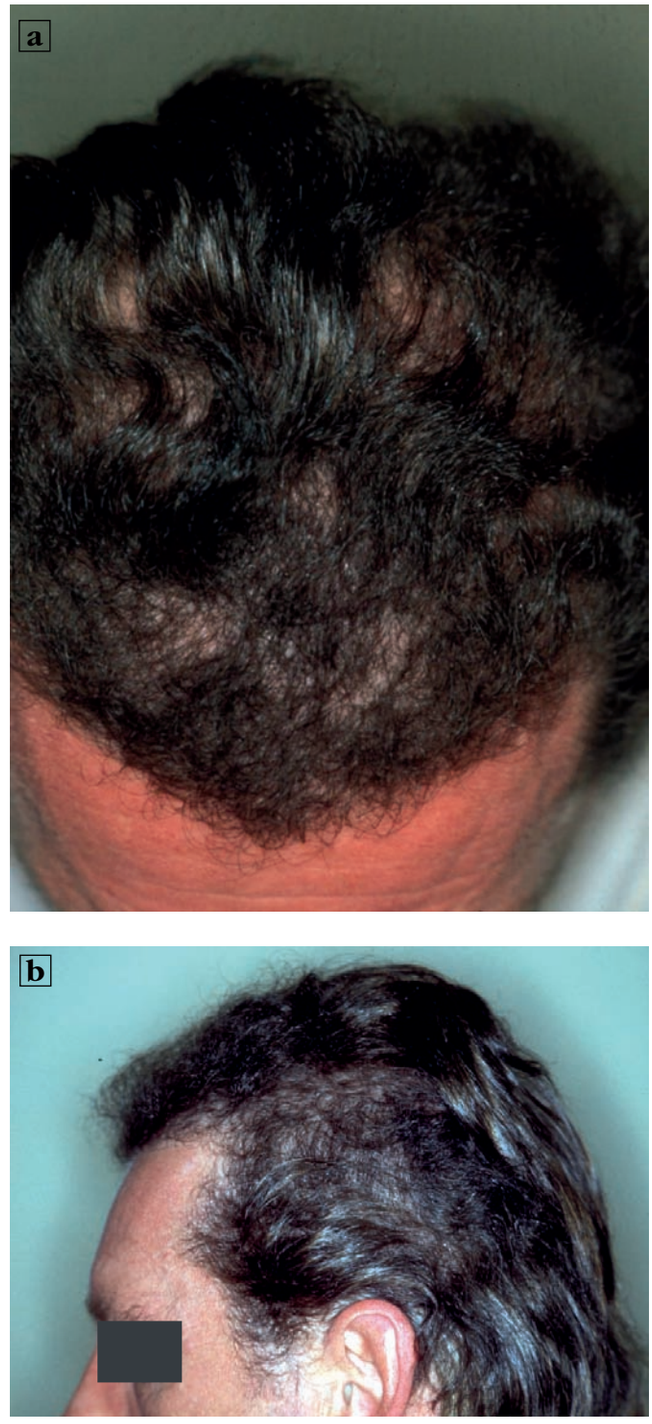


Figure 8.3a,b
Telogen effluvium and hair kinking in a patient treated with indinavir.

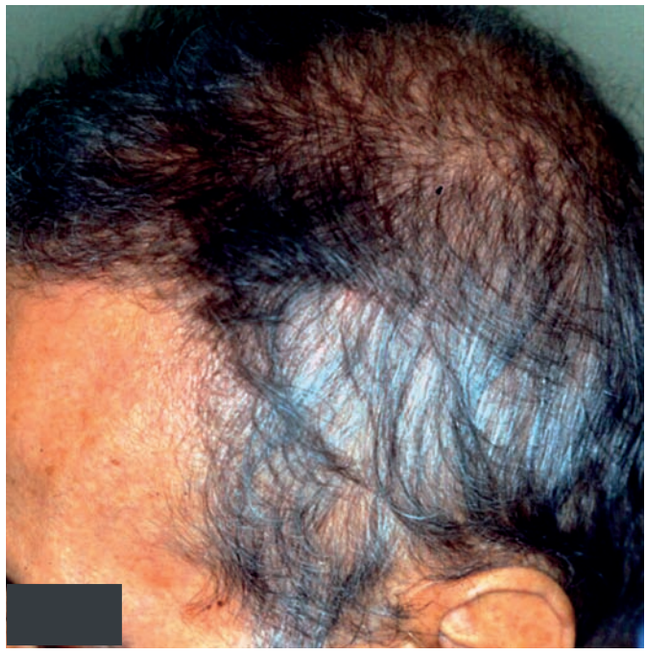


Figure 8.4
Severe telogen effluvium in a patient treated with interferon.

Minoxidil

Telogen effluvium occurs 2–3 months after topical minoxidil interruption. Hair loss is often severe and results from simultaneous telogen entry of all follicles that had prolonged their growth under minoxidil stimulation.

Treatment with 1 mg finasteride does not prevent telogen effluvium in patients interrupting minoxidil.

Hair loss may also occur at the beginning of minoxidil therapy; this might be due to the fact that initiation of anagen induced by minoxidil stimulates teloptosis.

Oral contraceptives

Interruption of oral contraceptives is frequently followed by telogen effluvium. This is due to the fact that estrogens contained in contraceptives prolong anagen duration and synchronize the cycle of scalp hair. This is followed by contemporary entry into rest of a large number of follicles after estrogen interruption.

Contraceptives containing androgenic progestants may induce or worsen androgenetic alopecia.

Radiation

Temporary patchy alopecia resembling alopecia areata is a possible side effect of neurosurgical operations with fluoroscopic imaging (Figure 8.5). Anagen hair loss due to epilating doses of X-ray (300–600 Gy) involves the retroauricular areas which receive the highest doses during fluoroscopy.

Retinoids

Acitretin, etretinate and isotretinoin cause hair loss with visible alopecia in up to 20% of patients (Figure 8.6).

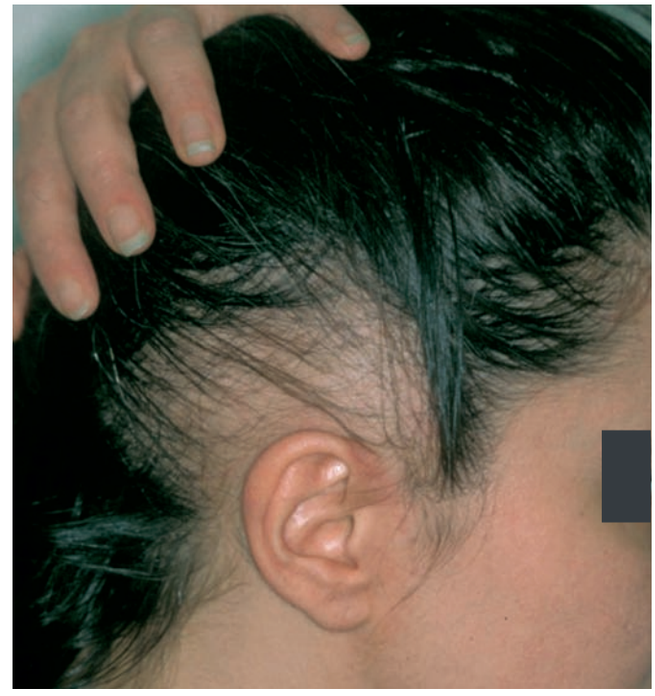


Figure 8.5
Temporary alopecia after microsurgery with fluoroscopy.

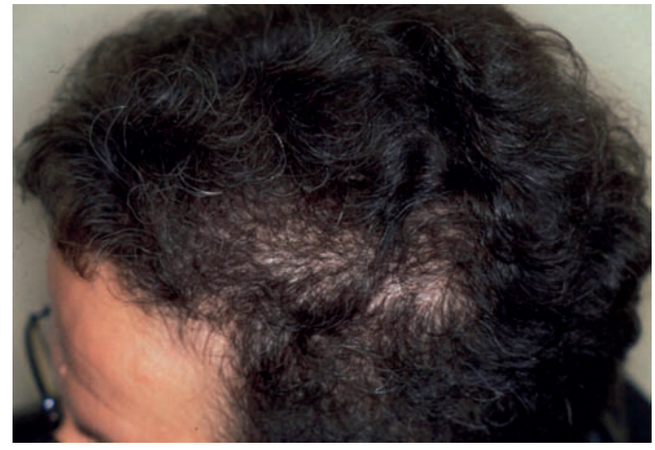


Figure 8.6
Hair loss during retinoid treatment.

The side effect is dose-related and may affect also body hairs. In some cases alopecia may be very severe and total alopecia has also been reported. Premature teloptosis probably has an important role in retinoid-induced hair loss.

Hair lightening and acquired hair kinking are other possible side effects of retinoids.

Retinol (vitamin A)

High dosages of vitamin A cause hair loss. In our experience mild hair loss is frequent in patients taking vitamin supplementations containing vitamin A.

Contemporary administration of vitamin E increases vitamin A toxicity.

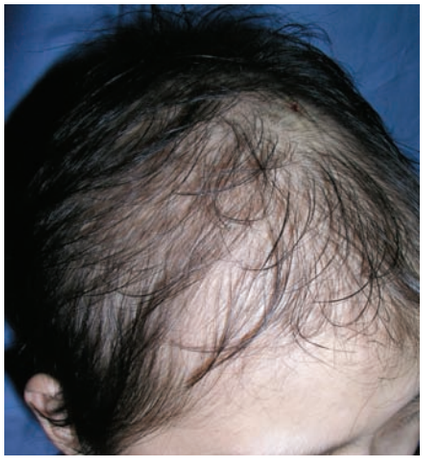
Permanent alopecia

Busulfan (Figures 8.7–8.9)

Busulfan conditioning for allogenic or autologous bone marrow transplantation produces permanent alopecia in up to 50% of patients.

Radiation

Radiotherapy of brain tumors often produces cicatricial alopecia. X-ray dosages >700 Gy permanently destroy the hair follicle (Figures 8.10–8.12).



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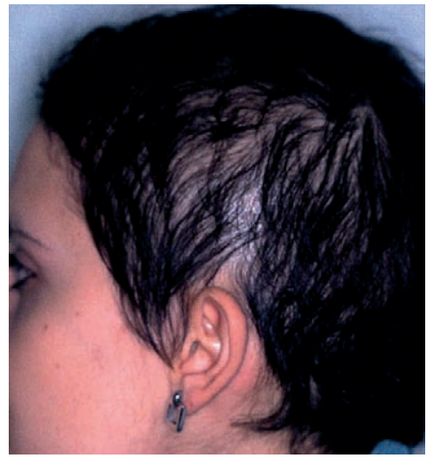

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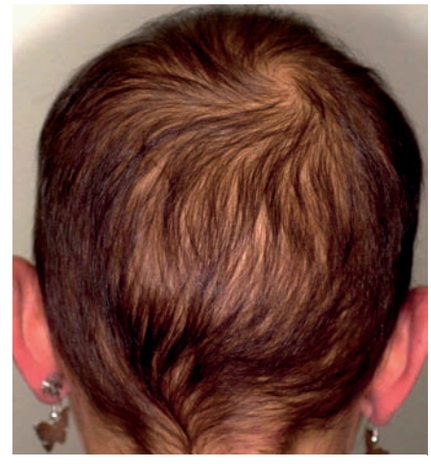

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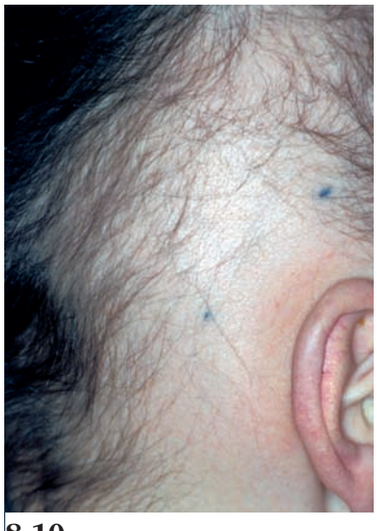
Figures 8.7–8.9
Permanent alopecia due to busulfan after bone marrow transplantation.



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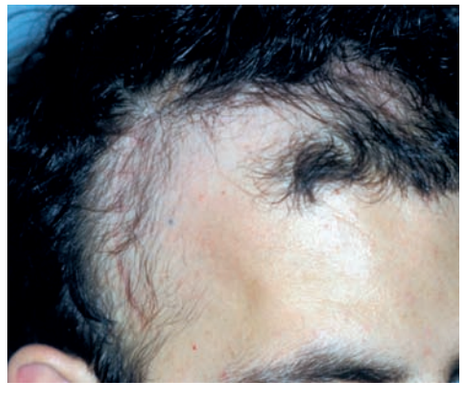

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Figures 8.10, 8.11
Cicatricial alopecia after radiotherapy.

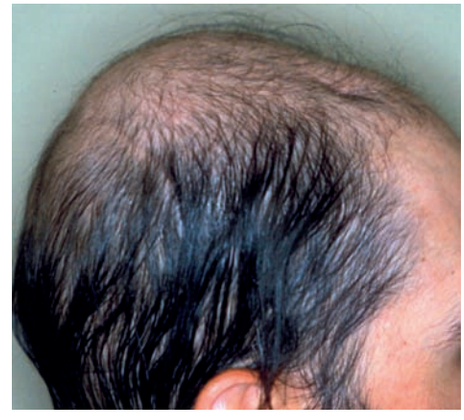


Figure 8.12
Diffuse cicatricial alopecia after radiotherapy.

Excessive hair growth (Table 8.4)

Drugs with androgenic effect may induce hirsutism in predisposed subjects. Excessive hair growth is often associated with other signs of hyperandrogenism, including acne, seborrhea and androgenetic alopecia.

Hypertrichosis is a reversible side effect of several drugs that stimulate the hair follicle to enter and prolong anagen with mechanisms which in most cases are still not completely understood. Hypertrichosis most commonly involves the face and upper trunk and is often associated with trichomegaly and elongation of the eyebrows (Figures 8.13, 8.14).

Cyclosporin A

Reversible hypertrichosis is common and most frequently affects the face and back. It is dose-related, occurring in up to 50% of patients taking high dosages of the drugs after transplantation.

It is rarely seen (3% of patients) with dosages <5 mg/kg/day (Figure 8.15).

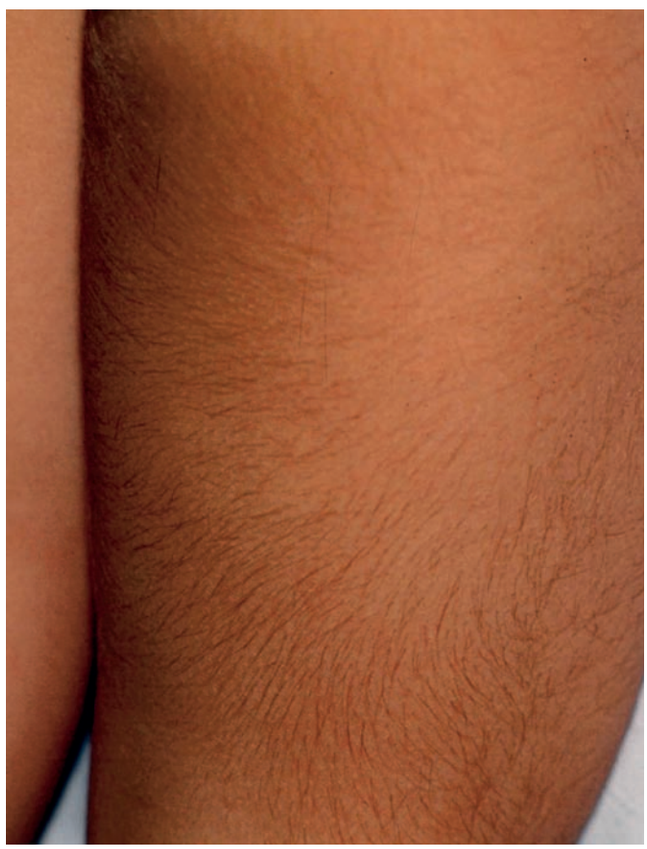
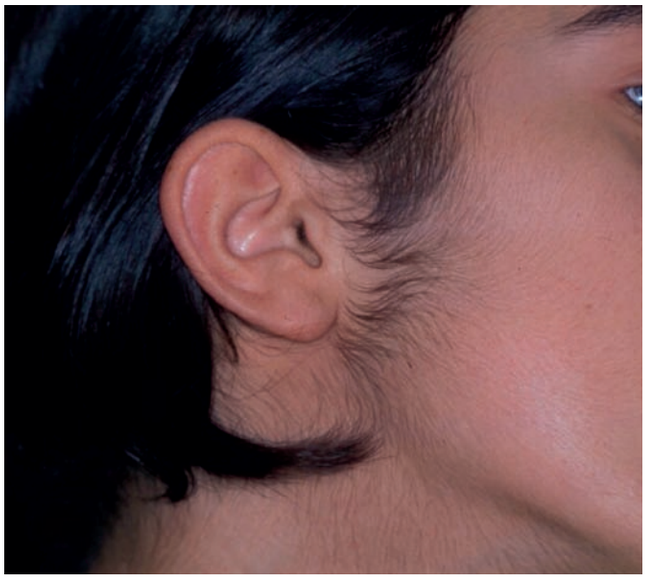


Figure 8.13
Anticonvulsant-induced hypertrichosis.

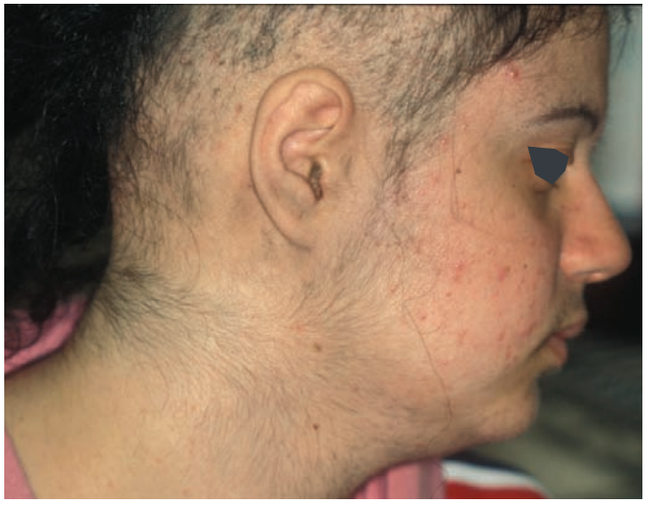
Table 8.4 Drugs reported to produce hirsutism/hypertrichosis

Hirsutism	*Hypertrichosis*
ACTH	Acetazolamide
Anabolizing steroids	Albendazole
Androgens	Benoxaprofen
Carbamazepine	Beta blockers (atenolol, betaxolol)
Danazol	Calcium antagonists (niphedipin, verapamil)
Methyrapone	Cetuximab
	Colestiramine
	Copper
	Cyclosporin A*
	Desipramine
	Diazoxide
	Diltiazem
	Erythropoietin
	Etambutole
	Etionamide
	Fenoterol
	Gentamycin
	Hexachlorobenzene
	Hydantoins*
	Immunoglobulins
	Indometacin
	Interferons
	Melphalan
	Mercury
	Methotrexate
	Methyldopa
	Methyrapone
	Minoxidil
	Mitomycin
	Mitoxantrone
	Nitrosureas
	Penicillamine
	Phenothiazines
	Photochemotherapy
	Pinacidil
	Procarbazine
	Prostaglandin analogs
	Psoralens
	Radiotherapy
	Retinoic acid (tretinoin)
	Sodium tetradecyl sulfate
	Steroids (systemic/topical)
	Streptomycin
	Tacrolimus*
	Temposide
	Thallium
	Thiotepa
	Triciclic antidepressants (imipramine, maprotiline)
	Vasoprexine
	Vinblastine
	Vincristine
	Zidovudine

*Also cause gingival hyperplasia.



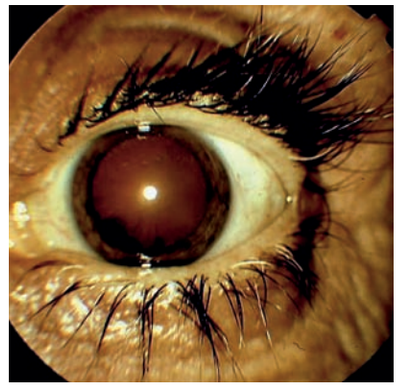
Figures 8.14
Anticonvulsant-induced hypertrichosis.



Figures 8.15
Hypertrichosis from Cyclosporin A

Prostaglandin analogs (Figure 8.16)

Several prostaglandin F analogs utilized for the topical treatment of glaucoma produce darkening of the eyelashes as well as increase in the eyelashes length and number. These include latanoprost, bimatoprost and travoprost.



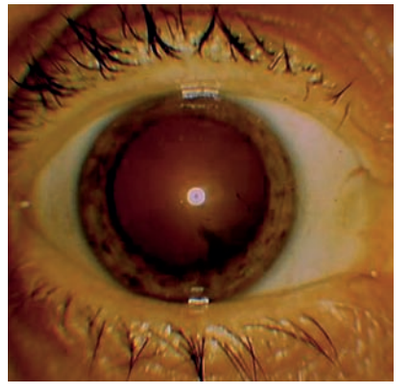


Figure 8.16a,b
Hypertrichosis of the eyelashes in a patient treated with topical bimatoprost for glaucoma of the left eye (a). Note normal length of the eyelashes of the right eye which was not treated.

a

b

Minoxidil (Figures 8.17–8.20)

Topical minoxidil often causes hypertrichosis of the face and neck due to contamination.

Hypertrichosis is more common in women and is not necessarily associated with hair pigmentation.

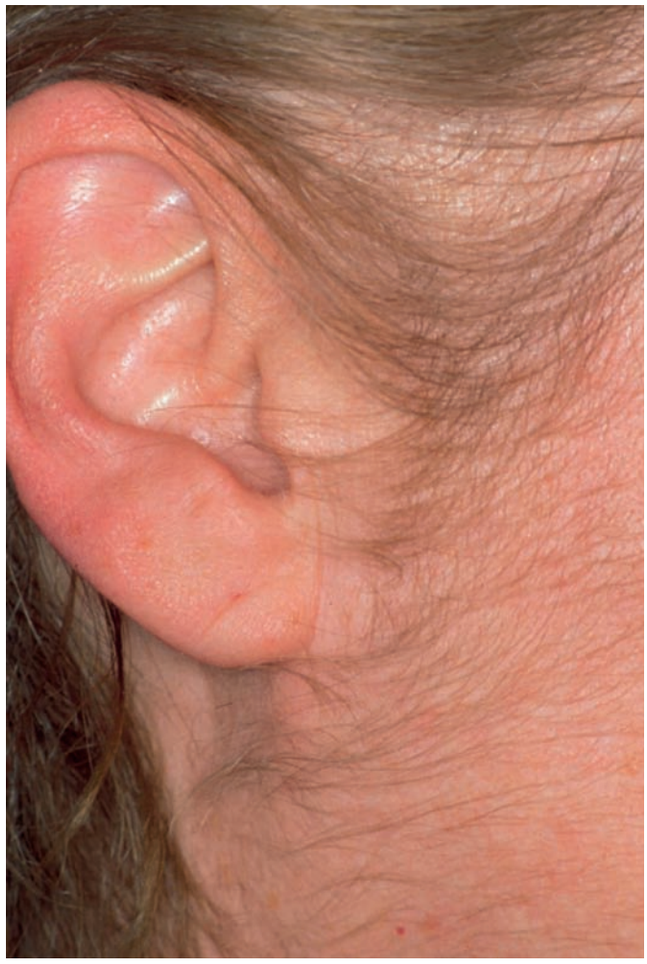
Some patients using 5% topical minoxidil develop diffuse hypertrichosis which is probably due to systemic absorption of the drug.

Changes in hair color (Table 8.5)

Antineoplastics

Hair regrowth after chemotherapy is often characterized by changes in hair color and shape. The hair is often darker and curlier than originally (Figure 8.21).

The flag-sign describes the alternation of horizontal dark and light bands along the hair shaft. This sign is also observed in patients with protein deficiency due to malnutrition (see page 174).



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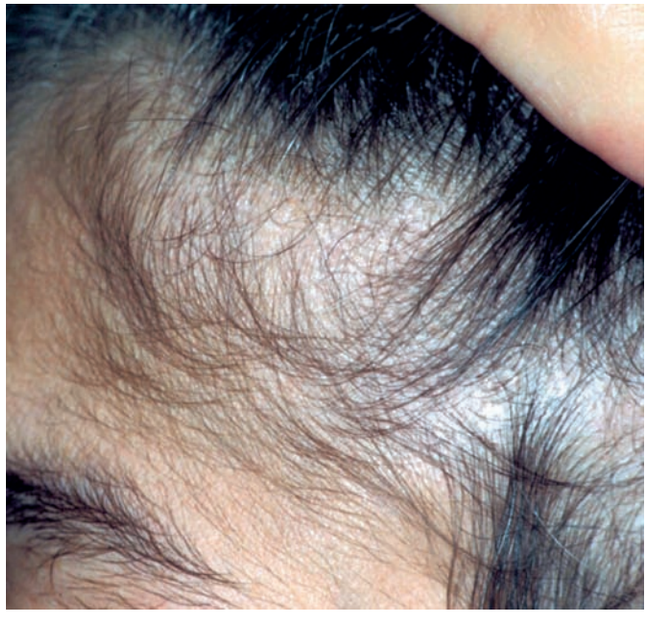


8.18

Figures 8.17, 8.18
Facial hypertrichosis due to topical minoxidil.

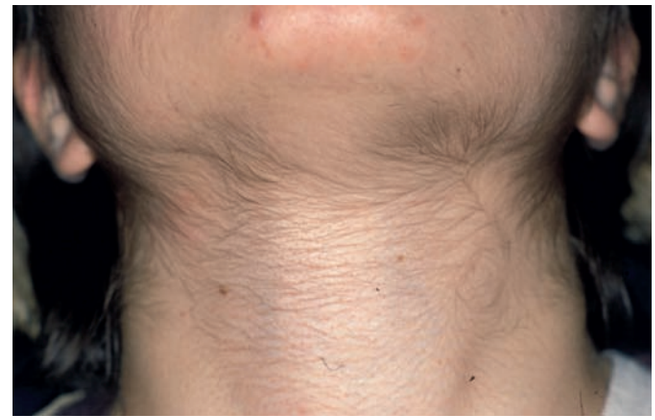


Figure 8.19
Facial and neck hypertrichosis due to topical minoxidil.

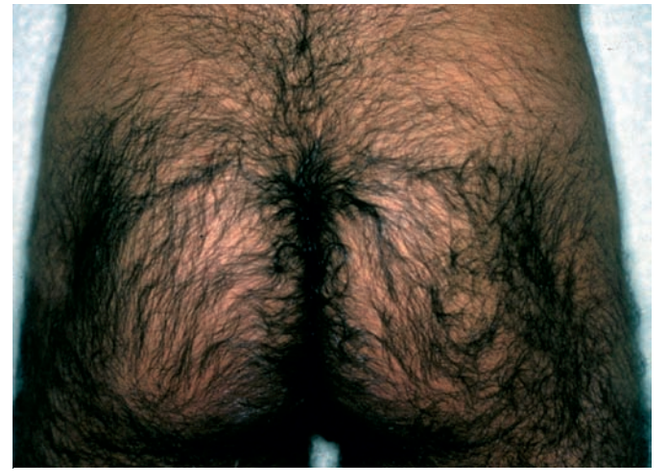


Figure 8.20
Generalized hypertrichosis due to topical minoxidil.

Table 8.5 Changes in hair color

Hair graying	*Hair darkening*
Althesin	Antineoplastics (flag-sign)
Benzoylperoxide	Arsenic
Butyrophenones	Bromocriptine
Chloroquine	Carbidopa
Cyclosporine A (poliosis)	Diazoxide
Etretinate	Estrogens
Hydroquinone	Minoxidil
Interferon α	Para-aminobenzoic acid
Mephenesine	Prostaglandin analogs
Phenols	Radiation (electron beam therapy)
Phenylthiourea	Tamoxifen
Triparanol	Verapamil
	Zidovudine

Figure 8.21
Hair regrowth after systemic chemotherapy. The hair is darker and curlier.

Chloroquine

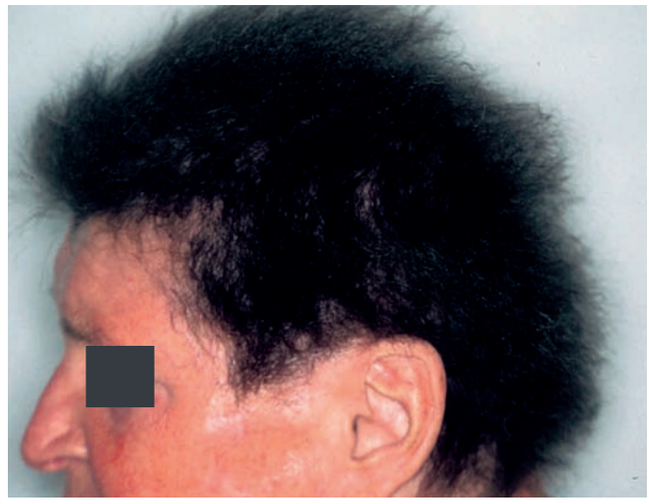
Reversible hair discoloration is a typical symptom of chloroquine treatment, which occurs in blond and red-haired individuals. Hair lightening starts 3–4 months after the beginning of treatment and initially affects the temples and the eyebrows. Hair discoloration is due to a toxic effect of chloroquine on pheomelanin synthesis with accumulation of non-melanized or poorly melanized melanosomes.

Changes in hair shape

Hair straightening or curling has been occasionally associated with drug-intake (Figures 8.22, 8.23, Table 8.6).

Diagnosis

- The history, including diseases and drugs, should explore the 4 months that preceded the onset of hair loss.



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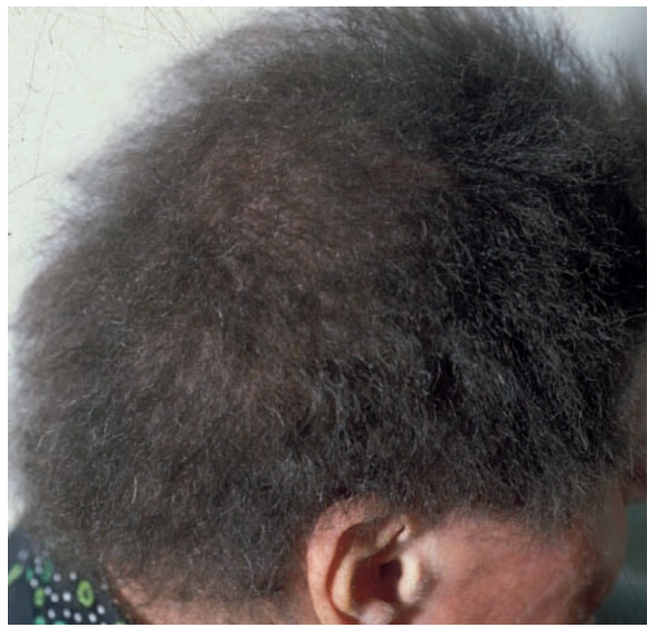

8.23

Figures 8.22, 8.23
Acquired hair kinking in a patient treated with acitretin. Note hair thinning due to retinoid-induced telogen effluvium.

Table 8.6 Changes in the hair shape/texture

Straightening	*Curling/kinking*
Interferon Lithium	Antineoplastics Indinavir Retinoids Valproic acid

- Take a pull test to confirm excessive shedding and examine the hair under the microscope.
- Anagen effluvium is easy to diagnose due to the acute and severe onset and involvement of most of the scalp: the microscopic examination of the hair permits to identify dystrophic hairs.
- Diagnosis of telogen effluvium is suggested by the clinical history and confirmed by a positive pull test with telogen hair roots.
- In telogen effluvium the causative role of the drug is often difficult to prove: the gradual normalization of

hair loss requires 2–3 months after drug discontinuation and induction of the side effect is also delayed with rechallenge.

Management

Drug-induced hair loss may precipitate or aggravate androgenetic alopecia in predisposed individuals.

Telogen effluvium

- Discontinue causative drug when possible.
- Prescribe finasteride or topical minoxidil in presence of androgenetic alopecia.

Anagen effluvium

- Efforts to reduce the severity of alopecia and promote faster regrowth are motivated by the severe impact of anticancer-induced hair loss on the patient's quality of life.
- Available options include scalp hypothermia and topical minoxidil.
- Promising possibilities include:
  - cyclin-dependent kinase 2 inhibitors
  - parathyroid hormone (PTH) related peptyde antagonist
  - p53 inhibitors.

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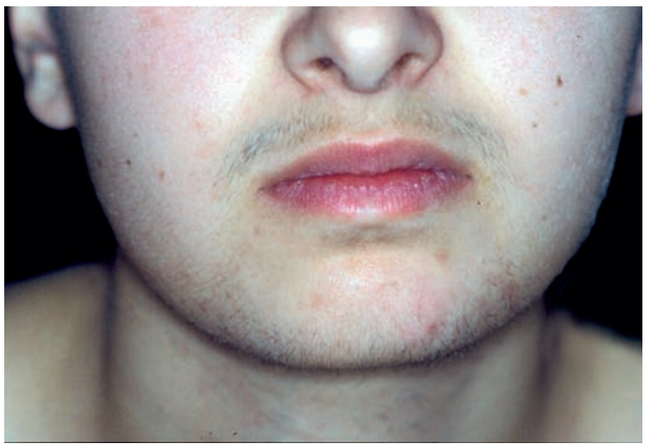
9 Hirsutism

Hirsutism describes excessive terminal hair with male distribution in a female (Figures 9.1, 9.2). Hirsutism is a common condition, affecting up to 10% of women. Prevalence of hirsutism is influenced by genetic and racial factors. It is frequent in Hispanic and Mediterranean women but rare in Asiatic and African women. Hirsutism may or not be associated with hyperandrogenism (Figure 9.3) and/or with acne and androgenetic alopecia. Idiopathic hirsutism accounts for about 15% of cases (Figure 9.4).



9.1

9.2

Figures 9.1, 9.2
Hirsutism: excessive terminal hair on lip and chin.

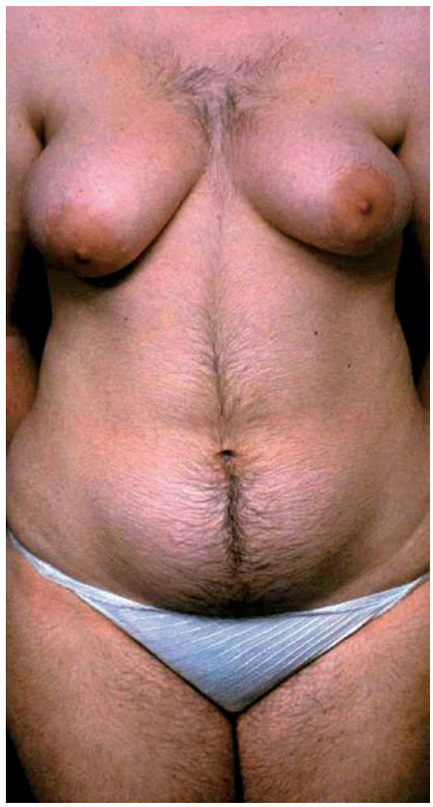


Figure 9.3
Severe hirsutism in a patient with hyperandrogenism.

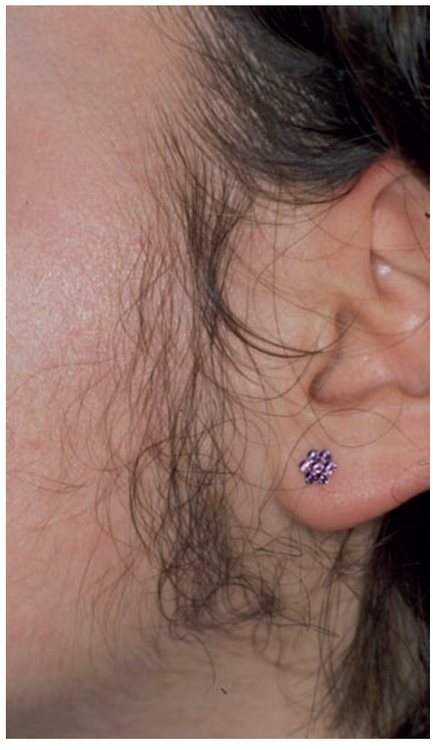


Figure 9.4
Idiopathic hirsutism.

Clinical features

The clinical evaluation of hirsutism is based on the Ferriman–Gallwey score that evaluates the presence of terminal hair in nine androgen-sensitive areas (Table 9.1): lip (Figure 9.5), chin (Figure 9.6), chest (Figure 9.7), upper abdomen, lower abdomen (Figure 9.8), upper

Table 9.1 Ferriman–Gallwey score for hirsutism (Figure 9.12)

<8 no hirsutism
8–15 mild-moderate hirsutism
>15 severe hirsutism

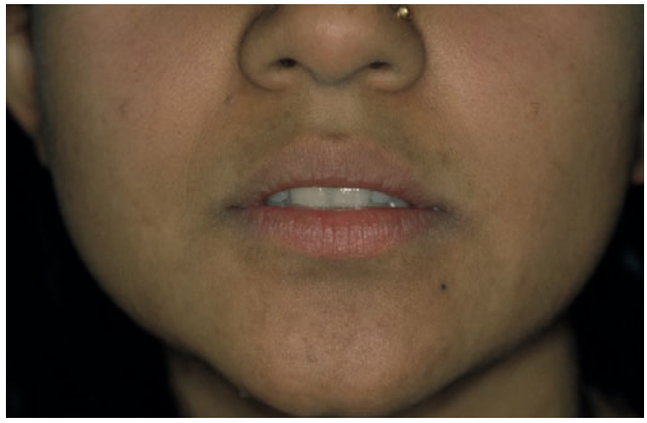
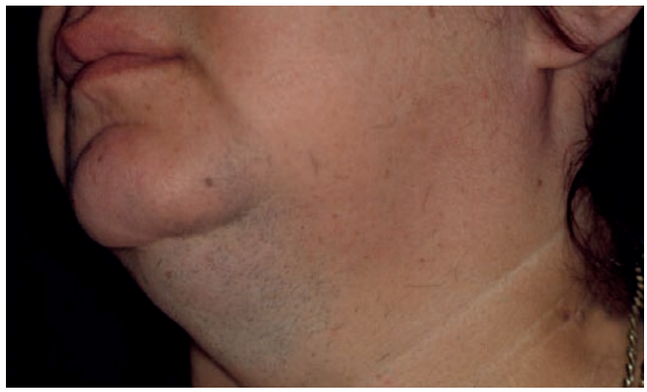


Figure 9.5
Hirsutism: excessive terminal hair on the upper lip.



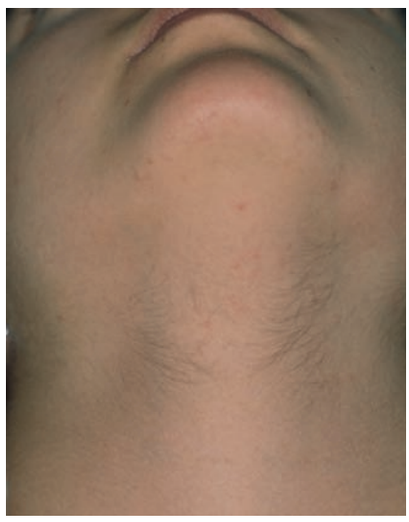


Figure 9.6a,b
Hirsutism: excessive terminal hair on the chin.

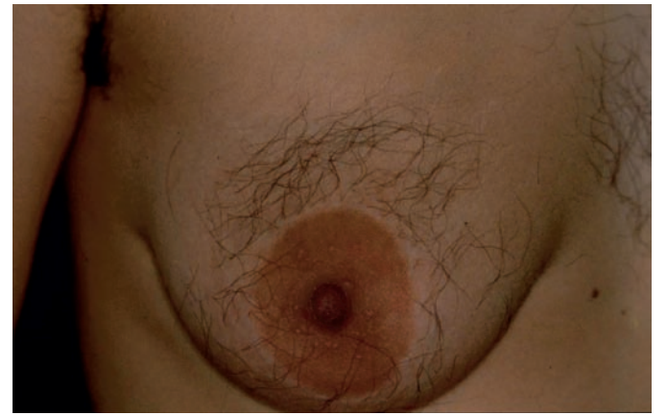


Figure 9.7
Hirsutism of chest and breast.

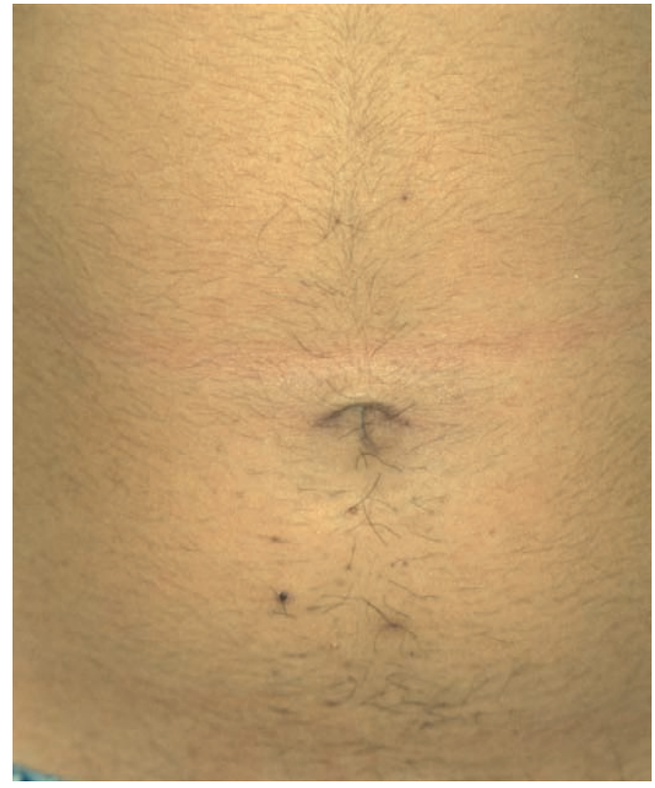


Figure 9.8
Hirsutism: terminal hair on the lower abdomen.

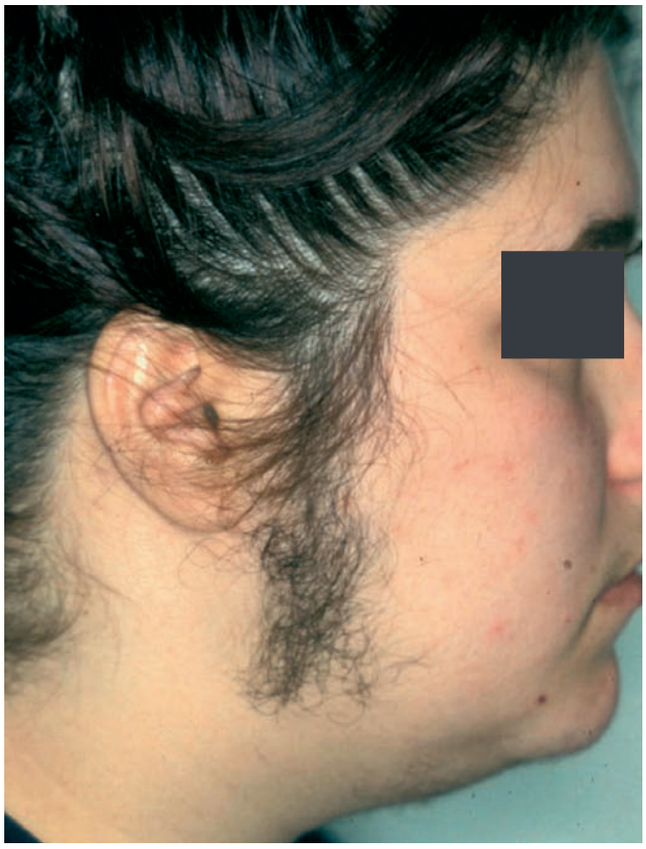


Figure 9.9
Hirsutism in a patient with polycystic ovary syndrome.

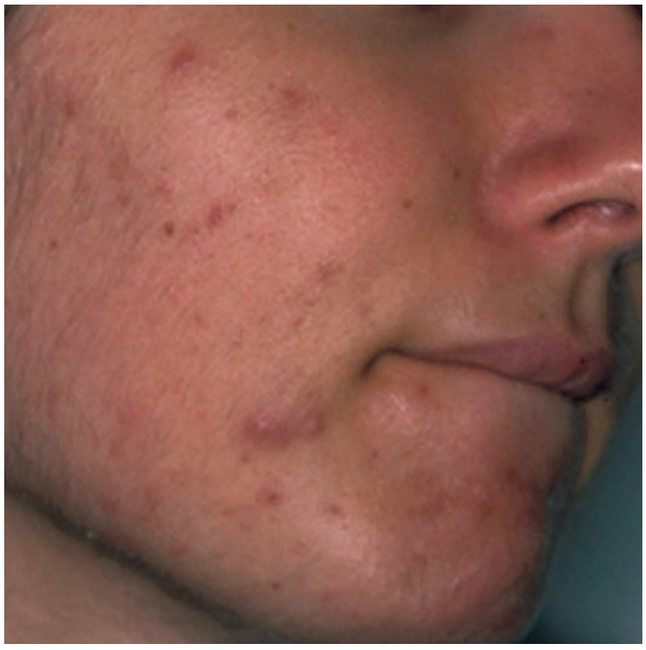


Figure 9.10
Hirsutism and acne in a patient with polycystic ovary syndrome.

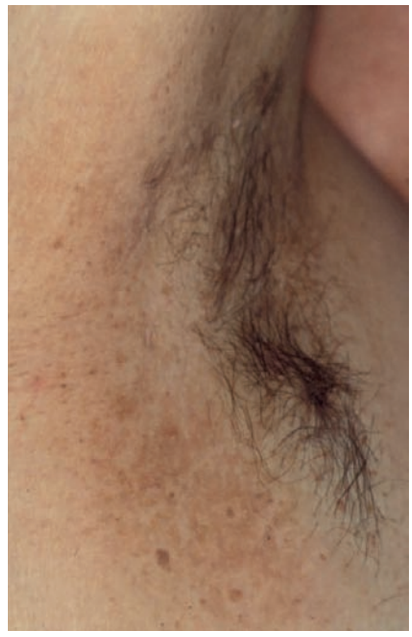


Figure 9.11
Acanthosis nigricans in a patient with PCOS.

back, lower back, upper arm, inner thigh. According with this scale each area has a score ranging from 0 to 4 depending on the presence, thickness and confluence of hair. A diagnosis of hirsutism can be made when the total score is higher than 8/36. A score higher than 15 is usually associated with other signs of hyperandrogenism.

The most common cause of hirsutism is polycystic ovarian syndrome (PCOS) (Figures 9.9–9.11, Table 9.2). Other causes are quite rare (Table 9.3). A diagnosis of idiopathic hirsutism requires demonstration of normal ovulatory cycles and normal androgenic levels (Table 9.4).

Table 9.5 shows laboratory work-up useful to evaluate the presence of hirsutism.

Table 9.2 PCOS: diagnostic criteria – presence of at least two elements

Oligo-anovulation
Hyperandrogenism (clinical and/or biochemical)
Micropolycystic ovaries (12 or more 2–9 mm follicles in each ovary and/or ovarian volume >10 ml)
Exclusion of specific etiologies

Table 9.3 Causes of hirsutism

Adrenal glands	Transitory adrenal hyperandrogenism Congenital adrenal hyperplasia Adrenal tumors Cushing syndrome
Ovaries	PCOS Tumors Hypertrichosis
Drugs	Androgens Oral contraceptives Anabolizing steroids
Hypophisis	Hypophisarian adenoma Hyperprolactinemia Pregnancy Idiopathic Obesity and insulin-resistance

Table 9.4 Diagnosis of idiopathic hirsutism

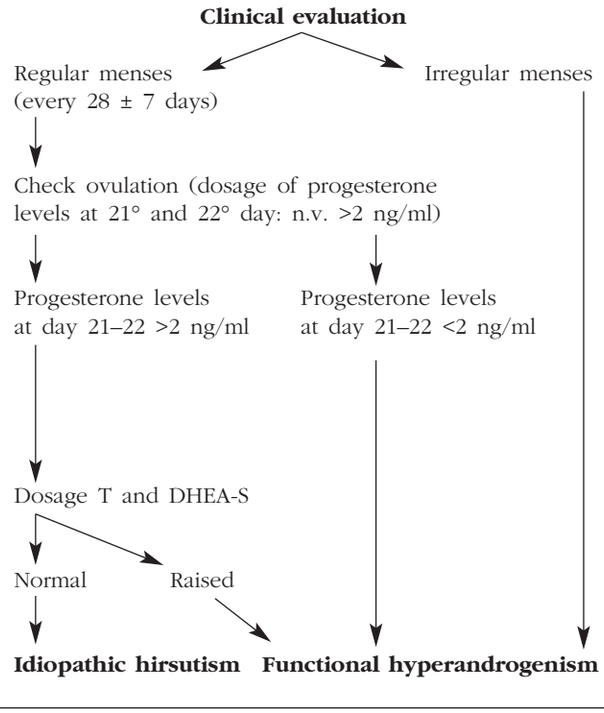


Table 9.5 Laboratory work up

Testosterone*	FSH
DHEA-S**	Prolactine
Androstenedione	SHBG
17-OH-progesterone	Cortisol
LH	Ovary scan

*Testosterone levels higher than 5 nmol/l = ovaric tumors.
**Very high DHEA-S levels = adrenal tumors.

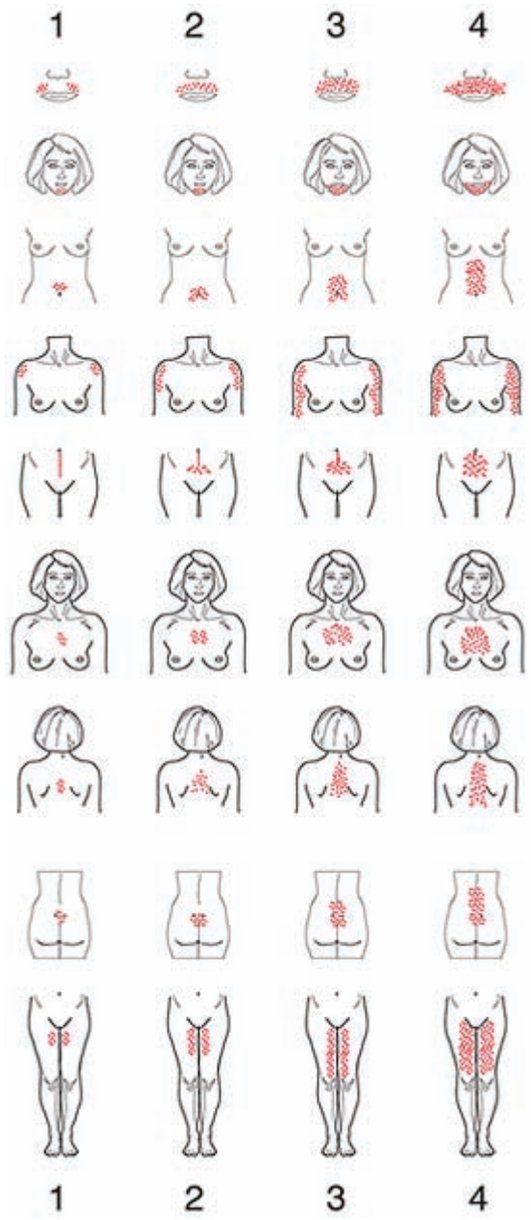


Figure 9.12
Ferriman–Gallwey scale.

Treatment

In most patients treatment should be prolonged for 3–4 years.

- Antiandrogens
  - Spironolacton 100 mg/day associated with contraceptive pill (A)
  - Finasteride 2.5–5 mg/day associated with contraceptive pill (A)
  - Flutamide 125–250 mg/day associated with contraceptive pill (A)
  - Cyproterone acetate 50–100 mg/day from days 5 to 14 of the menstrual cycle, associated with ethynil estradiol or contraceptive pill (A).
- Eflornithine cream (A).

Eflornithine inhibits irreversibly ornithine decarboxylase, an enzyme present in the hair follicle that stimulates hair growth. Topical application of a cream containing eflornithine twice a day produces significant improvement on excessive unwanted facial hair in women. This drug requires long-term use and drug interruption is followed by hair regrowth.

For temporary or permanent hair removal see page 87.

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Beigi A, Sobhi A, Zarrinkoub F. Finasteride versus cyproterone acetate-estrogen regimens in the treatment of hirsutism. *Int J Gynaecol Obstet* 2004; **87**:29–33.

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10 Hypertrichosis

The term hypertrichosis describes the presence of an excessive amount of hair in a non-androgen-dependent area. This results from the presence of terminal hairs in anatomical areas that are normally characterized by vellus hair.

Hypertrichosis can be congenital (Table 10.1) or acquired (Table 10.2), localized or generalized. In both cases hypertrichosis can be an isolated symptom or occur in association with other abnormalities (Table 10.3). Hypertrichosis can also be a feature of numerous genetic syndromes.

Localized hypertrichosis

Congenital melanocytic nevi

Large, coarse, terminal hairs are present in up to 95% of congenital giant melanocytic nevi (Figure 10.1). The presence of hair is not an indicator of possible malignant transformation.

Becker's nevus

Becker's nevus is an epidermal nevus characterized by irregular macular pigmentation with hypertrichosis

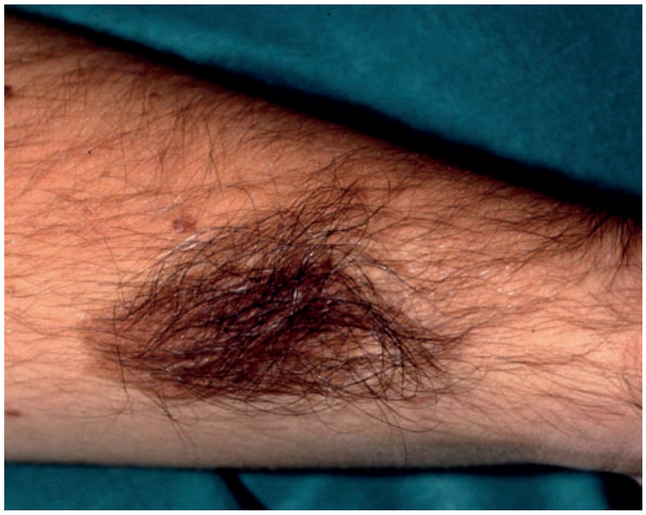


Figure 10.1
Congenital melanocytic nevus with hypertrichosis.

Table 10.1 Congenital hypertrichosis

Localized
Becker's nevus
Cervical hypertrichosis
- Anterior cervical hypertrichosis
- Posterior cervical hypertrichosis

Congenital melanocytic nevi
Faun tail (lumbosacral hypertrichosis; spinal hypertrichosis)
Hairy pinnae
Hairy palms and soles
Hemihypertrophy
- Isolated
- Beckwith–Wiedemann syndrome
- Neurofibromatosis
- Klippel–Trenaunay–Weber syndrome
- Proteus syndrome

Hypertrichosis cubiti (hairy elbows syndrome)
Neurofibroma
Nevoid hypertrichosis
Polythelia pilosa (hairy polythelia)
Primary multifocal localized hypertrichosis
Smooth muscle amartoma

Generalized
Fetal alchool syndrome
Fetal hydantoin
Hypertrichosis universalis
Hypertrichosis lanuginosa

Genetic syndromes
Ambras syndrome
Barber–Say syndrome
Byars–Jurkiewicz syndrome
Cantù syndrome (hypertrichosis, osteochondrodysplasia, cardiomegaly)
Coffin–Siris syndrome
Congenital generalized lipodystrophy (MIM: 269700, 272500)
Cornelia de Lange syndrome (MIM: 122470)
Cowden syndrome (MIM: 158350)
Craniofacial dysostosis
Crouzon craniofacial dysostosis (MIM: 123500)
Hemimaxillofacial dysplasia
Hypomelanosis of Ito
Laband syndrome (MIM: 135500)
Leprechaunism (MIM: 246200)
MELAS syndrome (mitochondrial, encephalomyopathy, lactic acidosis, stroke-like episodes)
Mucopolysaccharidoses
Oliver–MacFarlane syndrome
Porphyrias (congenital)
Rubinstein–Taybi syndrome
Seip–Berardinelli syndrome (MIM: 269700, 272500)
Schinzel–Giedion syndrome (MIM: 269150)
Stiff skin syndrome (MIM: 184900, 260530)
Trisomia 18 (Edwards syndrome)
Winchester's syndrome (MIM: 277950)

Table 10.2 Acquired hypertrichosis

Localized
Acromegaly
AIDS
Cast wearing
Cushing syndrome
Drugs (see page 67) (Figures 10.15–10.20)
Gonococcal arthritis
Hyperthyroidism
Leukemia
Myeloma
Porphyrias (acquired)
Postinflammatory
- Burns
- Contact dermatitis
- Epidermolysis bullosa
- Erysipela
- Erythema nodosum
- Friction
- Insect bites
- Osteomyelitis (chronic)
- Scleroderma
- Trombophlebitis
- UV irradiation
- Vaccination site
- Lymphedema

Reflex sympathetic dystrophy
Scars
Scoliosis
Transient adrenal hyperandrogenism
Traumas

Generalized
Acquired hypertrichosis lanuginosa
Acrodynia
Anorexia nervosa (Figure 10.21)
Brain tumors
Dermatomyositis (juvenile)
Drugs
Encephalitis
Head injuries
Hypothyroidism (congenital) (may be associated with rolled hairs)
Malnutrition
POEMS syndrome (polyneuropathy, organomegaly, endocrinopathy, M-protein, skin changes)
Traumatic shock
Tuberculosis (children)

Table 10.3 Localized hypertrichosis: possible associations

Becker's nevus
Ipsilateral breast and areolar hypoplasia
Ipsilateral limb hypoplasia
Pectus carenatus
Spina bifida
Accessory scrotum
Morphea

Hypertrichosis cubiti
Short stature

Cervical hypertrichosis
Anterior
- Peripheral neuropathy
- Retinal changes

Posterior
- Kyphoscoliosis

Faun tail
Meningioma
Spina bifida
Traction bands
Diastematomyelia
Myelomeningocele
Dermal sinus tract

Hairy pinnae
AIDS
XYY syndrome
Diabetes

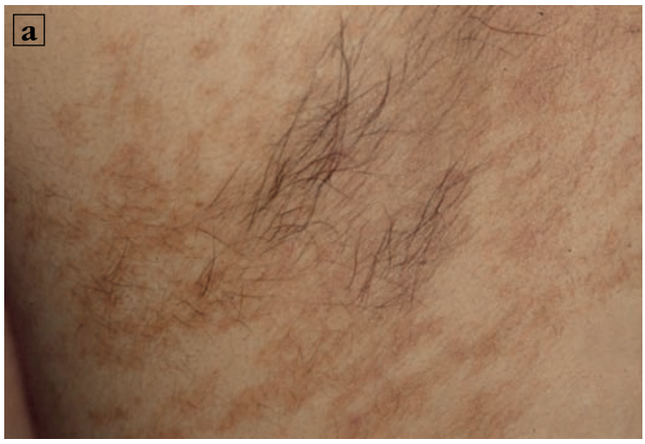
Nevoid hypertrichosis
Cutaneous meningioma

Congenital trichomegaly
Olivier–MacFarlane syndrome
Cornelia de Lange sindrome
Rubinstein–Taybi syndrome

(Figure 10.2). The pigmentation, which is light brown in color, usually develops in childhood or at puberty, most commonly involving the trunk or the upper arm. Hypertrichosis always appears after puberty, usually 2–3 years after the onset of the pigmentation in about 50% of cases. Hypertrichosis of Becker's nevus appears to be androgen-dependent and androgen receptors have been found in the nevus. Although Becker's nevus is reported to occur much more frequently in males than in females (10:1), some authors believe that Becker's nevus in females is often undiagnosed since it is not associated with hypertrichosis.

Hypertrichosis cubiti (hairy elbows syndrome)

This condition, that is usually autosomal dominant, but may be genetically heterogeneous, is characterized by the presence of lanugo hair on the the extensor surface of the elbows extending from mid humerus to mid



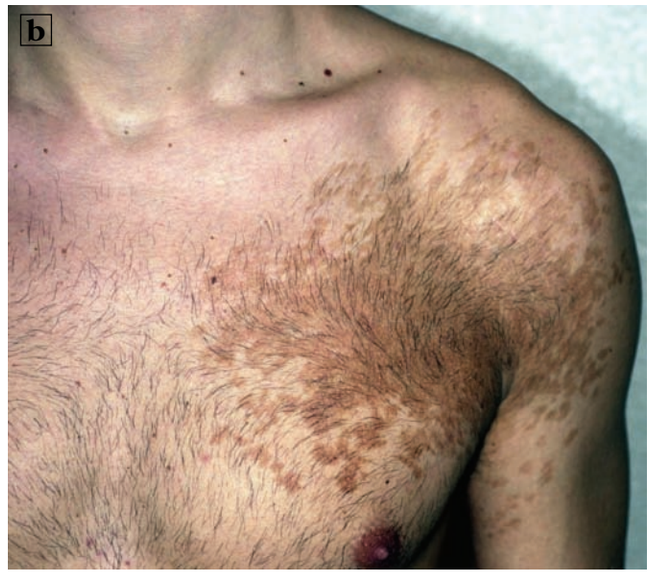


Figure 10.2a,b
Localized hypertrichosis and skin hyperpigmentation in Becker's nevus.

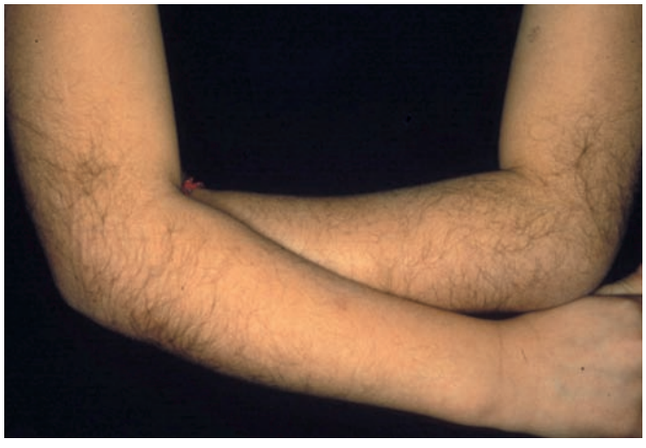


Figure 10.3
Hypertrichosis cubiti.

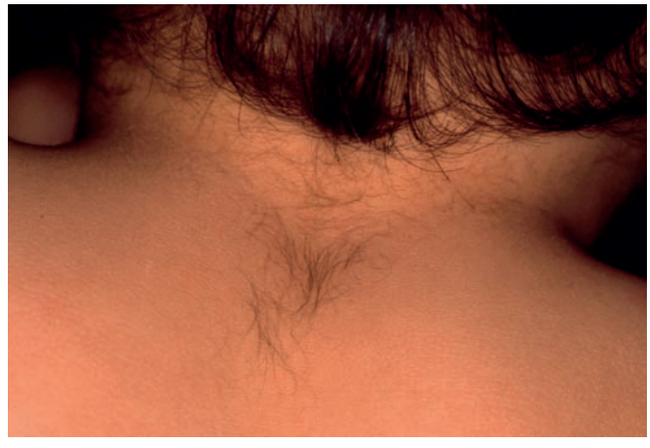


Figure 10.4
Posterior cervical hypertrichosis in a patient with Cornelia de Lange syndrome.

forearm (Figure 10.3). Hypertrichosis cubiti, which is typically bilateral and is usually present since the first months of life to become more evident during childhood, often disappears in adult life.

Cervical hypertrichosis

This may be localized in the anterior or in the posterior side of the neck and is present from birth.

- Anterior cervical hypertrichosis: in anterior cervical hypertrichosis a tuft of terminal hair is present 1–4 cm above the sternal notch. The mode of inheritance is possibly autosomal recessive.
- Posterior cervical hypertrichosis: a tuft of terminal hair is present over the cervical vertebras (Figure 10.4).

An X-linked recessive as well as an autosomal dominant inheritance have been reported.

Faun tail (lumbosacral hypertrichosis; spinal hypertrichosis)

Faun tail describes the presence of a patch of long terminal hair on the lumbosacral region (Figure 10.5). The condition is usually evident at birth or soon afterward. Since prompt diagnosis of the neurological abnormalities

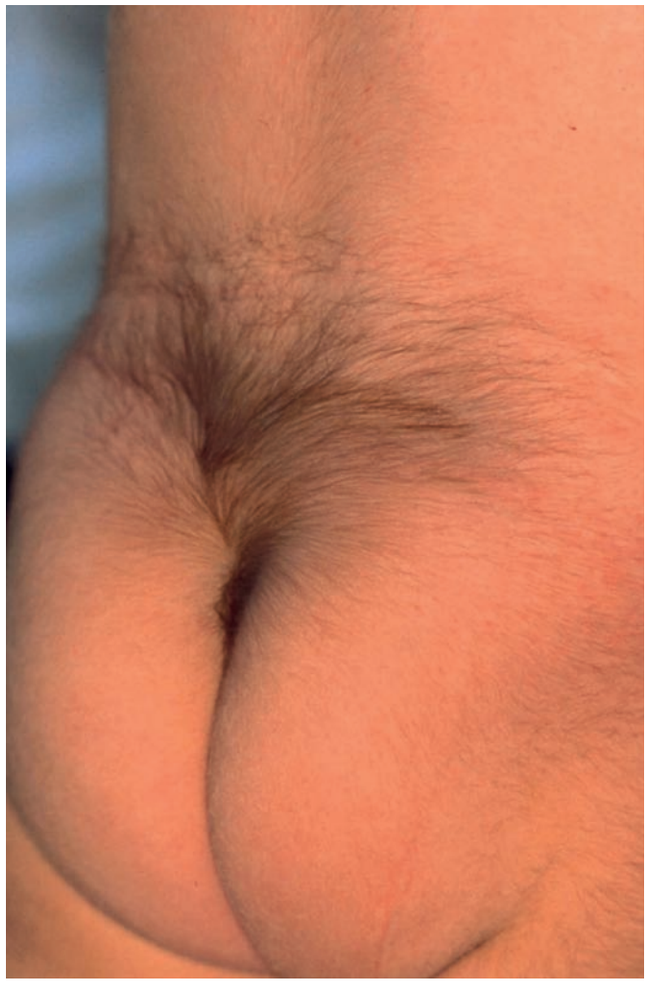


Figure 10.5
Faun tail.

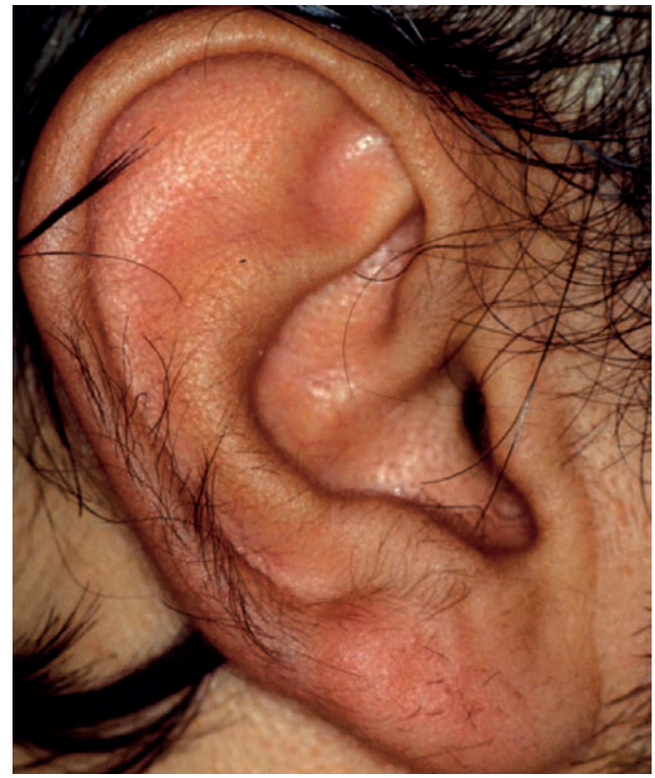
is essential for preventing definitive damage to the nerves, a full neurological and radiological workup is mandatory in all children with faun tail.

Hairy pinnae

The presence of coarse terminal hair on the pinnae (Figures 10.6, 10.7) is a genetic trait that is more frequently observed in Indians but has also been reported in Italians and in other Mediterranean populations. Hairy ears usually become evident after the age of 18.

Hairy palms and soles

In this hereditary condition patches of hairs are present on areas that are normally devoid of hair follicles.



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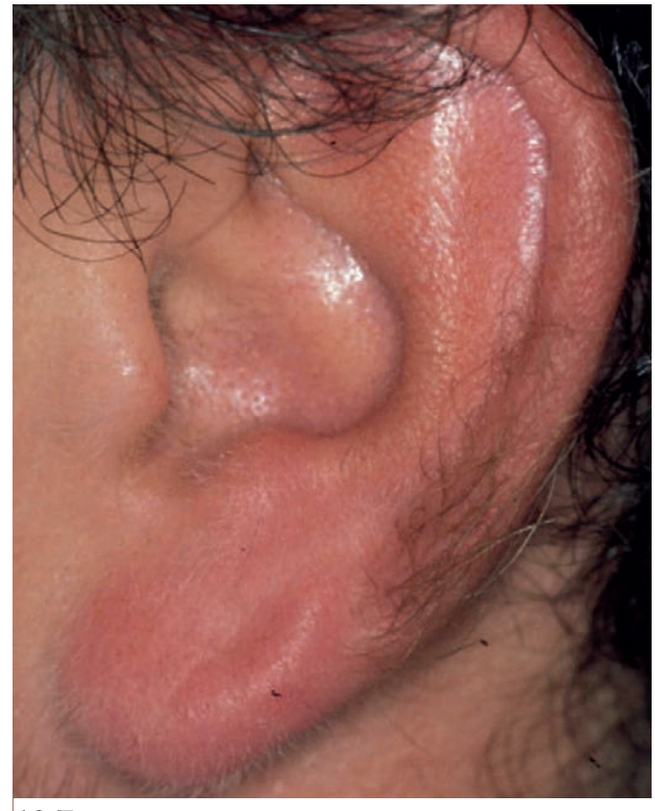


10.7

Figures 10.6, 10.7
Helix hypertrichosis (hair pinnae).

Polythelia pilosa (hairy polythelia)

This is a form of aberrant mammary tissue. Single or multiple tufts of hair occur along the mammary line on the chest and abdomen. The patches of hair are not associated with skin pigmentation or structures of areola or nipple. The condition may be symmetrical.

Nevoid hypertrichosis

This uncommon form of congenital hypertrichosis is characterized by single or multiple patches of terminal hair on apparently normal skin. The scalp can also be involved. Hypertrichosis may spontaneously disappear.

Porphyria

Hypertrichosis of the malar region (Figure 10.8) is a typical feature of porphyria cutanea tarda. Congenital or erythropoietic porphyria are typically associated with hypertrichosis of the face and limbs.

Trichomegaly

Elongation and thickening of the eyelashes is common in HIV patients. It has also been reported in neoplastic patients, in systemic lupus erythematosus and as a side effect of eyedrops containing prostaglandin analogs (Figure 10.9).

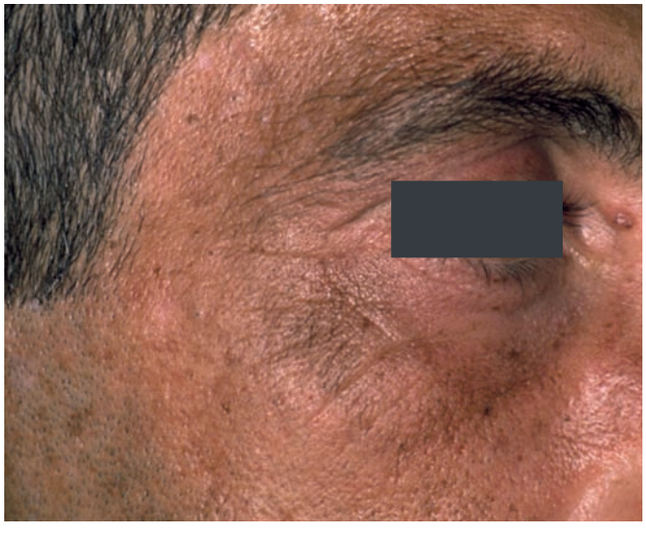


Figure 10.8
Malar hypertrichosis in porphyria cutanea tarda.

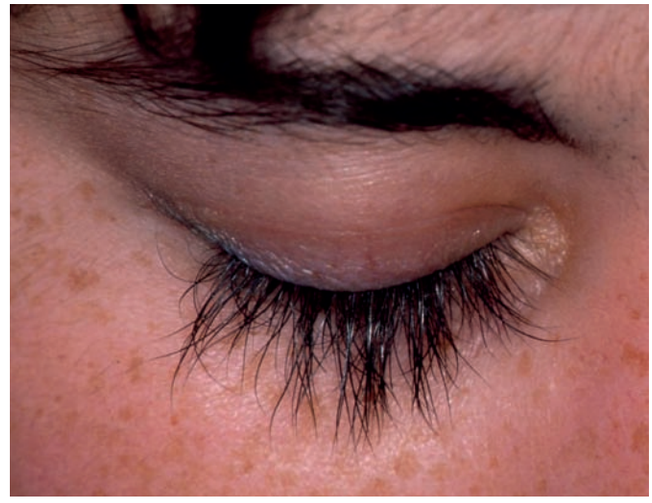


Figure 10.9
Hypertrichosis of the eyelashes in a patient treated with eyedrops containing prostaglandin analogs.

Post-inflammatory/post-traumatic hypertrichosis

Chronic inflammation, scratching and mechanical friction (cast wearing) cause localized hypertrichosis and hyperpigmentation (Figure 10.10).

Prepubertal hypertrichosis

Hypertrichosis of the limbs and/or the back (Figure 10.11) is common in young children. The etiology is

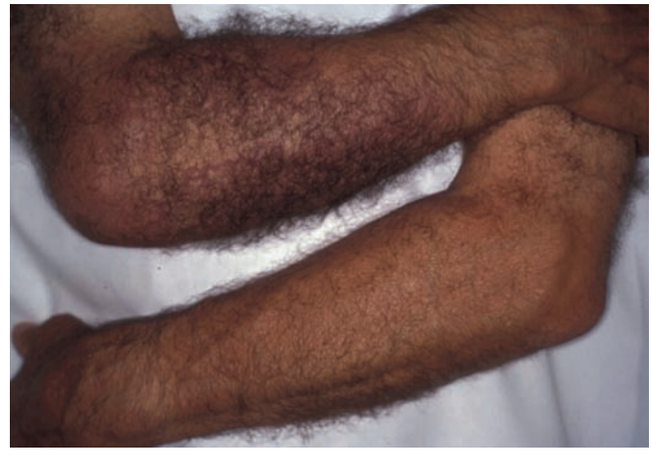


Figure 10.10
Hypertrichosis of the right arm following cast wearing.

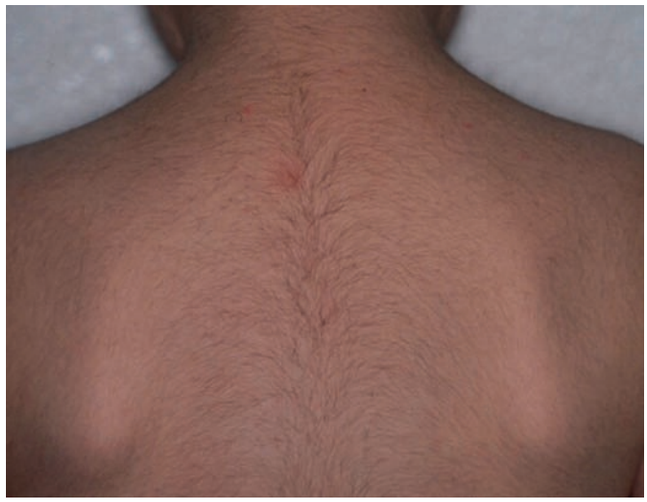


Figure 10.11
Prepubertal hypertrichosis.

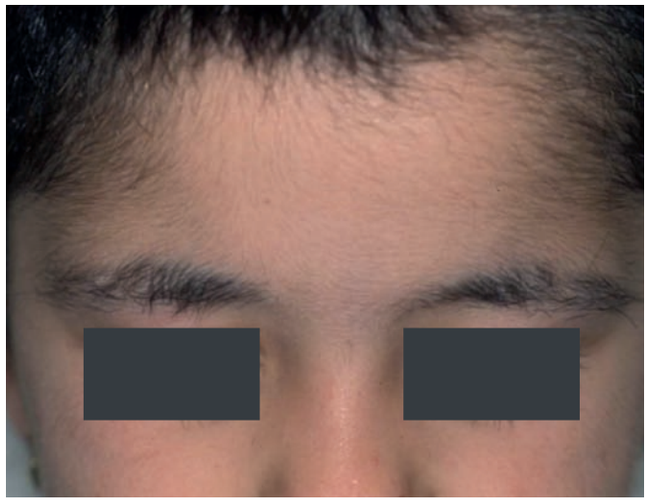


Figure 10.12
Idiopathic hypertrichosis in a young child.

unknown and it is not clear if it is an abnormal entity or an extreme form of the normal range of hair growth (Figure 10.12).

Generalized hypertrichosis

Hypertrichosis universalis

This variety of hypertrichosis is not rare in men of Mediterranean areas. The hair distribution is normal, but the density and length of the hair is above the normal range (Figure 10.13).

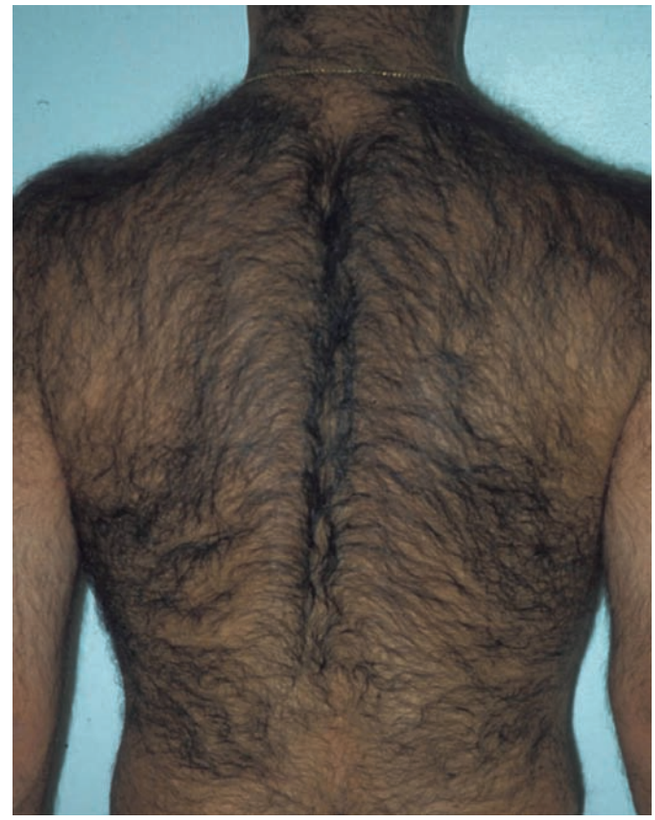


Figure 10.13
Hypertrichosis universalis.

Congenital hypertrichosis lanuginosa (MIM: 145700, 307150)

Hypertrichosis lanuginosa is an exceedingly rare disorder that is most commonly transmitted by an autosomal dominant trait and has been associated with abnormalities of chromosome 8q.

Hypertrichosis is present at birth and affects all the skin surface except for the palms, soles, lips, gland penis and distal phalanges. The abnormal hairs, that may be blond to black in color, are lanugo-type hairs that continue to grow and may reach the length of 5–10 cm (Figure 10.14). In some families the hairs are lost in childhood, in others they persist into adult life.

Hypertrichosis associated with gingival hyperplasia represents a different condition (MIM: 135400) even though the distribution and appearance of the hypertrichosis is similar to that of hypertrichosis lanuginosa. Gingival hyperplasia appears in early childhood and progresses to completely obscure the teeth.

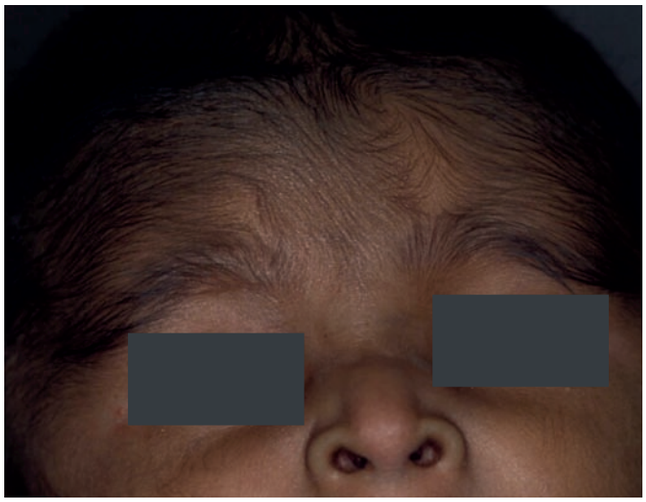
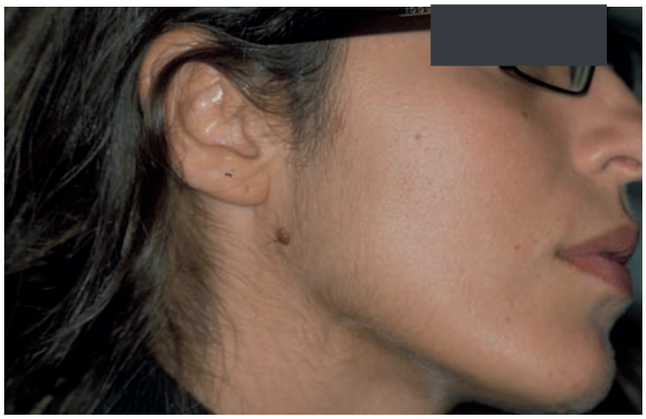


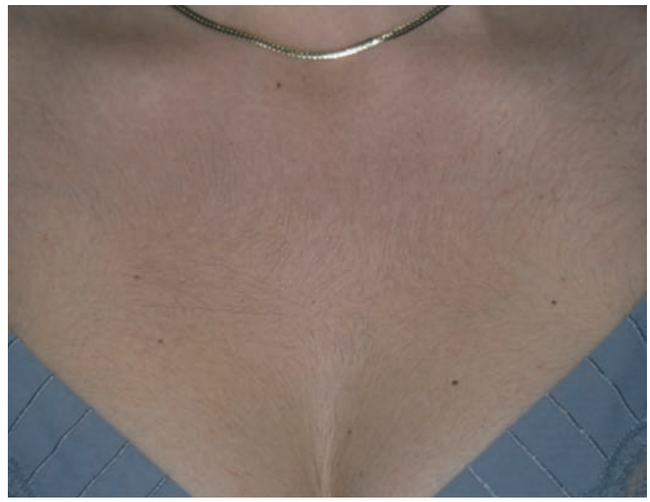
Figure 10.14
Severe hypertrichosis of the face in an Indian boy.

10.15



10.16

Figures 10.15, 10.16
Mild facial hypertrichosis due to topical minoxidil.



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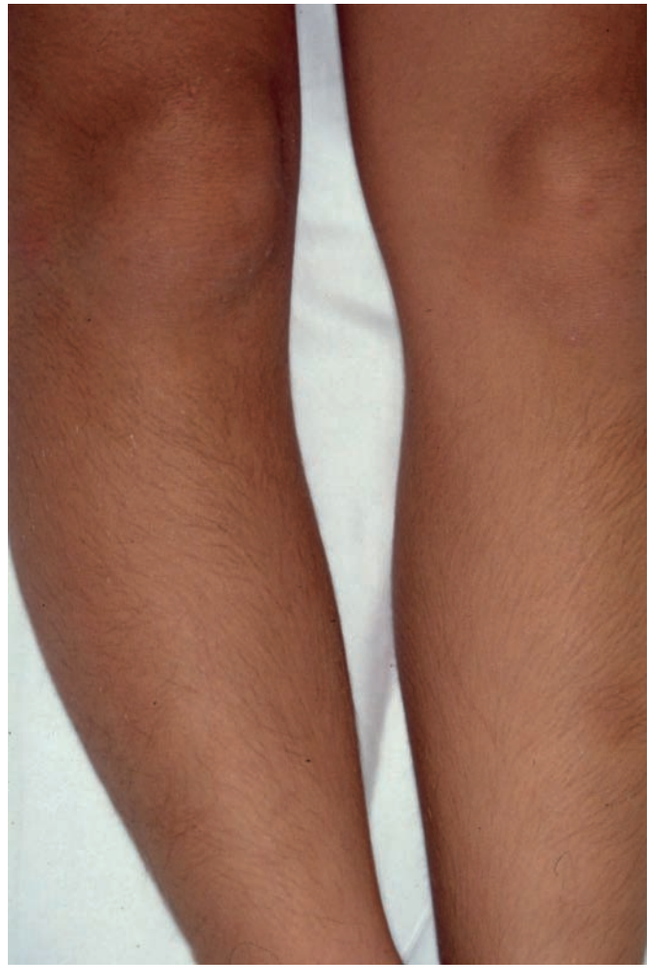

10.18

Figures 10.17, 10.18
Diffuse mild hypertrichosis in a patient undergoing anticonvulsant treatment.

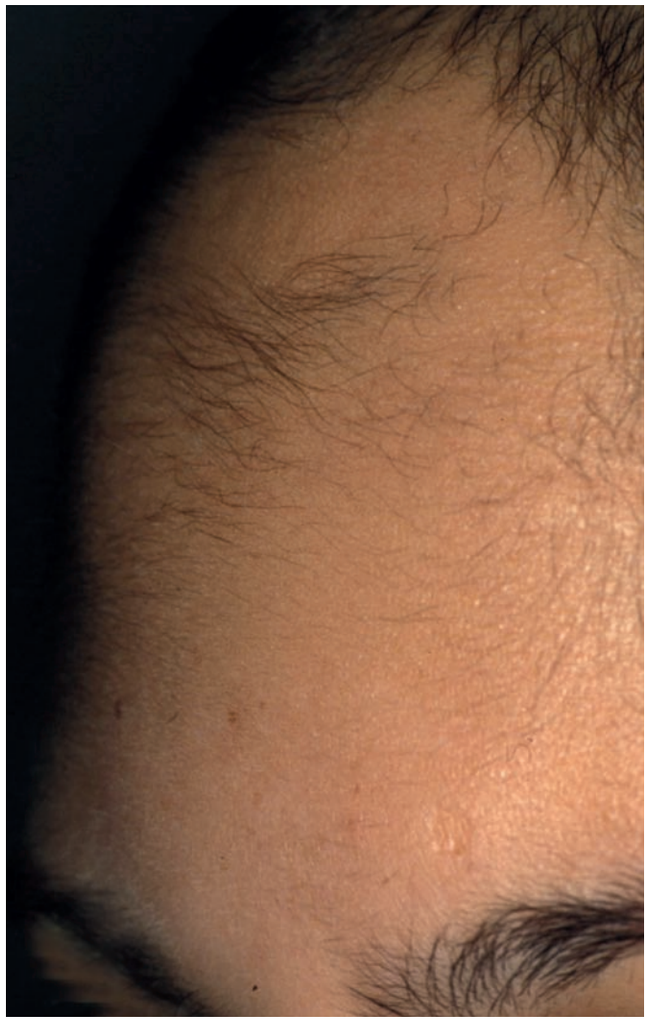

Figure 10.19
Hypertrichosis of the forehead in a patient with alopecia areata treated with systemic steroids.

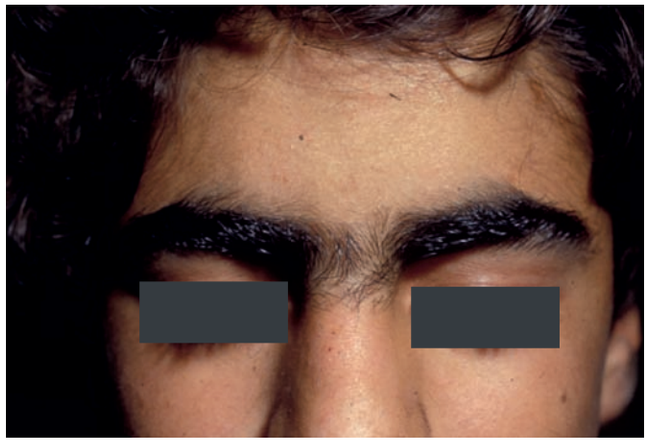

Figure 10.20
Severe facial hypertrichosis in a patient treated with systemic CyA.

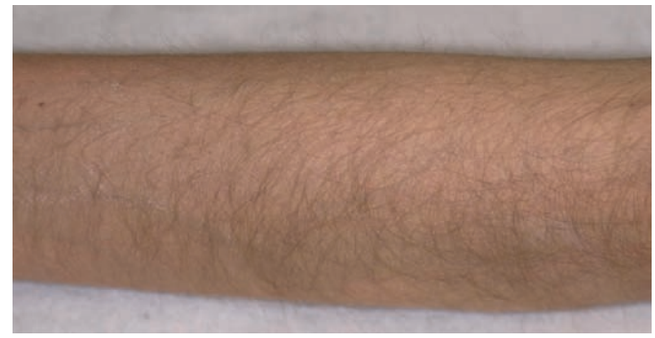

Figure 10.21
Arm hypertrichosis and skin xerosis in anorexia.

Acquired hypertrichosis lanuginosa

This is a rare paraneoplastic syndrome most commonly associated with neoplasms of the lung and bowel. Hypertrichosis develops rapidly with a typical cranio-caudal spread and may precede the other symptoms of tumors by several months. All the skin, except for palms and soles, present lanugo-like hairs that may reach the length of 10–15 cm.

Diagnosis

- Acquired hypertrichoses are most commonly iatrogenic, metabolic, nutritional or paraneoplastic.
- Metabolic and general assessment can help clinical diagnosis.

Treatment (Table 10.4)

- Several depilation (removal of hair at some point along its shaft) and epilation (removal of the entire hair shaft) techniques can be utilized depending on site, patient's age and severity (Figure 10.22a,b).
- Epilation lasts longer than depilation and may cause enough damage to the follicle to provide partial long-term permanent removal.
- Lasers (B): most lasers target melanin (Ruby, Alexandrite, Diode) and are suitable only for dark hair. The Q-switched Nd:YAG is less effective, but suitable also for light hair. Lasers are rapid but expensive. Laser treatment produces delayed hair regrowth and gradual thinning of the treated hairs. Lasers produce a maximum of 30–50% hair reduction after 6 months.

Table 10.4 Treatment of hypertrichosis

	Pro	*Contra*
Bleaching	Simple, quick, painless	Not good for long hairs or dark skin Possible skin irritation
Shaving (Figure 10.23)	Simple, safe	Often not accepted due to the false belief that produces hair thickening and coarsening
Trimming	Simple, painless	Daily for good cosmetic results
Plucking	Longlasting	Slow method, painful folliculitis
Electric plucking	Longlasting	Painful folliculitis, scarring (Figure 10.24)
Wax depilation	Longlasting	Painful folliculitis Requires minimum length
Chemical depilatories	Simple, effective	Irritation (Figure 10.25), unpleasant odor
Electrolysis	Effective, can be specifically targeted to a single hair	Operator-dependent, painful, scarring, dyschromias, suitable for small areas
Laser	Rapid, effective	Expensive, suitable only for dark hair
Eflornitine cream	Painless	Expensive, irritation, small areas

Figure 10.22a
Hair regrowth 1 day after shaving: note the acuminated hair tip.

Figure 10.22b
Hair regrowth 1 day after chemical depilatories: note the round tip.

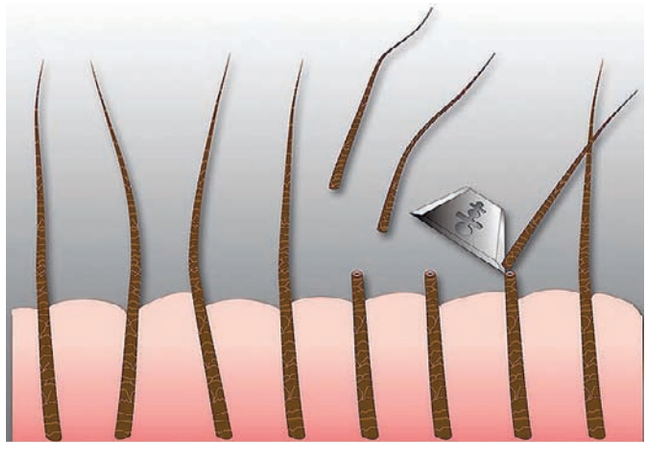

Figure 10.23
Shaving: regrowth occurs in 2–3 days and hair can be very sharp and stubbly because of the spiky tip.

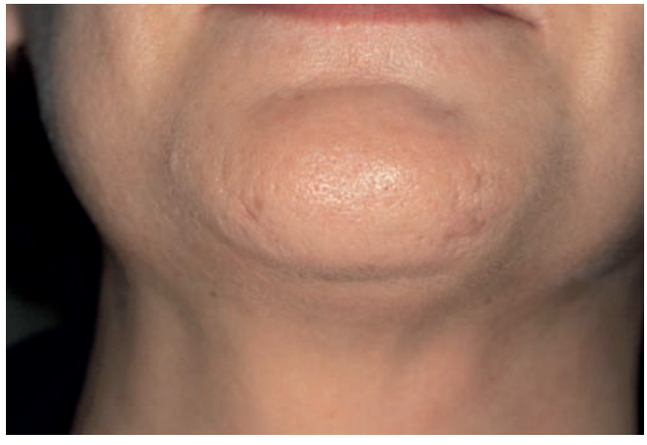

Figure 10.24
Chin scarring after hair electrolysis.

- Eflornitine cream (A) which is suitable for facial hypertrichosis, slows hair growth by inhibiting ornitine-decarboxylase which is essential for cell-growth. Efficacy is moderate.

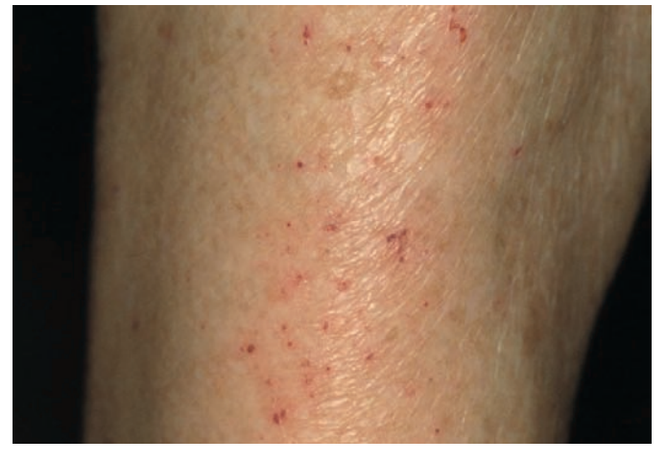

Figure 10.25
Irritative reaction from chemical depilatories.

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11 Hair diseases in children

In newborns hair density and thickness are often variable and the hair color is frequently lighter or darker than the definitive one. Changes from straight to curly and vice versa may occur in scalp hair at puberty. An unruly tuft of hair which streaks straight up is commonly observed (Figures 11.1, 11.2).

Table 11.1 lists hair loss in children according to the age of onset. Hair loss in children can be circumscribed or diffuse, transitory or permanent (Table 11.2).

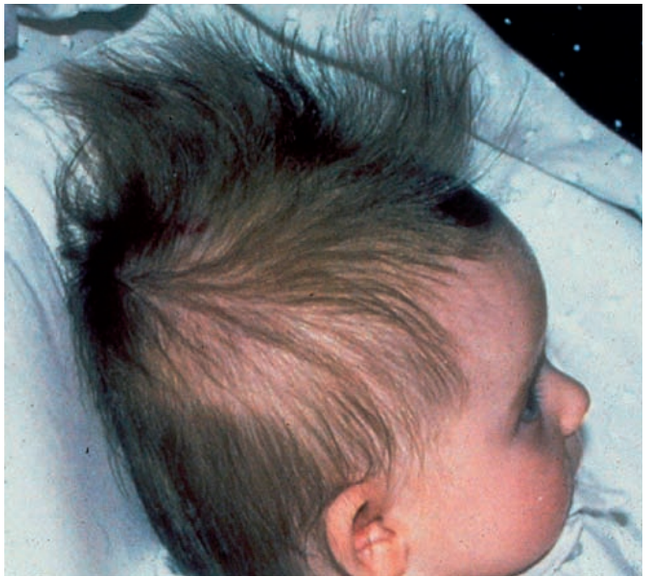


Figure 11.1
Antonella's daughter, Margherita, with straight up hair at the age of 5 months.

Figure 11.2
Margherita's long and straight hair at the age of 3 years.

Table 11.1 Hair loss in childhood: age of onset

At birth	*1st year*	*After*
Aplasia cutis congenita	Acrodermatitis enteropathica	Alopecia areata*
Epidermal nevi/sebaceous nevi	Alopecia due to congenital hair shaft disorders	Epidermolysis bullosa
Meningocele	Atrichia with papular lesions	Hereditary hypotrichosis simplex
	Ectodermal dysplasias	Incontinentia pigmenti
Simple transitory hypotrichosis	Occipital neonatal alopecia (2–3 months)	Loose anagen hair syndrome
		Marie–Unna hypotrichosis
		Short anagen syndrome
		Tinea capitis*
		Triangular congenital alopecia
		Trichotillomania

*Onset at birth or during the 1st year of life is possible.

Table 11.2 Hair loss in children

Circumscribed alopecia

Reversible	*Irreversible*
Alopecia areata (see page 37)	Aplasia cutis congenita
Occipital neonatal alopecia	Congenital triangular alopecia
Trichotillomania (see page 107)	Conradi–Hünerman chondrodysplasia punctata
Tinea capitis (see page 121)	Epidermal nevi
	Epidermolysis bullosa
	Incontinentia pigmenti
	Insect bites
	Meningocele
	Scalp tumors
	Sebaceous nevi

Diffuse alopecia

Reversible	*Irreversible*
Acrodermatite enteropathica	Atrichia with papular lesions
Alopecia areata	Ectodermal dysplasias
Hereditary hypotrichosis simplex	Trichothyodystrophy
Keratosis follicularis spinulosa decalvans	Tricho-rhino-phalangeal syndrome
Loose anagen hair syndrome	
Marie–Unna hypotrichosis	
Menkes' syndrome	
Moniletrix	
Netherton's syndrome	
Short anagen syndrome	
Trichotillomania (see page 000)	

Hair whorls

The angle followed by scalp hairs when they emerge from the scalp forms spiraling or non-spiraling patterns emanating from central whorls. The central point of the whorl is characterized by the divergent growth of hairs. Clockwise hair patterns are usually very evident in children and may be multiple. A single parietal scalp whorl is seen in 95% of infants. Seven per cent of children show a particular hair stream on the forehead: the cowlick (Figure 11.3).

Traumatic alopecia

Traumatic alopecia in newborns may result from mechanical traumas by fetal monitoring during delivery or obstetric procedures.

Scarring alopecia due to skin necrosis with dermal calcinosis may result from prolonged electroencephalographic monitoring.

Figure 11.3

Antonella's son, Lorenzo, showing the cowlick on the forehead.

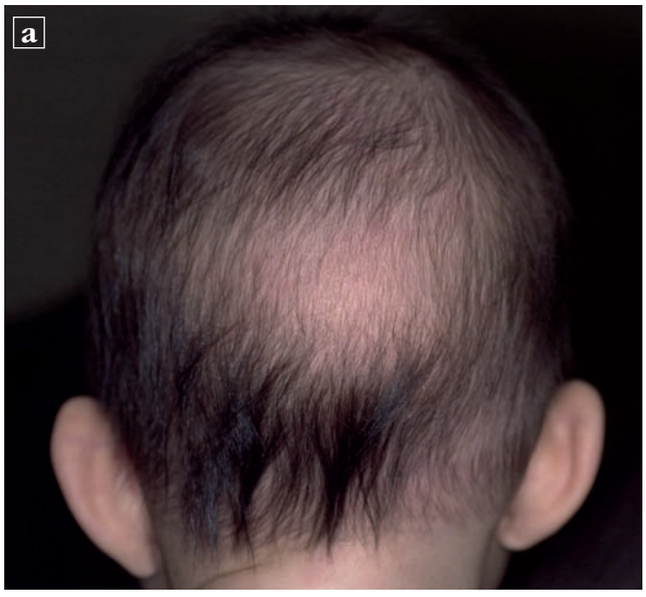


Figure 11.4a,b
Occipital neonatal alopecia.

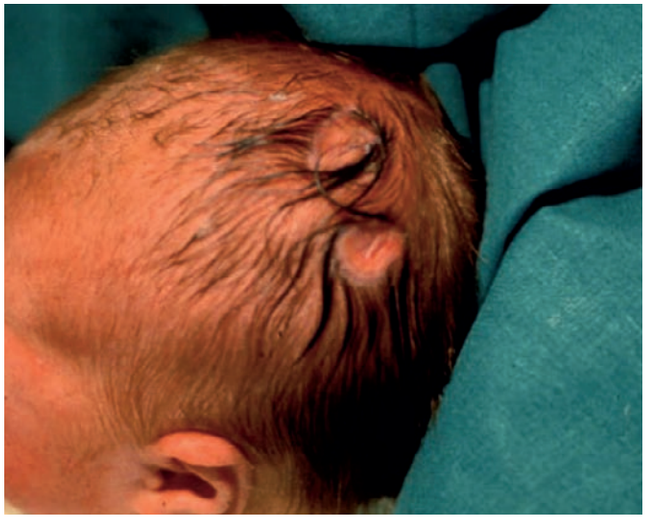


Figure 11.5
Meningocele surrounded by a tuft of long unruly hair.

Simple transitory hypotrichosis

High premature infants often show a physiologic hypotrichosis which spontaneously regresses in a few months.

Occipital neonatal alopecia

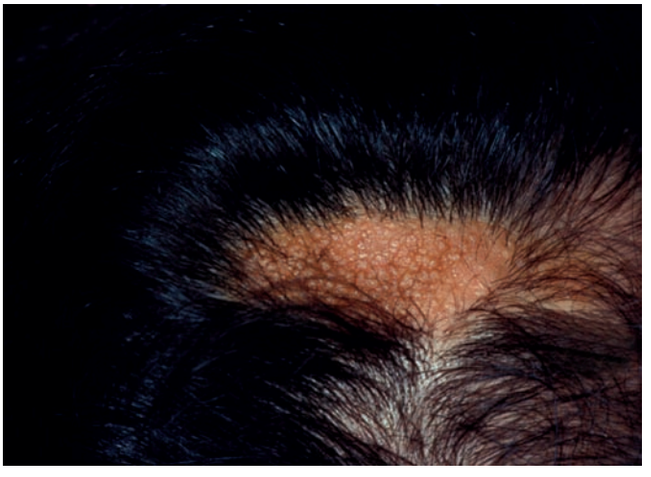
Occipital neonatal alopecia becomes evident by the third month of life (Figure 11.4a,b) and is often wrongly attributed to the friction on the pillow. It actually represents loss of telogen hairs of the intrauterine life. The condition regresses spontaneously.

Meningocele

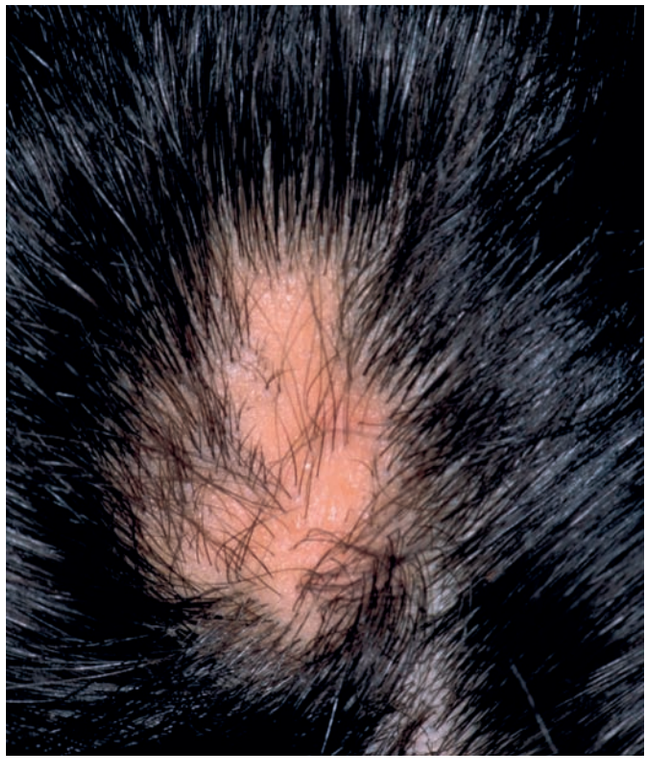
A congenital patch of alopecia or a tuft of unruly hair may be a sign of meningocele (Figure 11.5). There is no topographic correlation between site of hair loss and localization of the anomalous nervous tissue.

Sebaceous nevi

Sebaceous nevus is a common congenital lesion that is generally first noticed at birth. Before puberty, sebaceous nevus appears as a well demarcated plaque of yellowish papules with alopecia (Figure 11.6). After puberty the patch becomes verrucoid and micronodular and changes in color from yellow to dark-brown (Figures 11.7, 11.8). Although tumors may arise in nevus sebaceous, these are



Figures 11.6
Sebaceous nevus.



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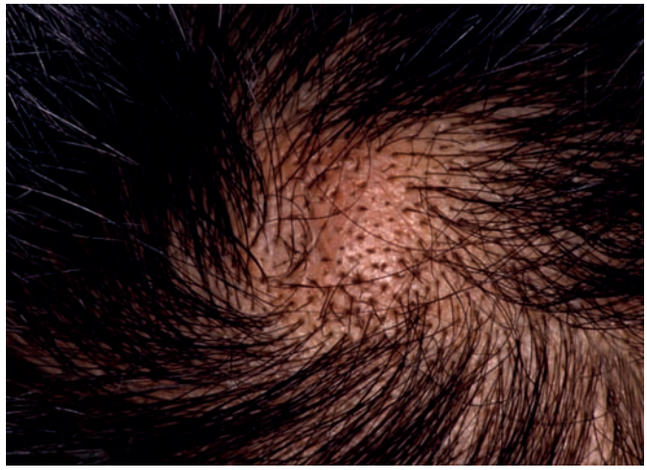


11.8

Figures 11.7, 11.8
Sebaceous nevus.

benign in most cases (Figure 11.9a,b). Prophylactic excision is not recommended.

Aplasia cutis congenita

The condition (Table 11.3) is rare, affecting 3/10 000 newborns. The scalp is involved in most cases with a

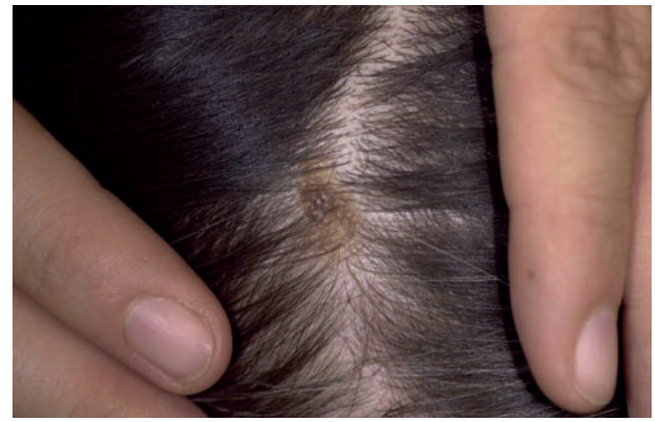


Figure 11.9a
Syringocystoadenoma papilliferum arising from a sebaceous nevus.

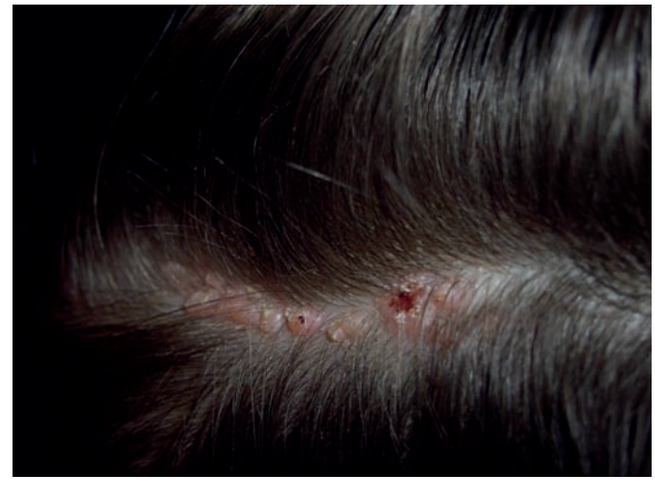


Figure 11.9b
Syringocystoadenoma papilliferum arising from a linear sebaceous nevus.

Table 11.3 Conditions associated with aplasia cutis congenita

Adams–Olivier syndrome
Dystrophic epidermolysis bullosa
Exposure to maternal methimazole
Focal dermal hypoplasia of Goltz
Intrauterine herpes simplex infection
Intrauterine varicella
Johanson–Blizard syndrome
Junctional epidermolysis bullosa
4p syndrome
Scalp ear nipple syndrome
Trisomy 13

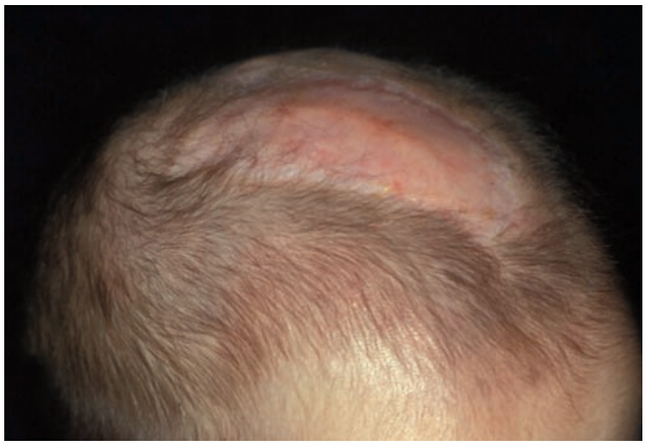
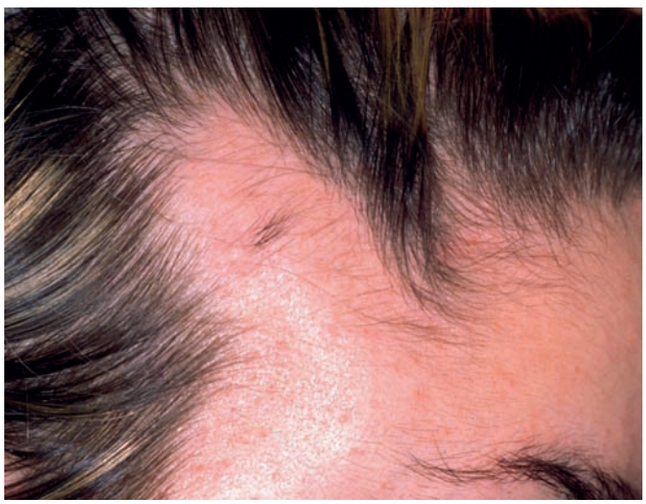


Figure 11.10
Aplasia cutis congenita.

patch of variable size of hair loss, characterized by atrophic skin (Figure 11.10). A defect in the underlying bone may be associated. Proposed etiologies include: infection, vascular malformations, traumatic, teratogenic and genetic factors. Differential diagnosis: sebaceous nevus, scalp hemangioma.

Congenital triangular alopecia

This condition is not rare and is usually diagnosed in childhood. A triangular or oval patch of alopecia involves the temporal region (Figures 11.11–11.14). The area may show vellus hair and remains stable in time. The condition may be bilateral.



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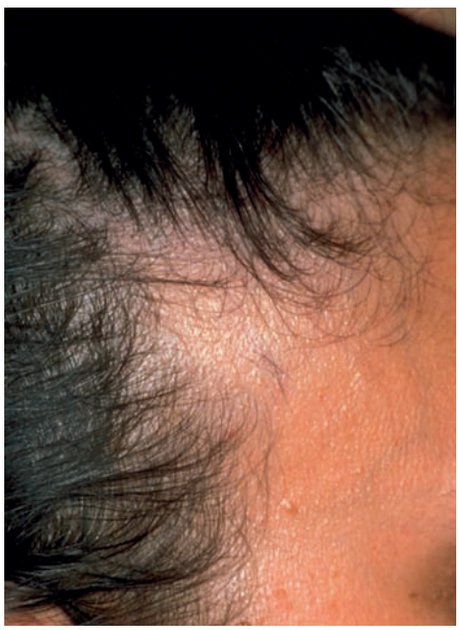

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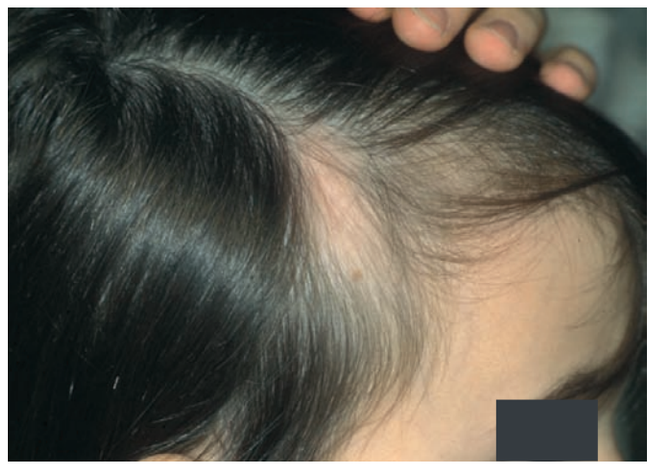

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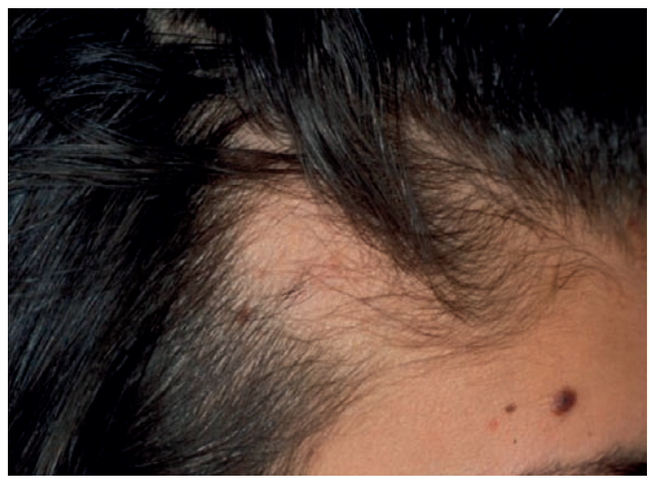


11.14

Figures 11.11–11.14
Congenital triangular alopecia.

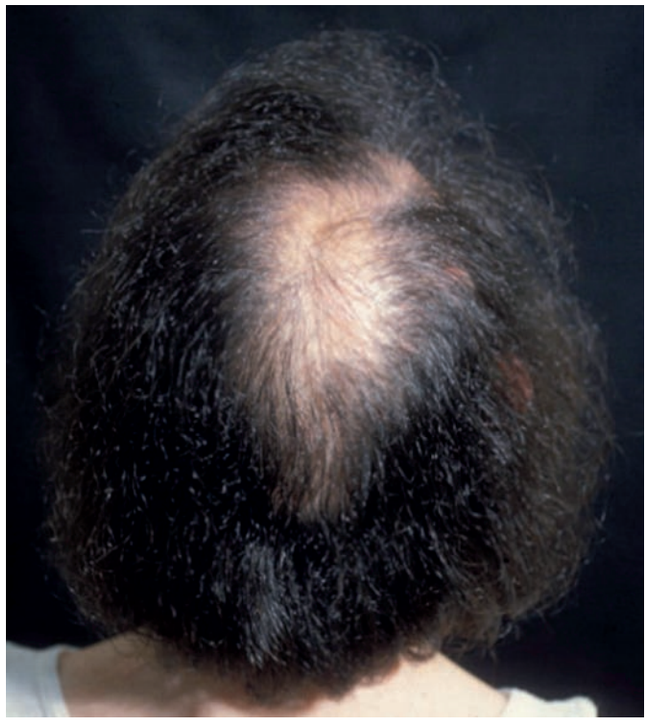


Figure 11.15
Epidermolysis bullosa.

Incontinentia pigmenti (MIM 308300)

Incontinentia pigmenti is a rare X-linked dominant disorder, due to mutation in the NEMO/IKKγ gene that affects mostly females and is usually lethal in males in utero. Alopecia of the vertex occurs in approximately 38% of patients and is usually the outcome of the vesicular stage of incontinentia pigmenti involving the scalp. Linear areas of alopecia following the Blaschko's lines can also occur.

Epidermolysis bullosa

Cicatricial alopecia may be a consequence of bullous lesions of the scalp (Figure 11.15). In generalized benign atrophic epidermolysis bullosa severe and progressive cicatricial alopecia is a predominant feature and characteristically shows a male pattern distribution.

Acrodermatitis enteropathica

Alopecia is a typical feature of acrodermatitis enteropathica and involves scalp, eyelashes and eyebrows. The hair color changes to red during the active phase of the disease.

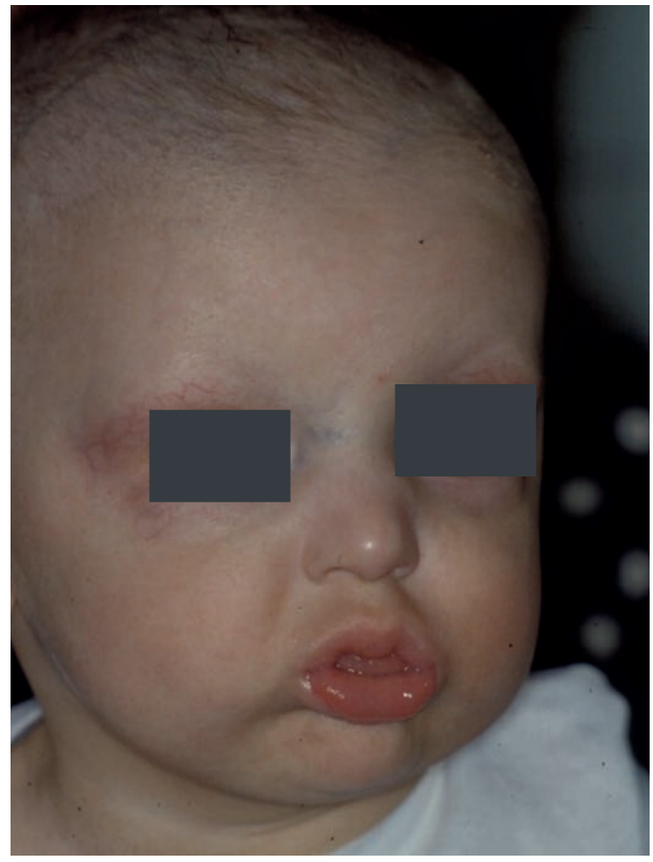


Figure 11.16
Hypoidrotic ectodermal dysplasia.

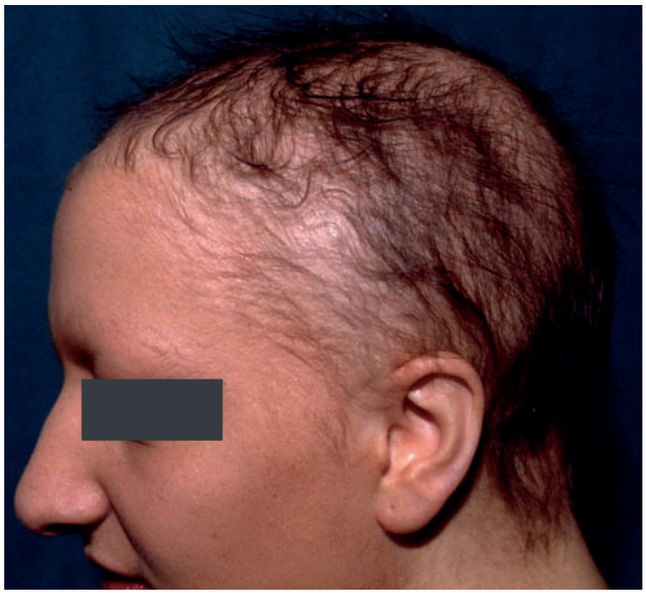
Ectodermal dysplasias

Hypohydrotic ectodermal displasias (MIM 305100; MIM 129490) (Figure 11.16)

These include X-linked and autosomal dominant/recessive forms which have been linked to the ectodysplasin (EDA) gene and to the downless (DL) gene, respectively. These genes have an important role in hair growth initiation. The phenotype appearance includes sparse, fine and blond scalp hair, hypodontia and dry skin with absent or inadequate sweating. Body hair is usually sparse or absent. Affected individuals have a characteristic face with saddle nose and relative frontal bossing. Nails are usually normal.

Hydrotic ectodermal dysplasia – Clouston's syndrome (MIM 129500)

This condition associates alopecia, nail abnormalities and palmoplantar keratoderma (Figures 11.17–11.19). Alopecia is often severe and may be total. When present, scalp hair is wiry, brittle and often pale in color. Body hair may also be affected.



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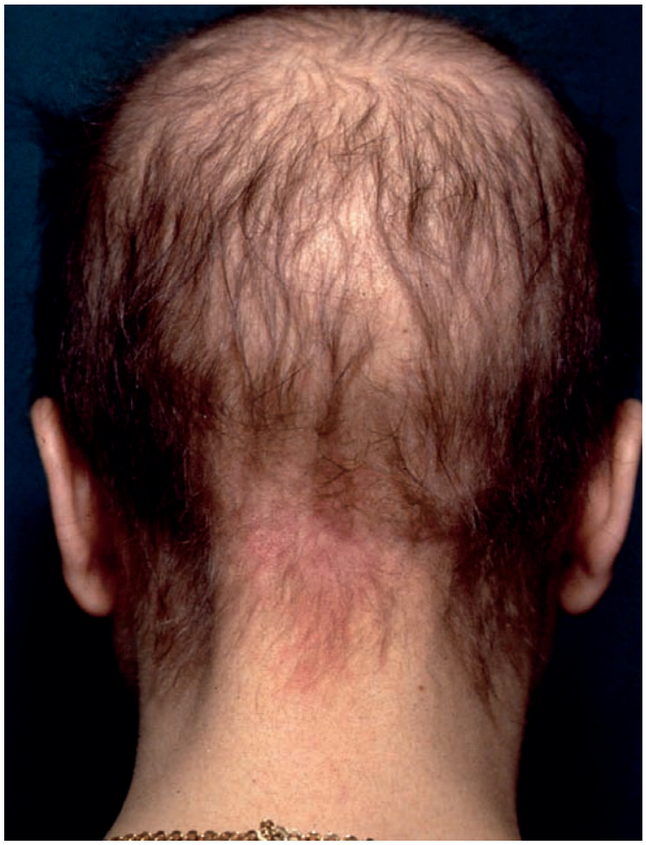


11.18

Figures 11.17, 11.18
Hydrotic ectodermal dysplasia.

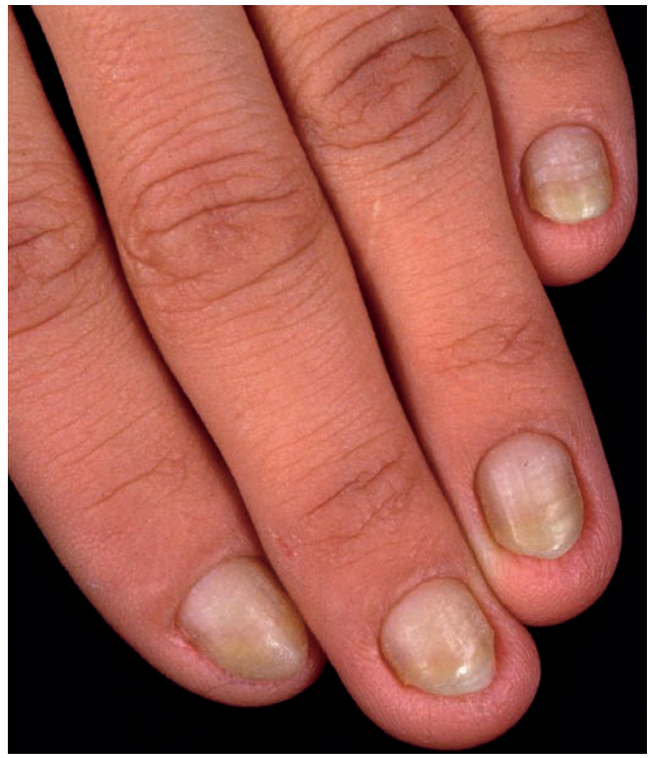


Figure 11.19
Nail abnormalities in hydrotic ectodermal dysplasia.

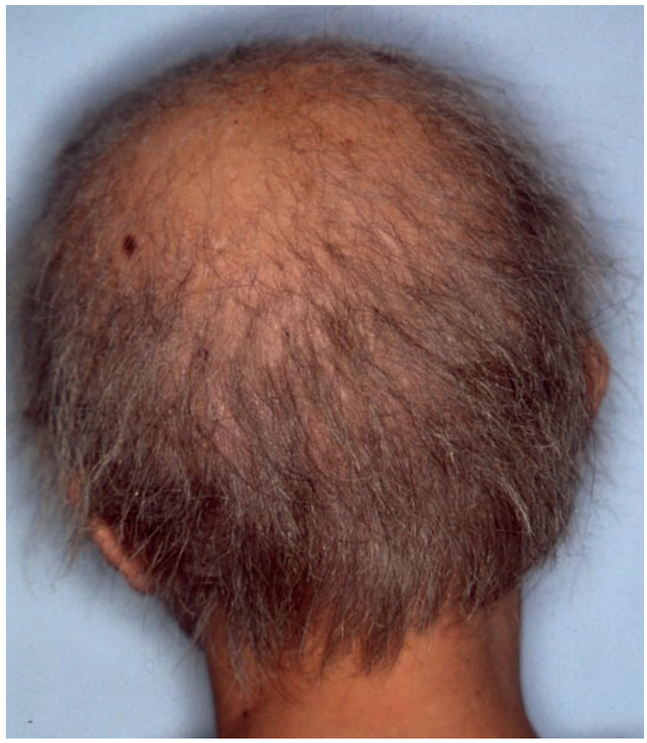
AEC syndrome – Ankyloblepharon, ectodermal defect and cleft-lip and palate (MIM 106260)

In this rare condition scalp erosions and recurrent scalp infections (impetigo, folliculitis) produce cicatricial alopecia (Figures 11.20, 11.21). The hair is usually coarse, wiry and often light in color. The nails are hypoplasic and dystrophic. Many features are similar to Rapp–Hodgkin syndrome (hypohidrotic ectodermal dysplasia and cleft lip and palate (MIM 129400) and some authors believe that they may represent variants of the same entity, although scalp erosions and infections are much rarer in the latter.

Tricho-rhino-phalangeal syndrome (MIM 150230; MIM 190350; MIM 275500)

Type 1 (TRPS 1)

This is an autosomal dominant disorder that has been mapped to chromosome 8q24 (Figure 11.22). Patients



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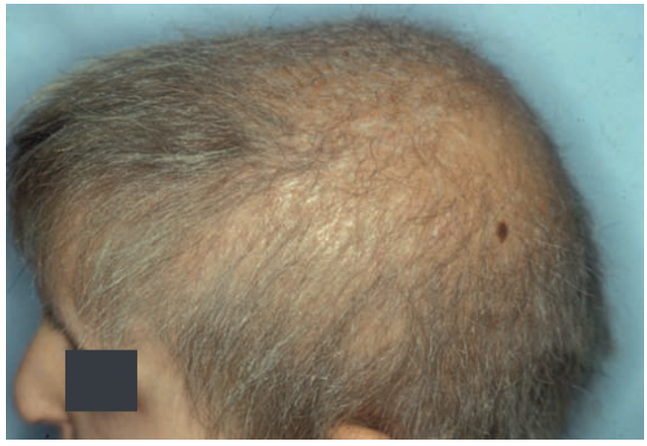


11.21

Figures 11.20, 11.21
Cicatricial alopecia in AEC syndrome.

show fine, sparse, slow-growing scalp hair, clino-brachydactyly of fingers and toes and a typical faces with pear-shaped nose and high philtrum. Nails may be thin and slow-growing.

Type 2 (TRPS 2)

Type 2 is also characterized by neurological and orthopedic abnormalities.

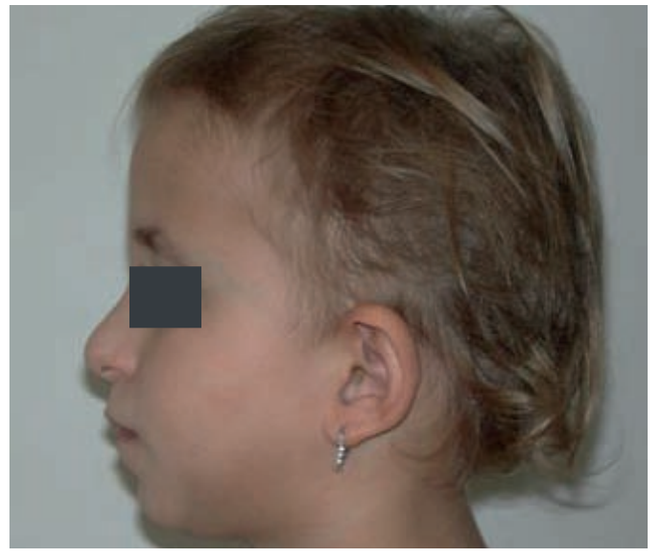


Figure 11.22
Tricho-rhino-phalangeal syndrome type I.

Marie–Unna hypotrichosis (MIM 146550)

This is a rare autosomal dominant disorder characterized by progressive alopecia of the scalp, eyebrows, eyelashes and body hair and structural defects of the hair shaft. The pattern of alopecia is similar to that of male androgenetic alopecia. The hair is coarse, unruly and has been compared to horse hair. Microscopic examination reveals irregular variations in the diameter, irregular twisting and irregular cuticles.

The disease has been recently mapped to chromosome 8p21.

Hereditary hypotrichosis simplex (MIM 146520)

Hereditary hypotrichosis simplex is an autosomal dominant condition that has been mapped to chromosome 6p21.3 (Figure 11.23). Hypotrichosis is evident from childhood and rapidly progresses leading to severe alopecia in early adulthood (Figures 11.24, 11.25). The pattern of hair loss resembles severe male AGA.

Atrichia with papular lesions (MIM 209500; MIM 203655)

Atrichia with papular lesions is an inherited condition caused by mutations in the human hairless gene that

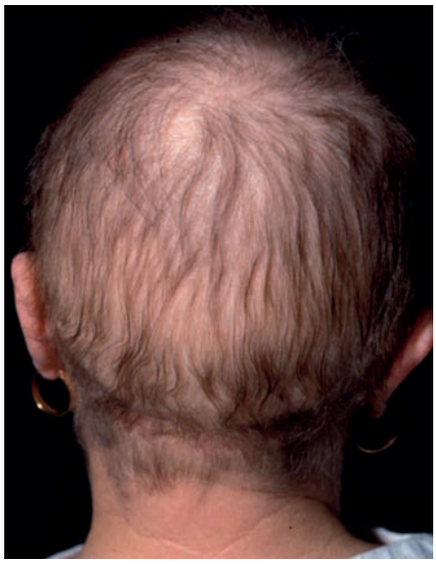


Figure 11.23
Hereditary hypotrichosis simplex.

Figure 11.24
Hereditary hypotrichosis simplex.

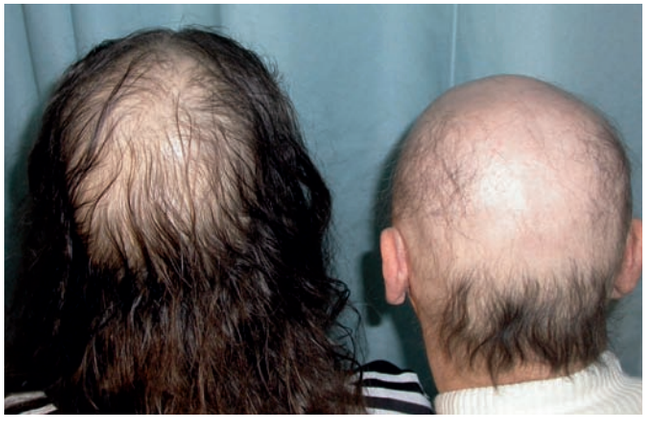
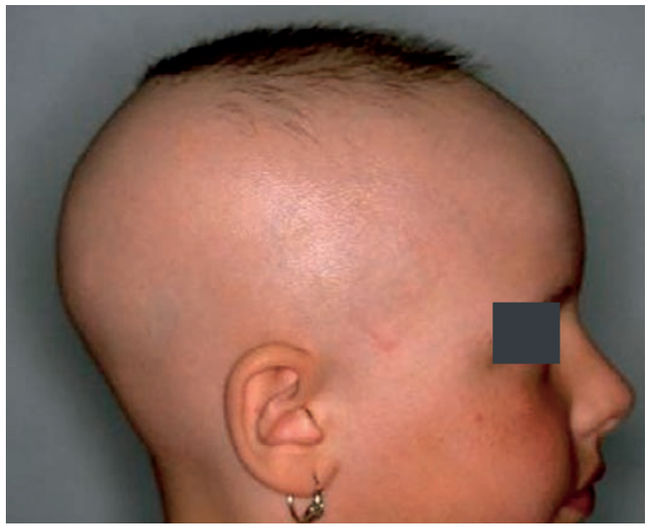


Figure 11.25
Hereditary hypotrichosis simplex: the same family as Figure 11.24 after several years.



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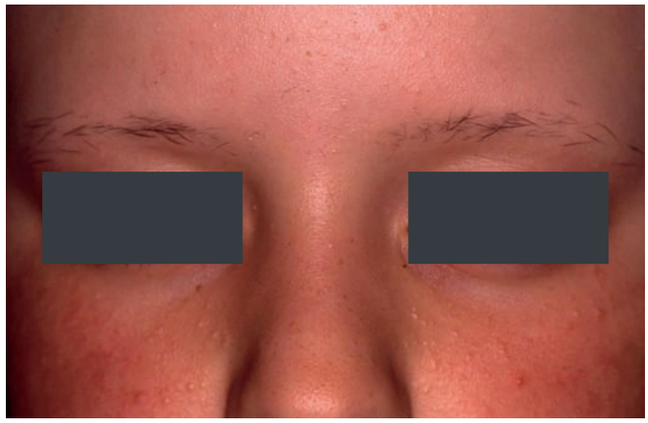


11.27

Figures 11.26, 11.27
Atrichia with papular lesions.

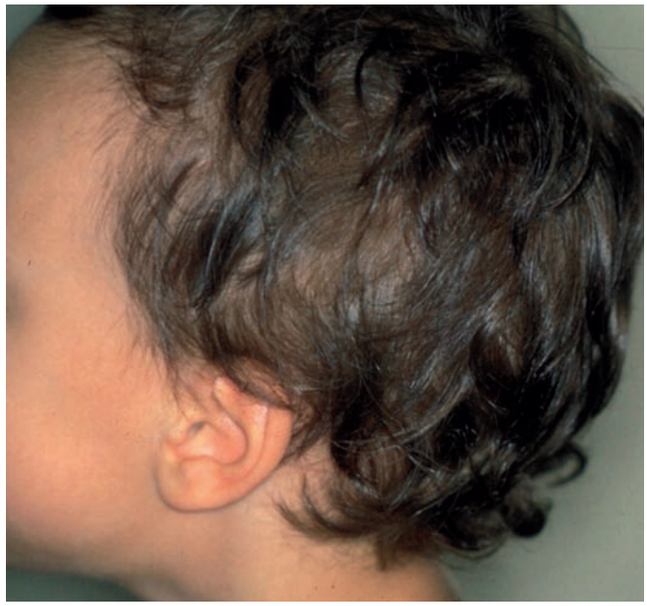
maps on chromosome 8p12 (Figures 11.26, 11.27). Individuals affected by atrichia with papular lesions have normal hair at birth and develop total or subtotal alopecia during the first years of life.

Eyebrows, eyelashes and body hair are also usually lost. Alopecia usually starts from the anterior portion of the scalp and spreads to involve the whole scalp along a fronto-caudal line.

Papular lesions due to follicular cysts develop in the scalp, face and limbs during the first years of life. The pathogenesis of the condition has been linked to premature apoptosis, failure in club hair formation and discontinuation between dermal papilla and hair follicle epithelium.

Short anagen syndrome

In this condition hair are short, sparse, and fine since birth (Figures 11.28, 11.29). Hair shortness is due to a short duration of the anagen phase and not to a slow hair growth. Shortening of anagen has also been reported in the tricho-dental syndrome. Improvement may occur after puberty.



11.28

11.29

Figures 11.28, 11.29
Short anagen syndrome.

Loose anagen hair syndrome (LAHS)

LAHS is a sporadic or familial hair disorder that primarily affects children. The condition is due to a defective anchorage of the hair shaft to the follicle resulting in easily and painless pluckable hair. LAHS is more frequent in females than in males. Mutations in the gene encoding for the companion-layer keratin have been reported in some families with LAHS. The typical patient is a young girl with short, blond hair that does not grow long. Diffuse thinning is frequent in association with irregular bald patches due to traumatic painless extraction of hair tufts. The hair is often dull, unruly or matted. LAHS is usually isolated, but may occur in association with hereditary or developmental disorders (Table 11.4).

Table 11.4 Conditions that may be associated with the LAH syndrome

Alopecia areata
Coloboma
Ectodermal dysplasia, ectrodatyly, cleft lip/palate (EEC) syndrome
HIV infection
Hypoidrotic ectodermal dysplasia
Nail patella syndrome
Noonan syndrome
Tricho-rhino-phalangeal syndrome
Trichotillomania

Three different varieties of LAHS have been divided by Olsen:

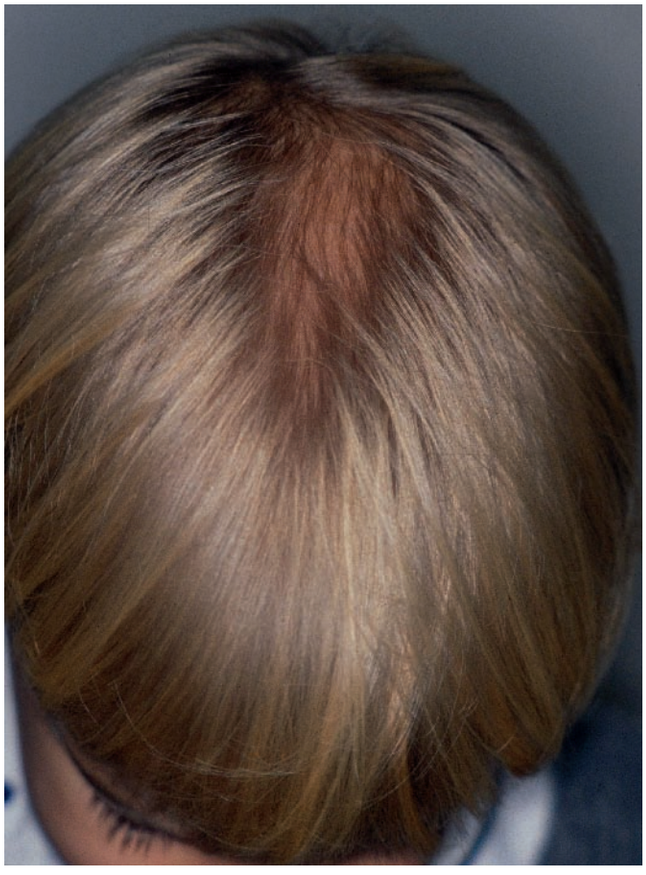
- type 1A; LAHS characterized by decreased hair density (Figures 11.30–11.32).
- type B; LAHS characterized by mainly unruly hair (Figure 11.33).
- type C; LAHS characterized by increased hair shedding (Figure 11.34).

Diagnosis is based on clinical features and presence of LA hair (LAH) at microscopic examination. LAH presents as anagen hair devoid of sheets; its bulb is often misshapen and its proximal portion often shows a ruffled cuticle. Since presence of LAH at the pull test or on trichogram may also occur in controls, the diagnosis of LAHS should be made only if the trichogram shows at least 70% LAH. A negative pull test does not exclude the diagnosis.

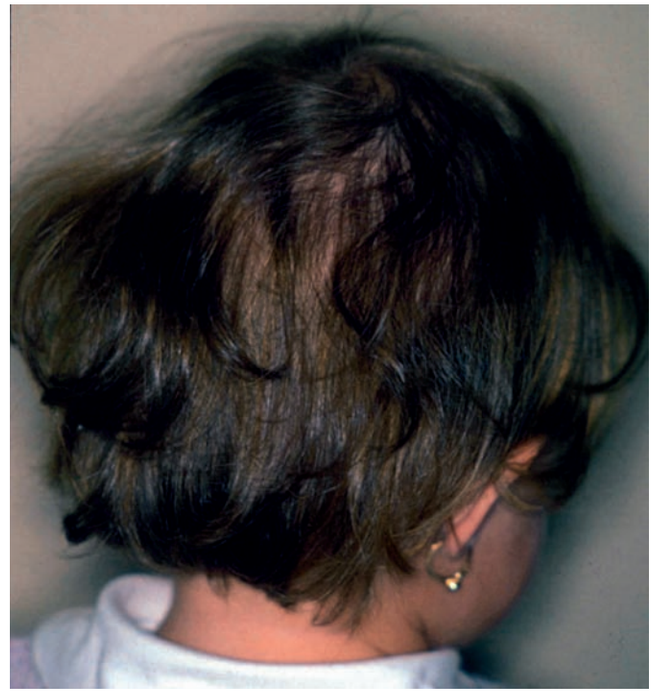
The condition usually improves spontaneously when the child grow up.

Table 11.5 Differential diagnosis between the most common causes of alopecia in children

	Clinical features	*Scalp*	*Diagnostic features*
Alopecia areata	Patches of complete hair loss	Normal, no inflammation or scales	Positive pull test with extraction of dystrophic anagen hairs Exclamation mark hairs Centrifugal spreading
Trichotillomania	Irregular alopecia with short broken hair	Normal, erosions and crusts may be present	Negative pull test Hair of various length
Tinea capitis	Irregular alopecia with short broken hair	Presence of inflammation and scales	Positive KOH and culture
Loose anagen hair syndrome	Diffuse hair thinning with/without irregular patches Short hair	Normal	Hair plucking produces painless extraction of anagen hairs devoid of sheets
Alopecia due to congenital hair shaft disorders	Alopecia with short broken hair on friction areas	Keratotic papules	Typical hair shaft abnormalities at microscopic examination



Figures 11.30
LAHS type A.



Figures 11.31
LAHS type A.

Figures 11.32
LAHS type A.

Figure 11.33
LAHS type B.

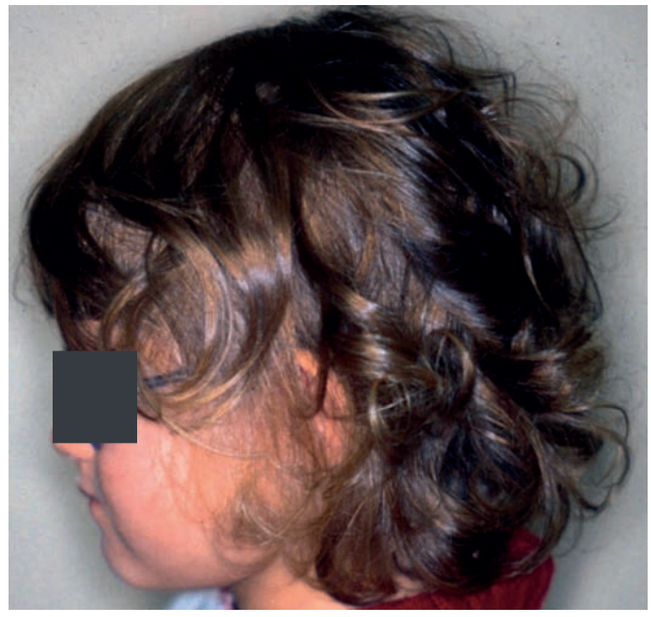


Figure 11.34
LAHS type C.

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12 Hair diseases in the elderly

Aging is associated with hair graying and reduction in the hair density. The number, thickness and growth rate of pigmented, but not white, hairs decrease with aging and the number of telogen hair increases. The hair becomes finer and its quality declines. There is a reduction in the overall capacity of the follicles to produce long hair.

Trichostasis spinulosa

This is a common disorder resulting from retention of bundles of vellus hairs within the follicle.

It is most commonly observed in the face of the elderly as comedo-like lesions, but can also affect young adults where it produces an itching papular eruption of the trunk and upper arms.

Circle hair

Circle hair is observed in elderly obese patients with abundant body hair and appears as perfectly round dark circles interspersed among the normal hair (Figure 12.1). Circle hairs correspond to thin, coiled hair with a subcorneal location. Circle hair mostly occurs on the back and abdomen.

Erosive pustular dermatosis of the scalp

See page 150.

Actinic damage

Multiple actinic keratoses and solar lentigines are very common in the sun-exposed scalp of patients with androgentic alopecia (Figures 12.2–12.4). Squamous cell carcinoma is a possible sequela.

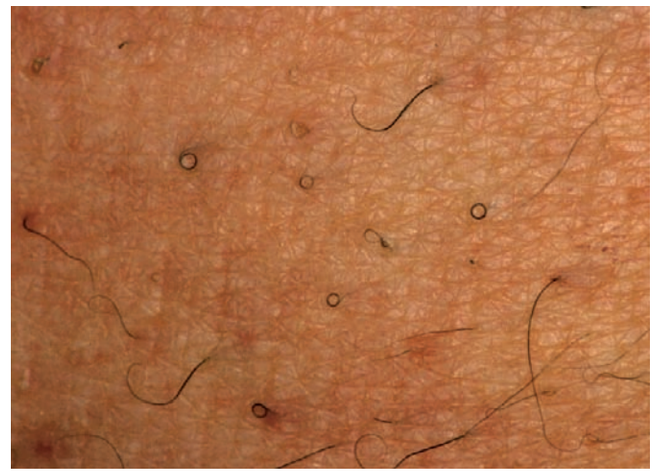
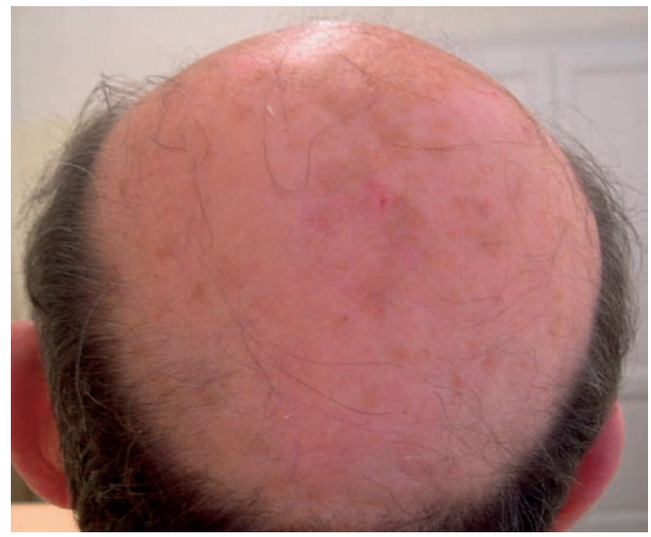
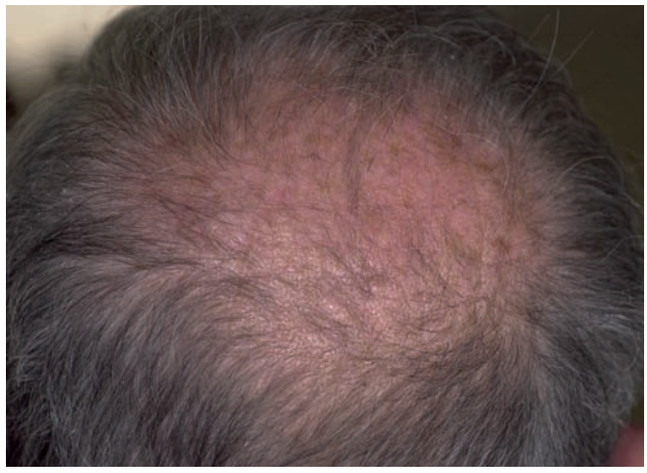


Figure 12.1
Circle hair.



Figures 12.2
Actinic keratoses and solar lentigines in a balding scalp.



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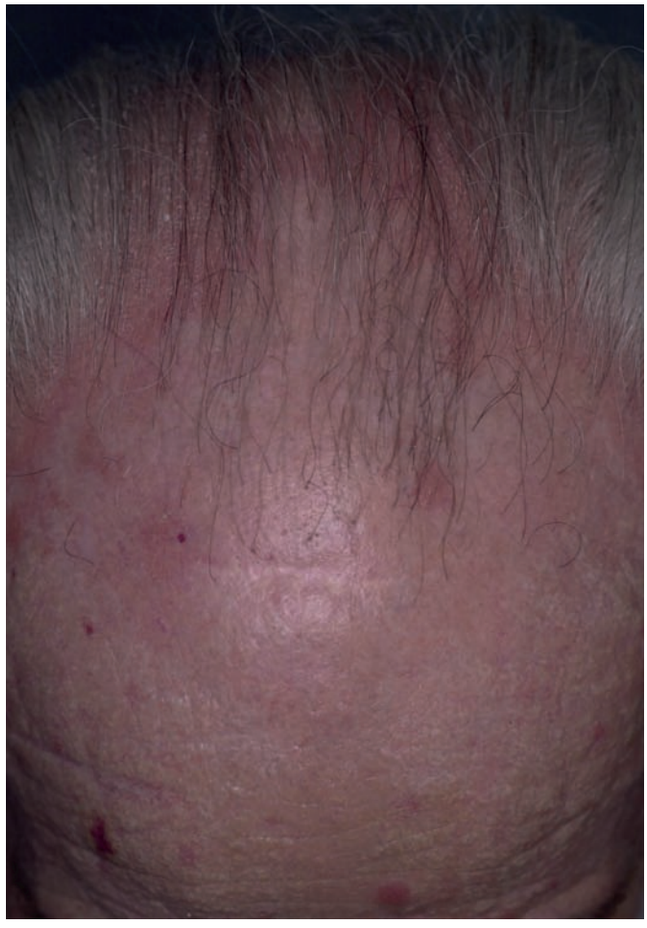

12.4

Figures 12.3, 12.4
Actinic keratoses and solar lentigines in a balding scalp.

Treatment

Since multiple lesione are the rule, topical imiquimod (A) or photodynamic therapy (A) are probably the best option.

Senile alopecia (Figure 12.5)

It occurs in both sexes, after the age of 50 years, as a slowly progressive diffuse hair thinning of the scalp and body hair. Family history for androgenetic alopecia is negative.

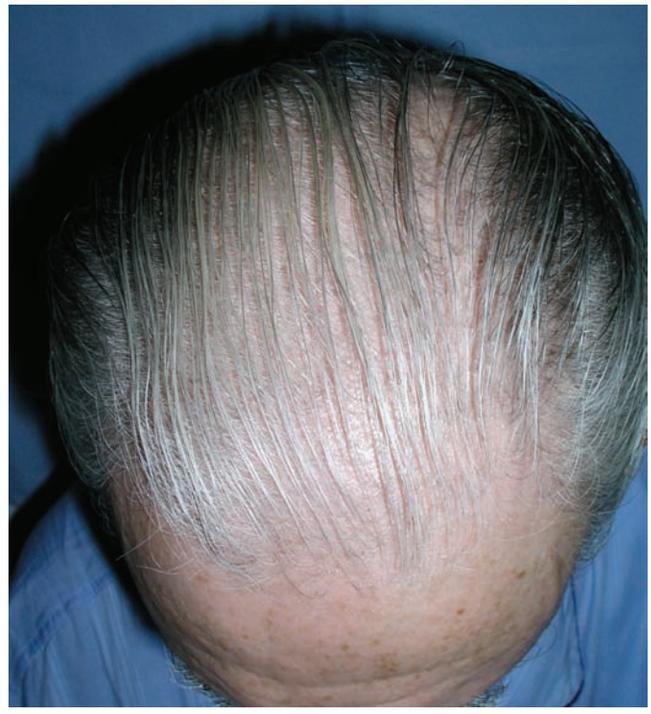

Figure 12.5
Senile alopecia.

Differential diagnosis

Androgenetic alopecia, especially in female patients.

Reference

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13 Traction alopecia

Traction alopecia may be a consequence of accidental traumas, styling procedures, itching dermatosis or of compulsive disorders (Table 13.1).

Table 13.1 Causes of traction alopecia

Mechanical	Accidental traumas	
	Styling	Back combing
		Braiding
		Brushing/combing
		Corn rowing
		Hair band
		Hair clipping
		Hot rollers
		Ponytails
		Rollers
Chemical	Bleaching	
	Permanent waving	
	Straightening	
Pressure	Coma	
	Immobilization	
	Prolonged anesthesia	
Itching dermatosis		
Habit tics		
Compulsive disorders	Trichoteiromania	
	Trichotillomania	

Mechanical traumas

Accidental traumas

Accidental traumas may pull out tufts of hair resulting in patchy alopecia (Figures 13.1, 13.2). Hair extraction is associated with pain and skin injury.

In patients affected by loose anagen hair syndrome, hair extraction is easier and painless and alopecia may develop even with a mild trauma such as hair grasping during playing (see page 98).

Hair styling

Chronic traction due to hair styling produces alopecia of the scalp margins, the pattern of alopecia depending on the styling procedures. The patch usually presents short

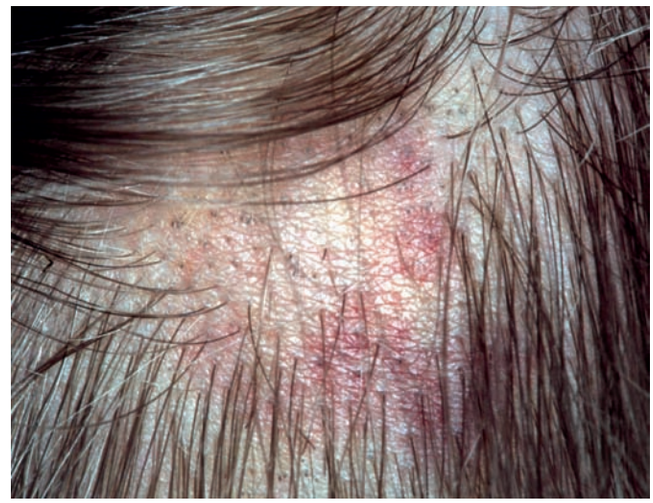


Figure 13.1
Traction alopecia after trauma.

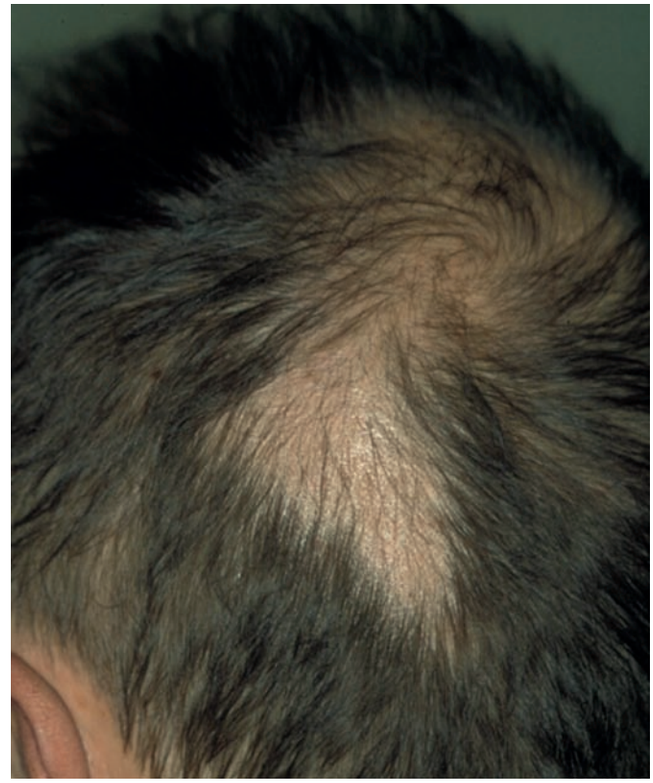
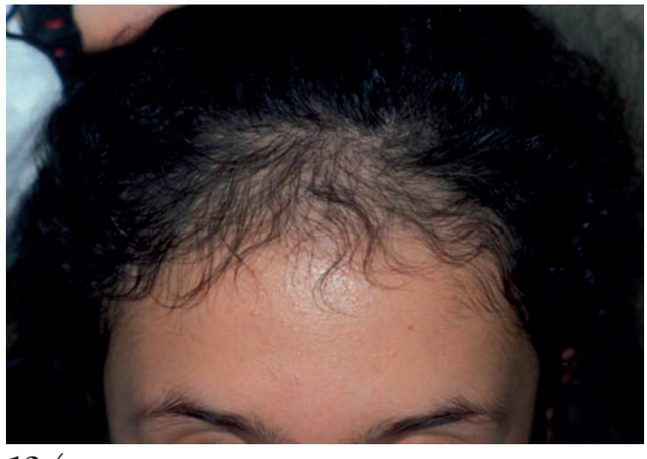


Figure 13.2
Traction alopecia after an aggression.

vellus hairs, broken hairs and dense intermediate hairs at its periphery (Figures 13.3, 13.4). African-Americans are most frequently affected by traction marginal alopecia because of the hair styling (Figure 13.5).

13.3



13.4

13.5

Figures 13.3–13.5
Traction alopecia due to hair styling.

Hair casts are often present in the hairs surrounding the alopecic area (Figure 13.6). Tension headache is very common in women wearing a ponytail (more than 50% of cases).

Figure 13.6
Hair casts.

Friction

Repetitive friction produces hair breakage and patchy hair loss (Figure 13.7a,b). This may be a consequence of scratching, rubbing or even overzealous application of hair lotions. Scalp or body hair may be involved (Table 13.2).

Anterolateral leg alopecia is a common form of alopecia that mostly affects middle-aged and elderly men. The anterior and lateral aspects of the legs show well-circumscribed patches of alopecia.

The etiology of anterior leg alopecia is still unclear, but traumas such as sock and trouser friction and leg crossing probably play a role.

Alopecia of the posterior surface of the legs has been reported after water sliding.

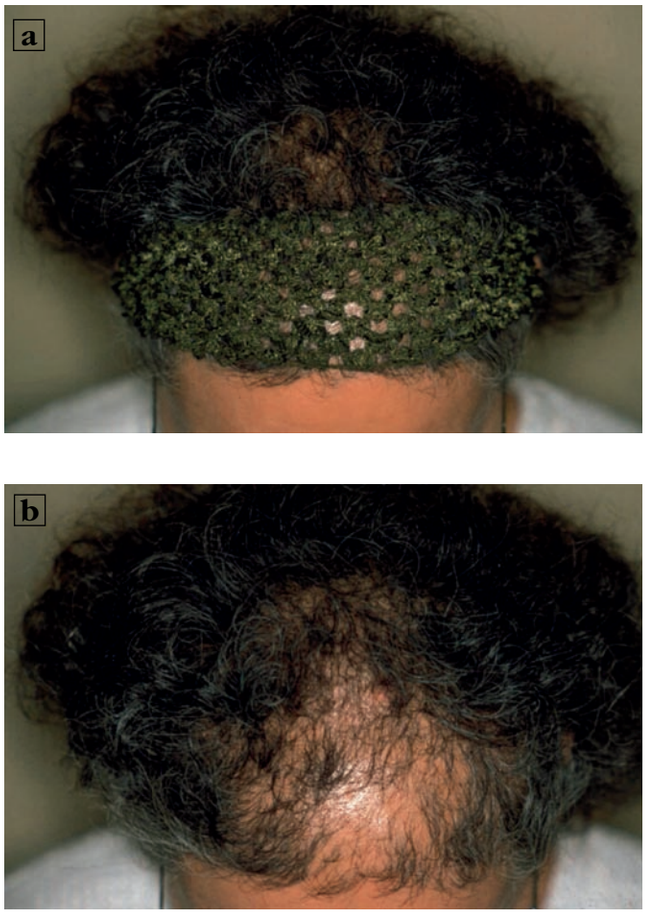


Figure 13.7a,b
Traction alopecia due to prolonged wearing of a hair band.

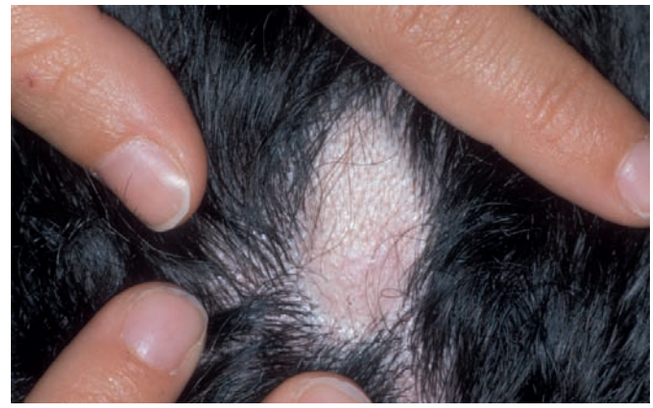
Table 13.2 Friction alopecia

Body area	*Cause*
Anterolateral leg	Sock/trousers friction
	Leg crossing
Posterior leg	Water sliding
Inner thigh	Sitting cross-legged
Abdomen	Tight clothes
	Sleeping habits
Scalp	Scalp massaging
	Break dancing

Traction alopecia can also be a consequence of recreational and sport activities such as break-dancing and gymnastics when rollover or spinning on the head is repeatedly done.

Pressure

A patchy alopecia can develop in the occipital scalp of patients who are immobilized to bed for prolonged periods during surgery because of systemic illness or coma (Figures 13.8–13.10).



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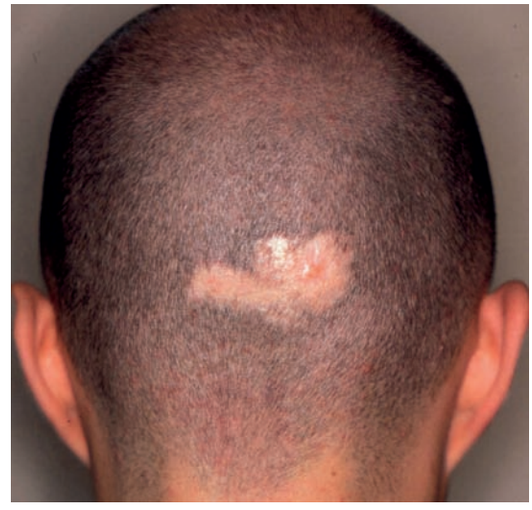

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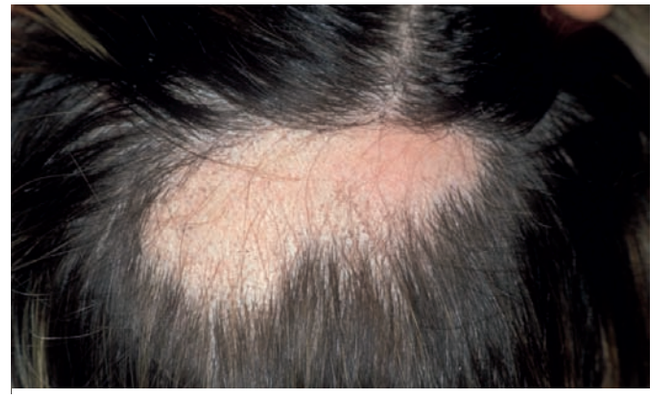
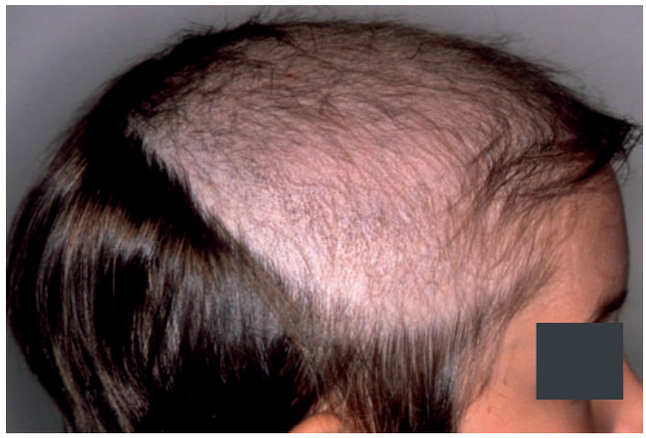


13.10

Figures 13.8–13.10
Pressure alopecia after coma.



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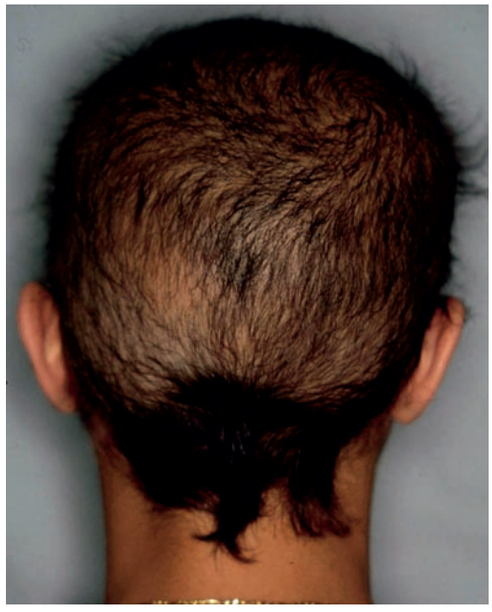

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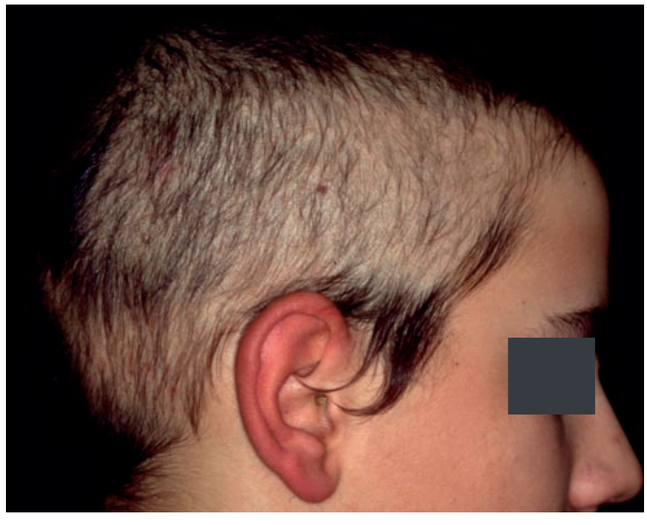


13.13

Figures 13.11–13.16
Trichotillomania.

The alopecia results from ischemia and appears 1–2 weeks after immobilization with loss of dystrophic hairs. The margin of the patch may show exclamation point hairs similar to those of alopecia areata.

Alopecia is usually irreversible since ischemia may induce skin necrosis with permanent follicle destruction.

Tension alopecia

Temporal alopecia is a common sequela of facial lifting. Alopecia, which is usually reversible, results from a traumatic damage to the hair follicles. It may be permanent in 2–3% of patients. Topical 2% minoxidil can effectively prevent this side effect of cervico-facial rhytidectomy.

Compulsive disorders

Trichotillomania

Trichotillomania (Figures 13.11–13.18) is a compulsive disorder that most commonly transitorily affects children, especially girls. Trichotillomania affects 0.5% of the population under the age of 18. In adults, trichotillomania is usually associated with psychiatric disorders and in most cases is chronic.

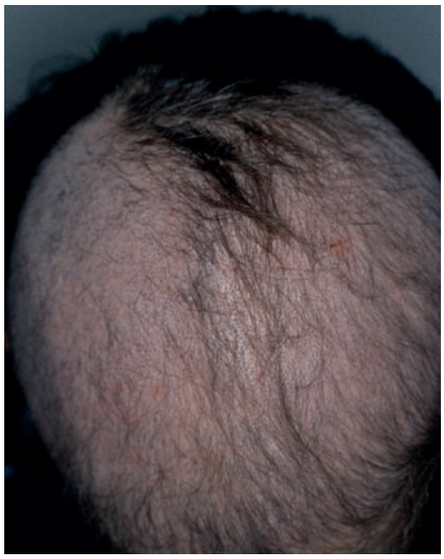
Trichotillomania affects the scalp in the majority of patients, but other terminal hair can be involved, especially the upper eyelashes. The frontal, parietal and occipital scalp are most commonly affected by patches with irregular size and shape, covered with broken hairs of various length. This produces a typical stubbly sensation at the light touch of the hair.

Complete hair loss due to plucking is often seen within the alopecic patch. The skin of the involved area may show folliculitis and escoriations due to scratching.

Patients with trichotillomania frequently do not admit their habit and parents of affected children are often reluctant to accept the diagnosis.

Trichoteiromania

Compulsive rubbing of the hair results in hair shaft fracturing with distal splitting. The affected hairs are broken at different lengths and give the impression of distal white tips. The condition differs from trichotillomania because the pathology does not show trichomalacia and increase of catagen hair.



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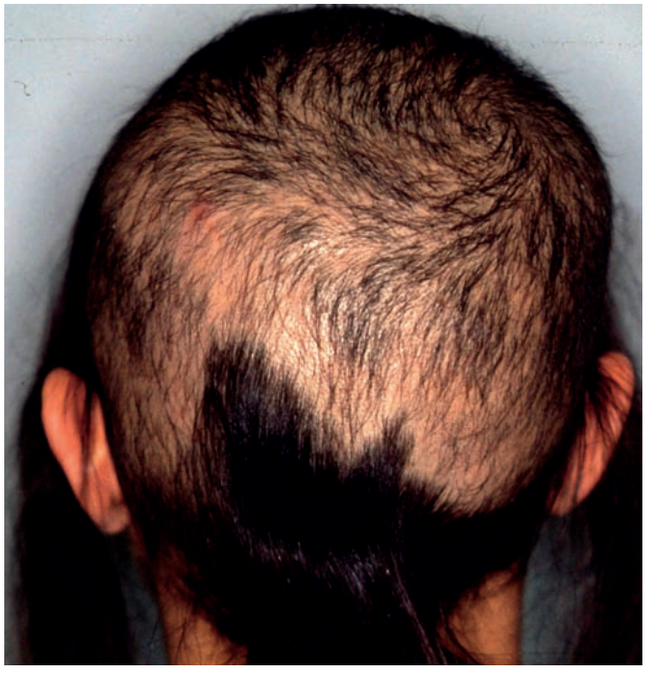

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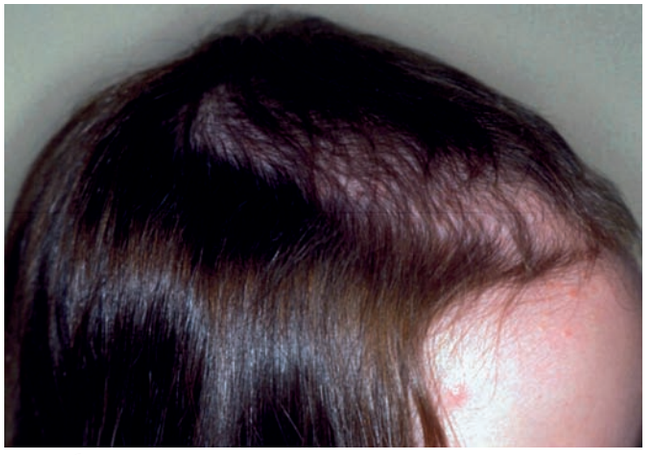

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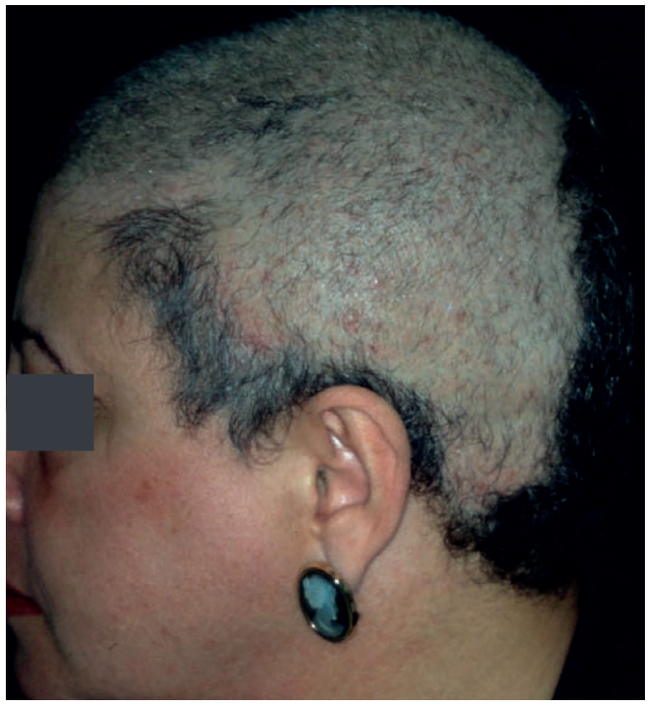

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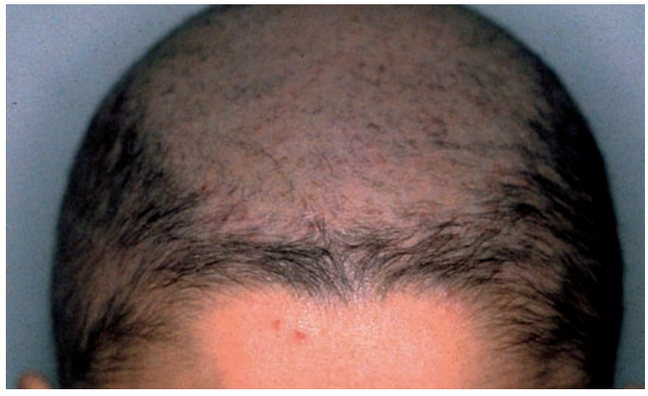

13.18

Figures 13.17, 13.18
Trichotillomania in an adult patient.

Diagnosis

- Clinical features are quite typical and the diagnosis is usually evident.
- A negative pull test is typical of traction alopecia.
- The trichogram shows reduction or absence of telogen roots. The few telogen roots are typically surrounded by the epithelial sac indicating that the hair was still attached to the follicular canal. The hair shafts may show irregular twisting, trichorrhexis nodosa and fractured ends.
- The hair window test can be utilized to confirm the diagnosis trichoteiromania.

13.19

13.20

Figures 13.19, 13.20
Scalp dermatoscopy shows trichomalacia.

- Dermascopy is diagnostic showing coiled fractured short hairs and trichomalacia (Figures 13.19, 13.20).
- Alopecia areata and trichotillomania may occasionally occur in association. Involvement of the lower eyelashes, which are short and difficult to grasp, is uncommon in trichotillomania and suggests alopecia areata.

Management

- Psychiatric referral is always advisable.
- Adults with trichotillomania may benefit from behavior therapy or psychotherapy.
- Pharmacological treatment: clomipramine 100–250 mg/day (B).

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14 Scarring alopecia

A large number of scalp disorders may destroy the hair follicles and result in cicatricial alopecia (Table 14.1). These include diseases that primarily affect the hair follicles as well as diseases that affect the dermis and secondarily cause follicular destruction.

Table 14.1 Causes of cicatricial alopecia

Follicular diseases	Lichen plano pilaris
	Frontal fibrosing alopecia
	Fibrosing alopecia in a pattern distribution
	Discoid lupus erythematosus
	Keratosis follicularis spinulosa decalvans
	Folliculitis decalvans
	Traction alopecia
Dermal fibrosis	Localized scleroderma
	Radiation
	Pemphigoid
	Chemical or physical injuries (Figure 14.45)
	Bacterial or fungal infections

The differential diagnosis between the diseases that cause cicatricial alopecia requires a pathological examination, since the clinical features are usually not diagnostic. The biopsy should be taken from scalp areas that show inflammatory signs, as biopsies taken from atrophic scalp areas only reveal follicular or dermal fibrosis. Direct immunofluorescence is important to distinguish LPP from DLE.

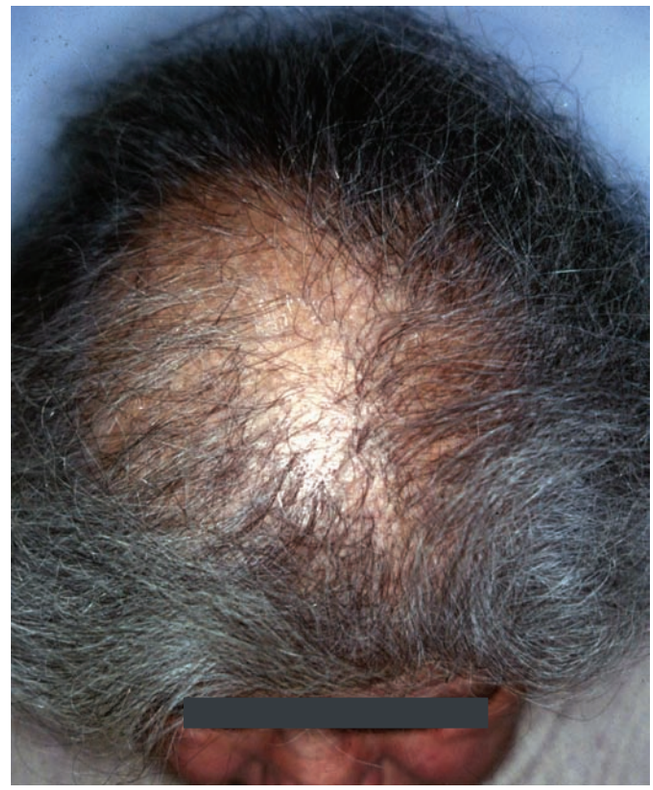


Figure 14.1
Lichen plano pilaris: diffuse hair thinning with irregular areas of cicatricial alopecia.

Lichen plano-pilaris (LPP)

This is the most common cause of cicatricial alopecia. LPP occurs in adults of both sex, but is more common in middle-aged females.

The scalp shows irregular areas of cicatricial alopecia (Figure 14.1). Closer examination of the follicles surrounding the alopecic areas shows perifollicular erythema and follicular plugging (Figures 14.2–14.5). Itching is a main feature and some patients may consult the doctor just because of scalp itching and increased hair loss. LPP may occasionally involve other hair-bearing areas including the axillae and pubis (Figure 14.6).

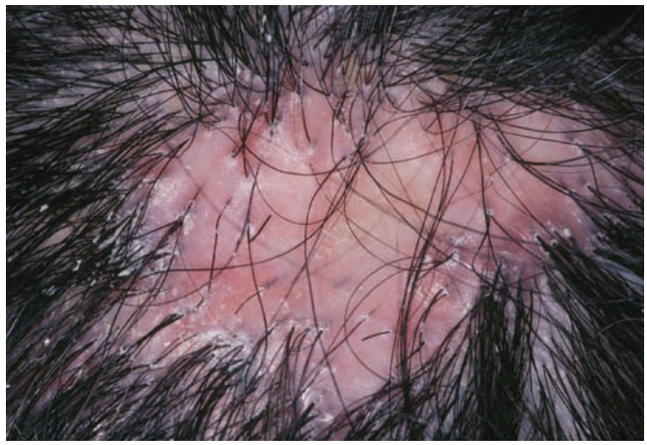
Diagnosis

In the early phases patients may not show patches of hair loss, but just small atrophic areas and the presence of pin-sized follicular papules around the hairs at the periphery (Figure 14.7).

In the late stages inflammatory signs may be completely absent.

The pull test typically shows painless extraction of anagen hair roots with thickened sheaths.

A biopsy for pathology and immunofluorescence is mandatory to confirm the diagnosis and should include some papular lesions.



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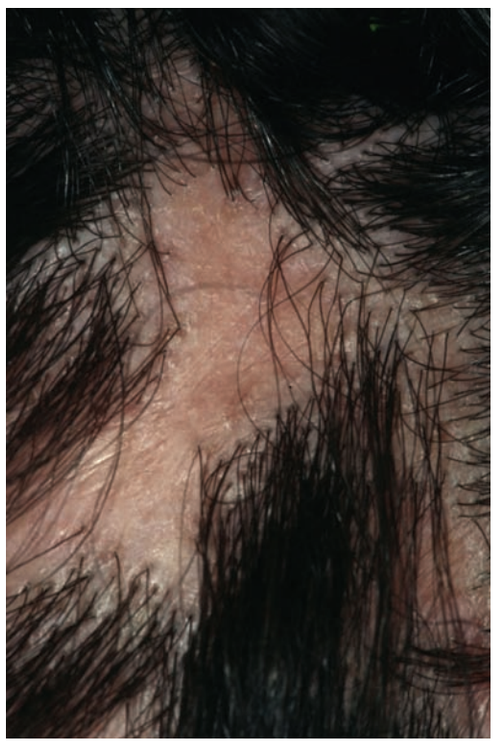

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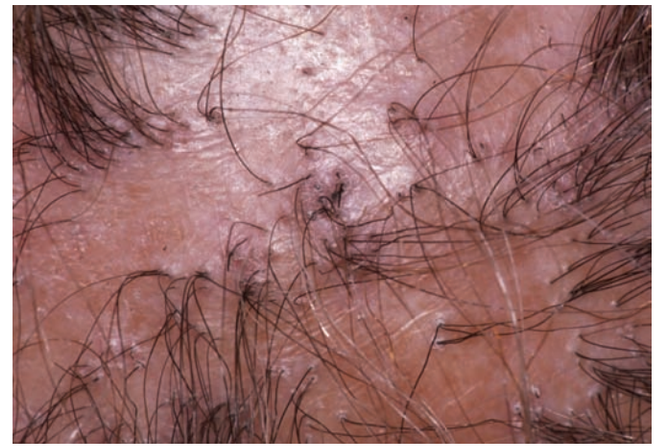

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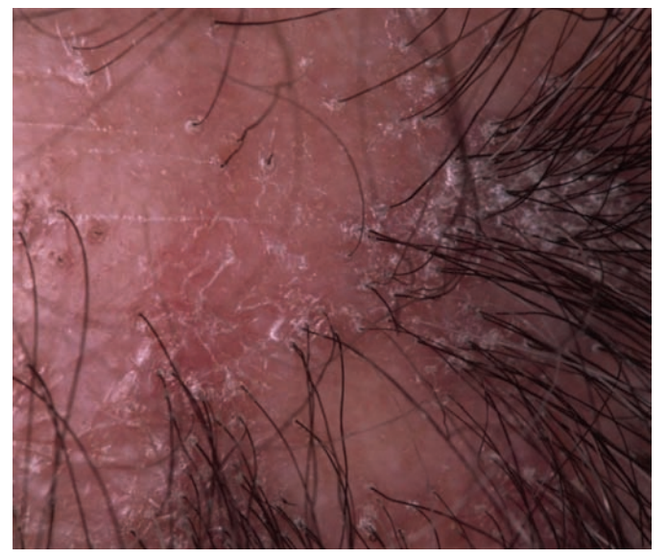


14.5

Figures 14.2–14.5

Lichen plano pilaris: cicatricial alopecia with perifollicular erythema and follicular plugging.

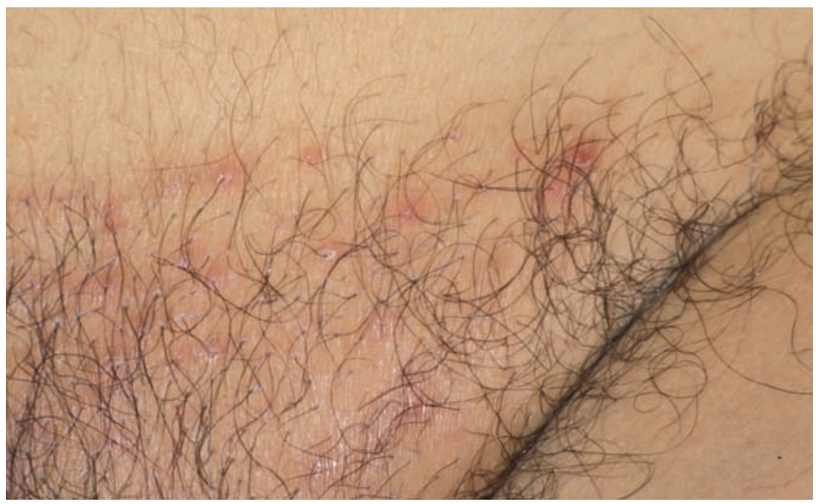


Figure 14.6

Lichen plano pilaris of the pubis.

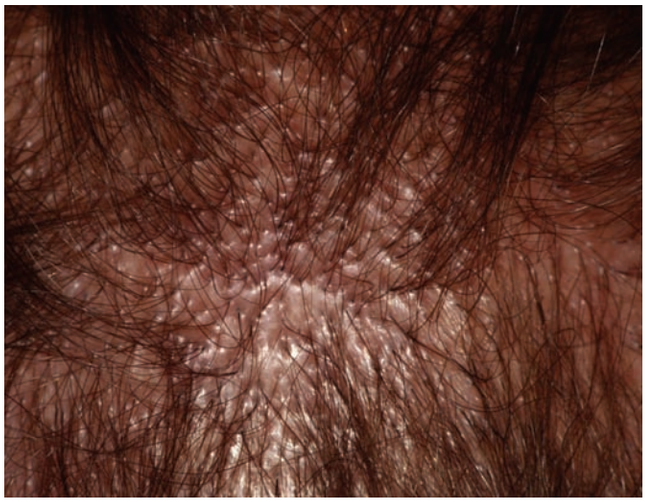


Figure 14.7
Lichen plano pilaris: follicular plugging and small areas of cicatricial alopecia in the early phases of the disease.

Prognosis

LPP responds scarcely to treatment which in most cases slows down, but does not arrest progression of the disease. Severe alopecia however is uncommon (Figure 14.8).

Treatment

- Systemic steroids (C)
  - Triamcinolone acetonide i.m. 0.5 mg/kg/month
  - Oral prednisone 25–50 mg/day
- Topical steroids are of doubtful value

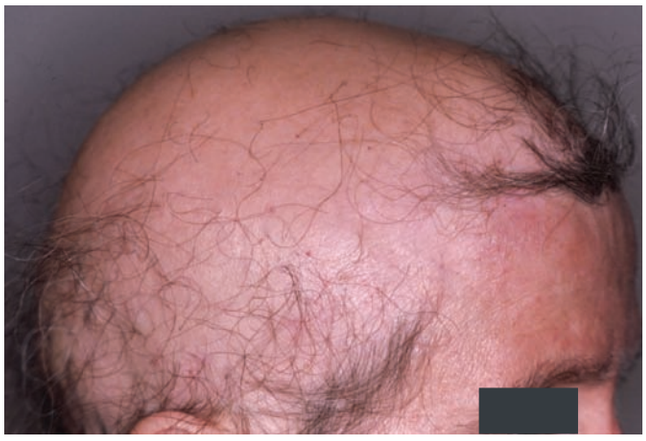


Figure 14.8
Lichen plano pilaris: diffuse scarring alopecia.

- Systemic antimalarials: e.g., cloroquine phosphate 150 mg/day (C), are scarcely effective in our hands
- Oral thalidomide 100 mg/day (E)
- Azathioprine 100 mg/day (E) in association with steroids
- 2% topical minoxidil (E) may prevent fibrosis and is useful in association with systemic steroids.
- Topical tacrolimus (E)

Frontal fibrosing alopecia

This condition typically affects postmenopausal women and it is considered a clinical variant of LPP. Hair loss typically involves the fronto-temporal hairline which recedes a few centimeters (Figures 14.9–14.11). Closer examination of the scalp margin reveals perifollicular erythema and small alopecic patches (Figures 14.12, 14.13). The eyebrows are frequently involved with partial or complete absence of hair (Figure 14.14). Involvement of other body regions such as axillae and groin may rarely occur as well as cicatricial alopecia of other scalp areas.

Diagnosis

Slowly progressive band-like hairline recession with eyebrow thinning.

Prognosis

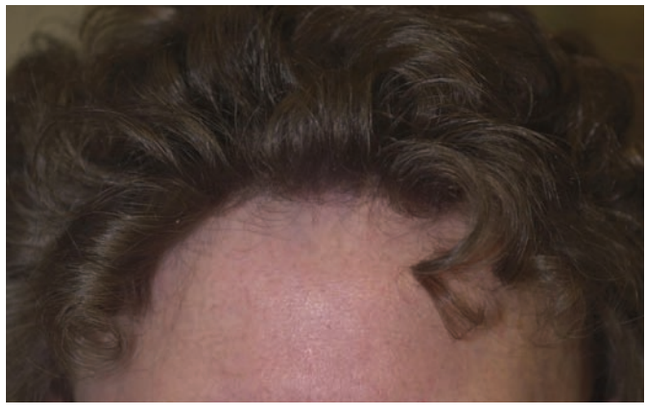
Progression is usually very slow.

Treatment

- Systemic steroids are not effective
- Finasteride 2.5 mg/day (E).

Differential diagnosis

Alopecia areata.



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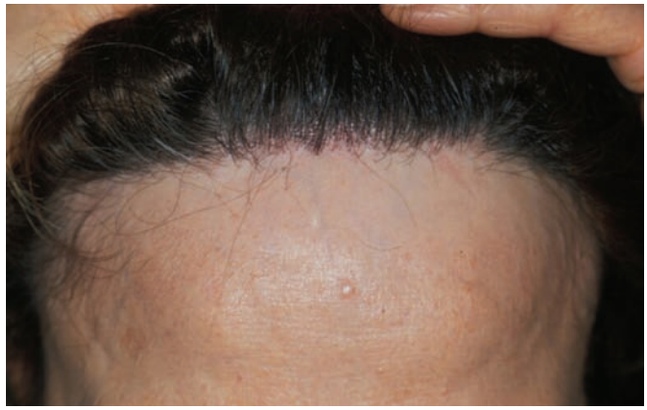

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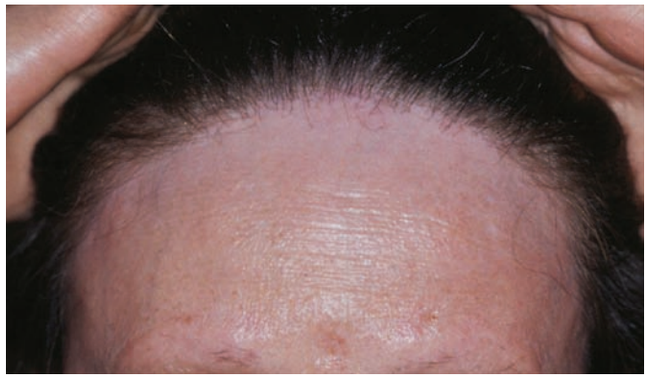
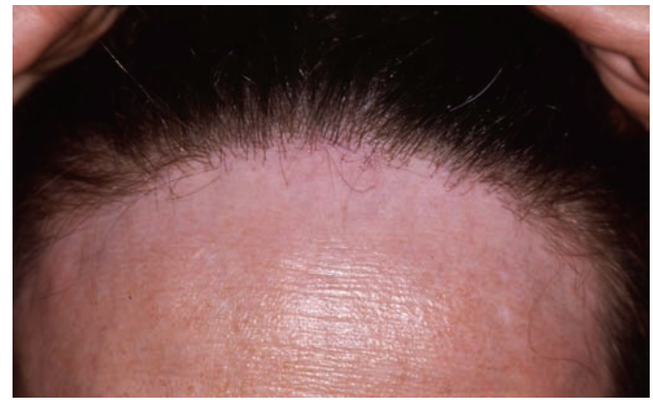


14.11

Figures 14.9–14.11
Frontal fibrosing alopecia: cicatricial alopecia of the frontotemporal hair line.



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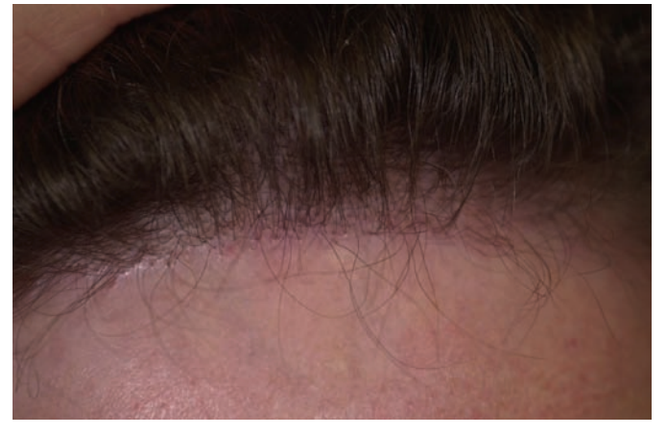


14.13

Figures 14.12, 14.13
Frontal fibrosing alopecia: perifollicular erythema and follicular plugging at the scalp margin.

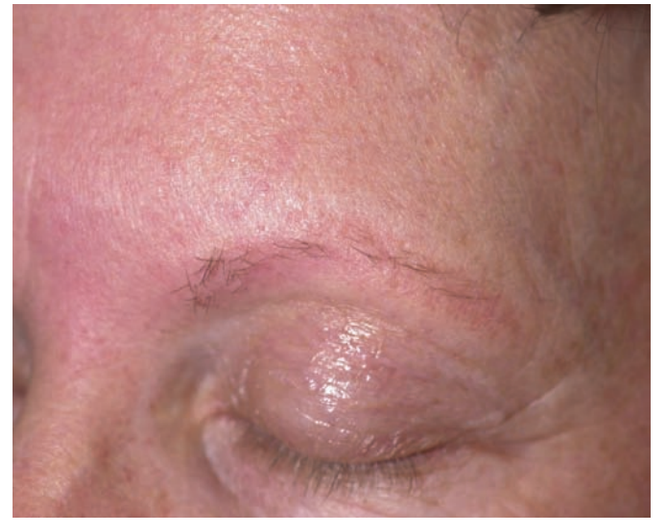


Figure 14.14
Frontal fibrosing alopecia: partial absence of the eyebrows.

Fibrosing alopecia in a pattern distribution

This is a variety of lichen planus pilaris affecting the androgen-dependent areas of the scalp.

The pathology shows selective involvement of miniaturized follicles. Women are most commonly affected. Patients show thinning of the centroparietal scalp hair associated with inflammatory follicular lesions of the surrounding hair and cicatricial alopecia. Frontal fibrosing alopecia may be associated. Itching and pain are complained of by half of patients.

Diagnosis

Scarring alopecia is localized to the androgen-dependent scalp.

Treatment

- Finasteride 1 mg/day (E)
- Oral antiandrogens (E): cyproterone acetate 10 mg as a single treatment.

Graham–Little syndrome

This rare syndrome is characterized by progressive scarring alopecia, loss of pubic and axillary hair and rapid development of horny follicular papules on the limbs and trunk.

Pseudopelade of Brocq

This entity probably represents a variety of lichen planopilaris and is characterized by irregularly or geometrically shaped hypopigmented patches of cicatricial alopecia. Keratotic papules and follicular plugging are absent. The lesions have been described as 'foot prints in the snow' (Figure 14.15).

Follicular degeneration syndrome

See page 162.

Perifolliculitis capitis abscedens et suffodiens (dissecting folliculitis)

See page 149.

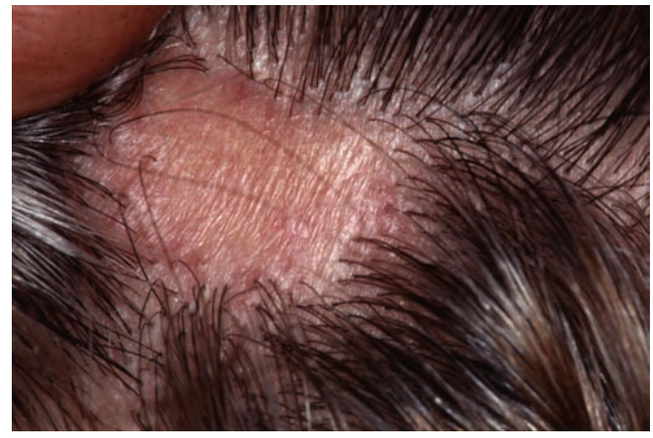
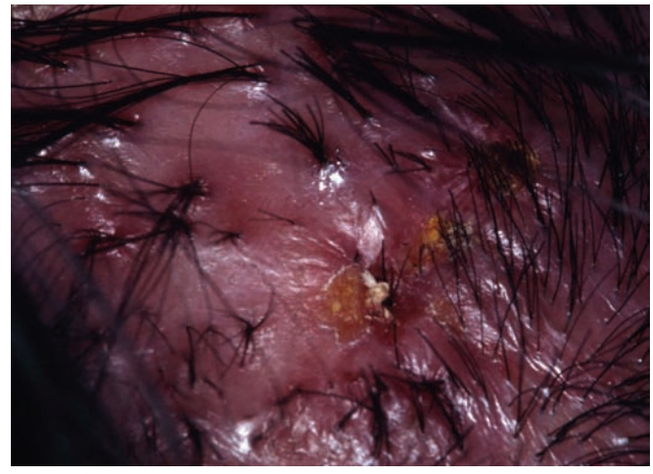


Figure 14.15

Pseudopelade of Brocq. Irregular patches of cicatricial alopecia in the absence of inflammatory signs.



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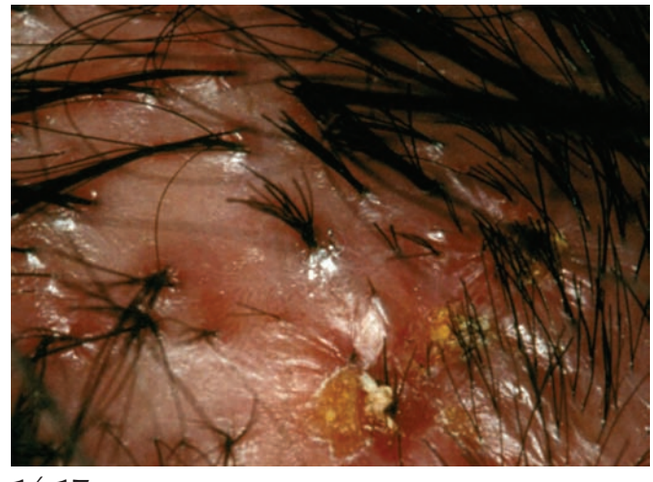

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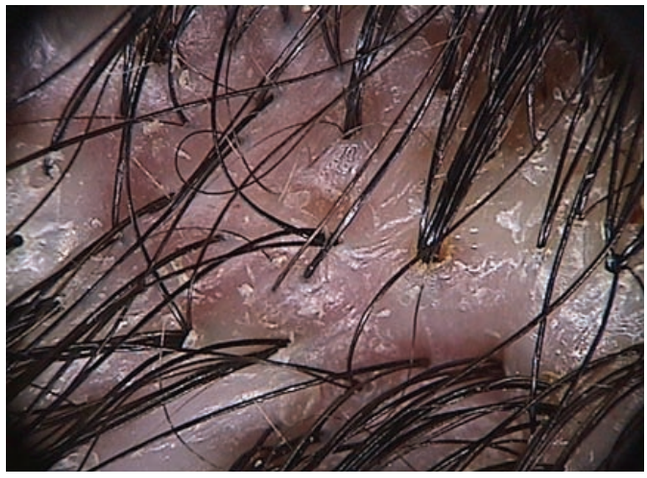
Figures 14.16, 14.17

Tufted folliculitis: several hairs emerge from the same follicular ostium.



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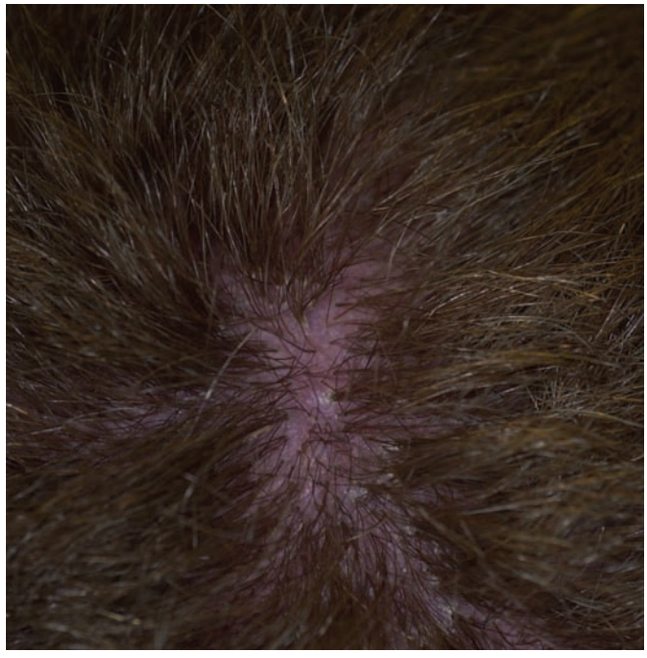
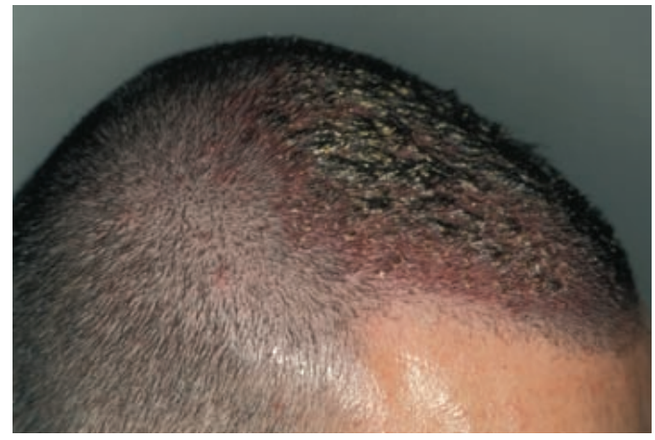


14.19

Figures 14.18, 14.19
Tufted folliculitis: several hairs emerge from the same follicular ostium.

Tufted folliculitis

Although often reported as an individual entity, tufted folliculitis represents the outcome of several forms of cicatricial alopecia. They are more common and severe in folliculitis decalvans. Tufts of five to 15 hairs emerge together from a cicatricial scalp which shows mild to severe inflammatory changes (Figures 14.16–14.19). Tufted folliculitis is caused by clustering of adjacent follicular units as a consequence of dermal fibrosis and retention of telogen hairs.



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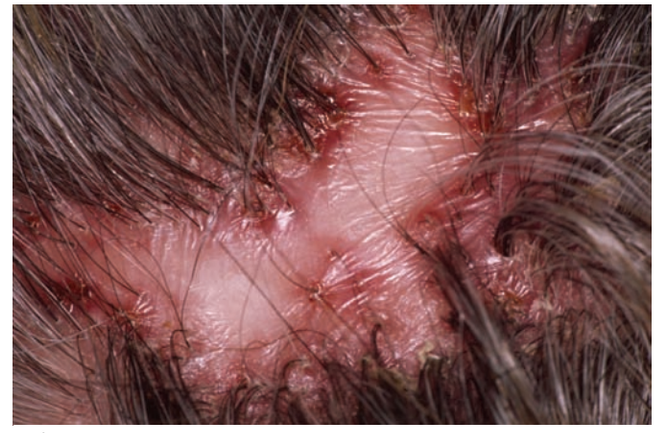

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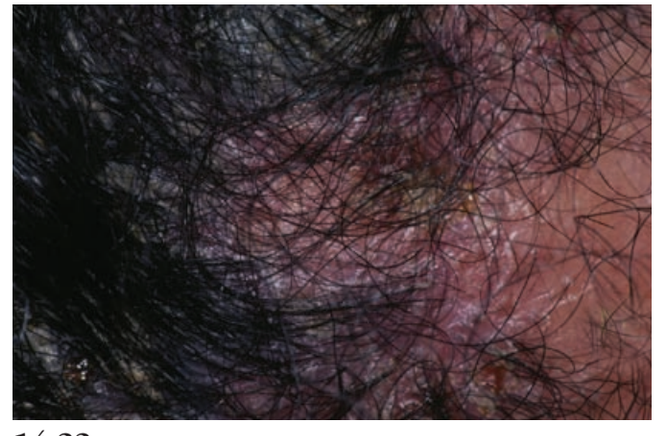

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Figures 14.20–14.22
Folliculitis decalvans: isolated follicular pustules lesions with tufted folliculitis.

Folliculitis decalvans

This term includes a spectrum of scalp disorders characterized by acute inflammatory changes with or without

Figure 14.23
Folliculitis decalvans: isolated follicular pustules lesions with tufted folliculitis.

pustules. The scalp shows patchy or diffuse (Figures 14.20–14.23) areas of papulopustular lesions that often coalesce to form exudative crusted areas. Relapsing inflammatory episodes result in cicatricial alopecia and tufted folliculitis. The condition possibly reflects an abnormal host response to bacterial antigens and *Staphylococcus aureus* can be isolated from the active lesions.

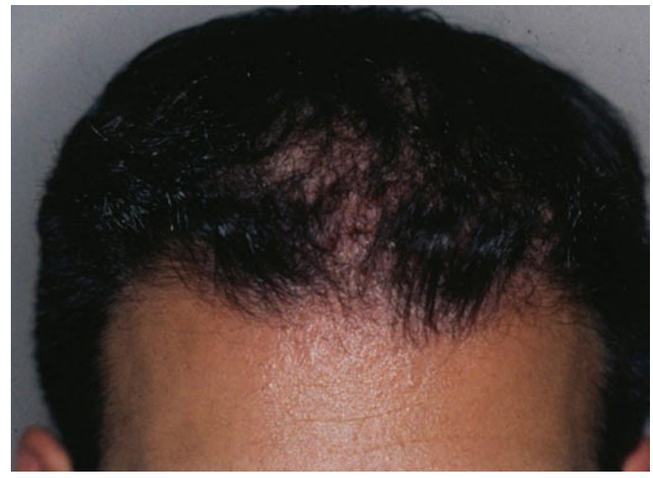
Papulopustular lesions only occur around the hair and the inflammation subsides when the follicles are destroyed and cicatricial alopecia established. Shaving of the scalp may improve the disease. Centrifugal progression commonly occurs (Figures 14.24–14.26).

Prognosis

The disease is progressive and often relapses after interruption of antibiotic treatment. In some cases it may be limited to a circumscribed area and in others may involve a large portion of the scalp.

Treatment

- Scalp shaving
- Oral rifampicin 300 mg/day + clyndamicin 300 mg/day for 10 weeks (C)
- High-potency topical steroids (E)
- Tetracyclines (E)
- Oral/topic fusidic acid (E)
- Isotretinoin 0.5 mg/kg/day (E) is scarcely effective and may even worsen the disease.



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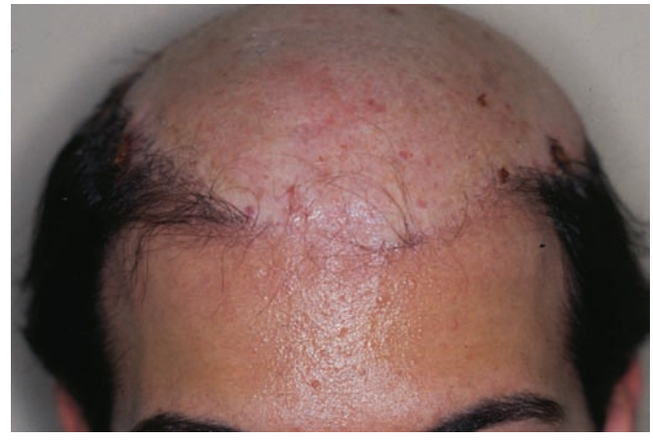

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Figures 14.24, 14.25
Folliculitis decalvans: centrifugal progression of the disease with diffuse cicatricial alopecia.

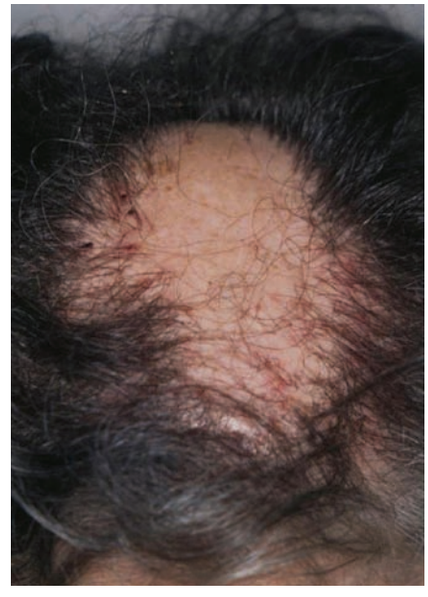


Figure 14.26
Folliculitis decalvans: diffuse scarring alopecia.

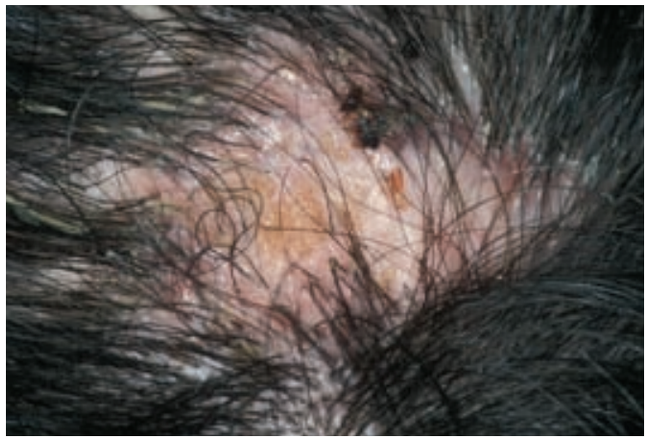


Figure 14.27
Keratosis follicularis spinulosa decalvans: cicatricial alopecia with follicular papules and pustules.

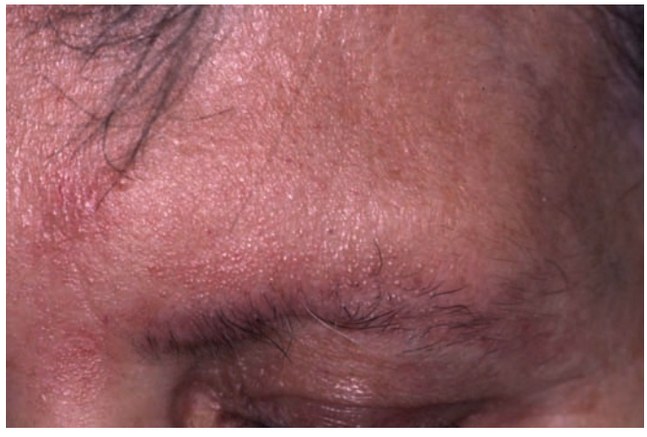


Figure 14.28
Keratosis follicularis spinulosa decalvans: eyebrows alopecia.

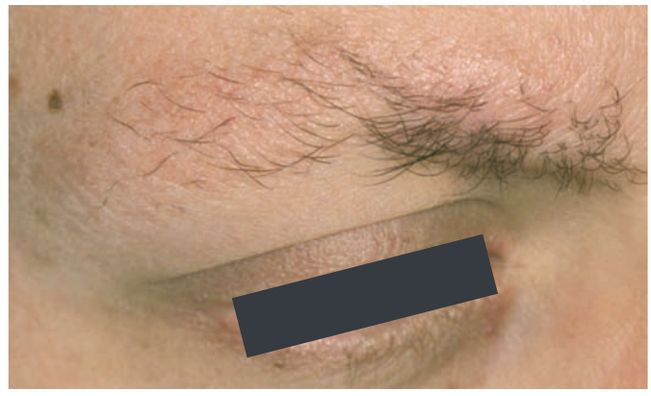


Figure 14.29
Keratosis follicularis spinulosa decalvans: eyebrows alopecia.

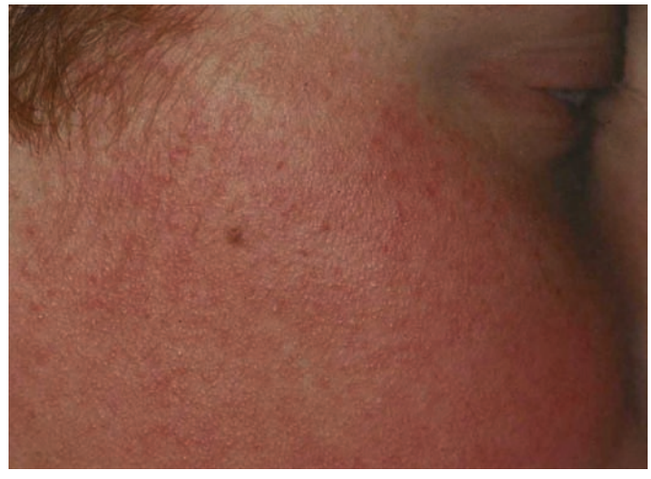


Figure 14.30
Keratosis follicularis spinulosa decalvans: follicular papules on the cheek.

Keratosis follicularis spinulosa decalvans (MIM 308800)

This inherited condition usually becomes evident in infancy. The disease has been mapped to Xp21–p23.

The scalp presents follicular keratotic papules and pustules producing progressive cicatricial alopecia (Figure 14.27). Follicular papules are also evident on the eyebrows (Figures 14.28, 14.29) and cheeks (Figure 14.30). The disease is slowly progressive and can produce severe alopecia.

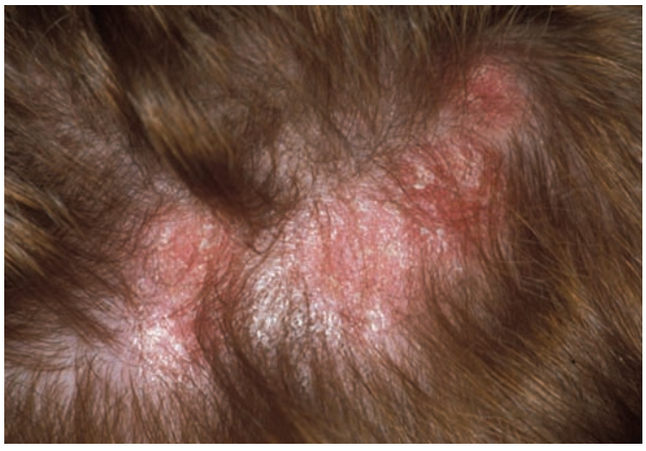
Treatment

All treatments are scarcely effective and the disease is slowly progressive.

- Oral retinoids 0.5 mg/kg/day (C)
- Dapsone 100 mg/day (E)
- Antibiotics (E)

Discoid lupus erythematosus (DLE)

DLE is rare and may be limited to the scalp. Cicatricial alopecia is associated with active inflammatory lesions characterized by erythema, edema, scaling with follicular hyperkeratosis, atrophy and teleangectasia (Figures 14.31–14.35).



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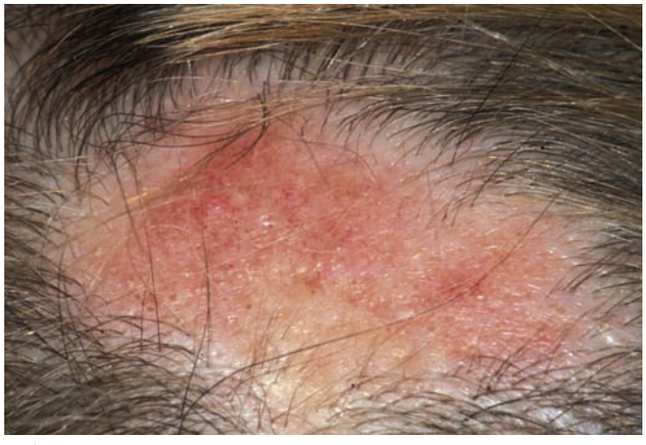

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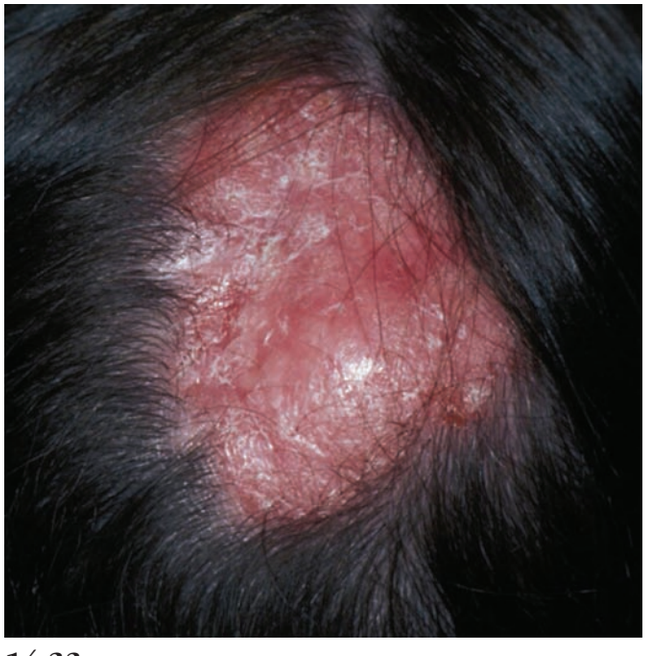

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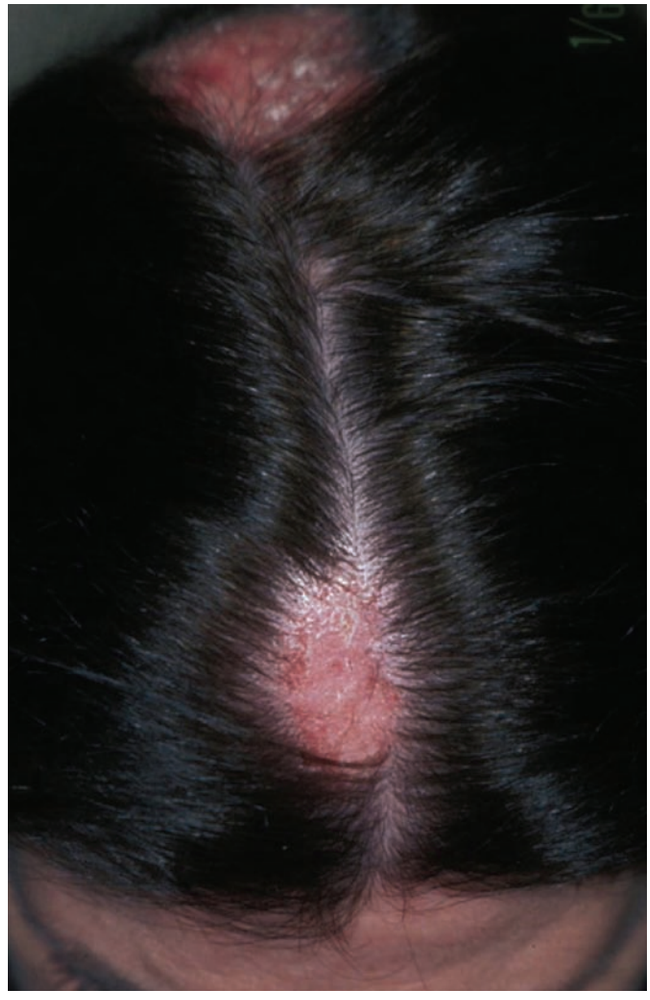

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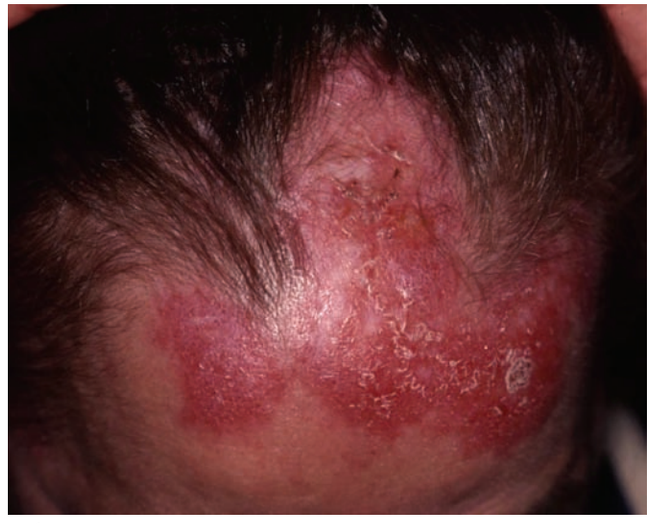


14.35

Figures 14.31–14.35

Discoid lupus erythematosus: erythema, edema and scaling with skin atrophy and telangiectasias.

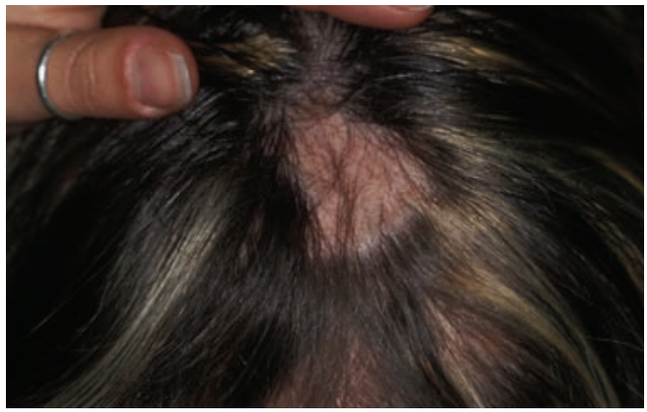


Figure 14.36
Lupus tumidus.

In some cases, however, lupus panniculitis of the scalp usually produces ulceration and scarring.

Hair regrowth may occur in the deep variant of lupus panniculitis which affects the lower portion of the follicles but not the isthmus. Hair loss may be temporary and closely simulate alopecia areata, but scalp tenderness and inflammation are typical.

Tumid DLE may or not produce alopecia (Figure 14.36). The disease usually presents in patches where erythema and edema are prominent features. Scarring is not the rule.

Diagnosis

The pull test from active lesions reveals anagen hair with hyperkeratotic sheaths.

The biopsy for histopathology and immunofluorescence should be taken from an inflamed area.

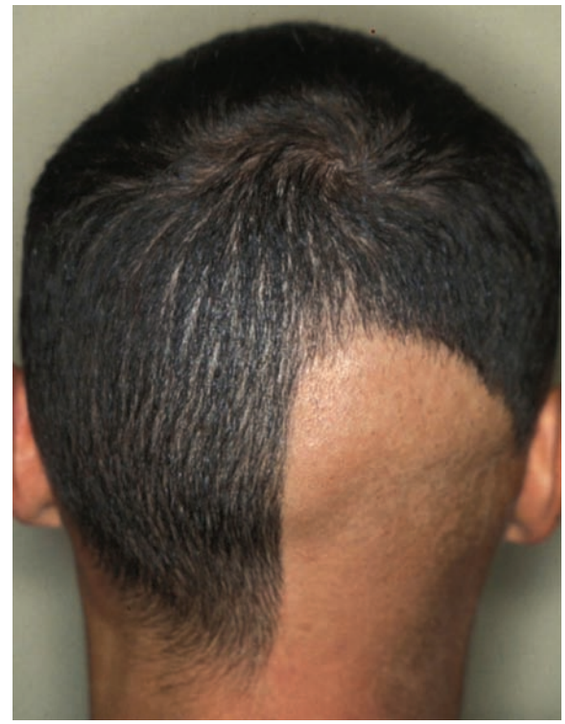
Treatment

Patients should wear a hat to avoid sun exposure.

- Antimalarials (B): hydroxychloroquine 400 mg/day; chloroquine 200 mg/day
- Thalidomide 100–300 mg/day (E)
- Systemic steroids (B): oral prednisone 0.5 mg/kg/day
- High-potency topical steroids: clobetasol propionate 0.05% (B)
- Topical imiquimod (E)
- Topical tacrolimus.

Radiation

Scarring alopecia may be a consequence of irradiation, when the dosage of X-rays exceeds 700 Gy (Figures



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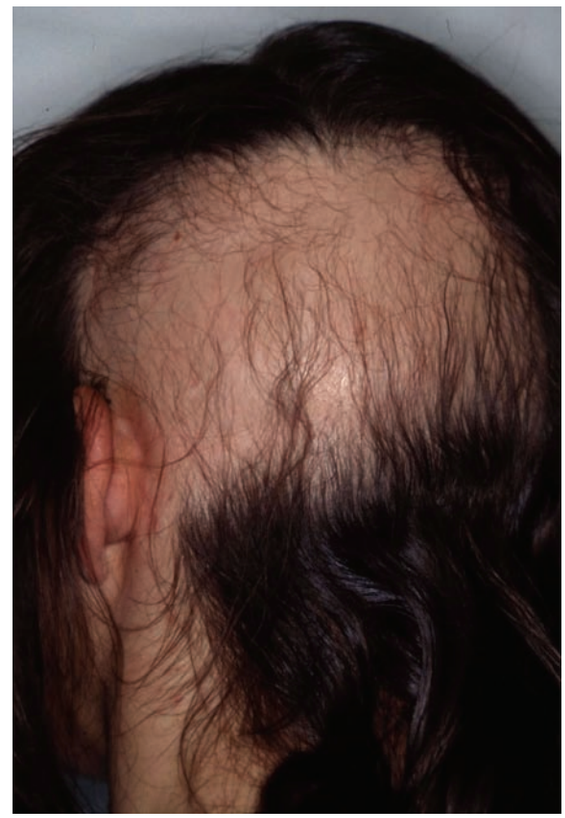


14.38

Figures 14.37, 14.38
Scarring alopecia following scalp irradiation for a brain tumor.

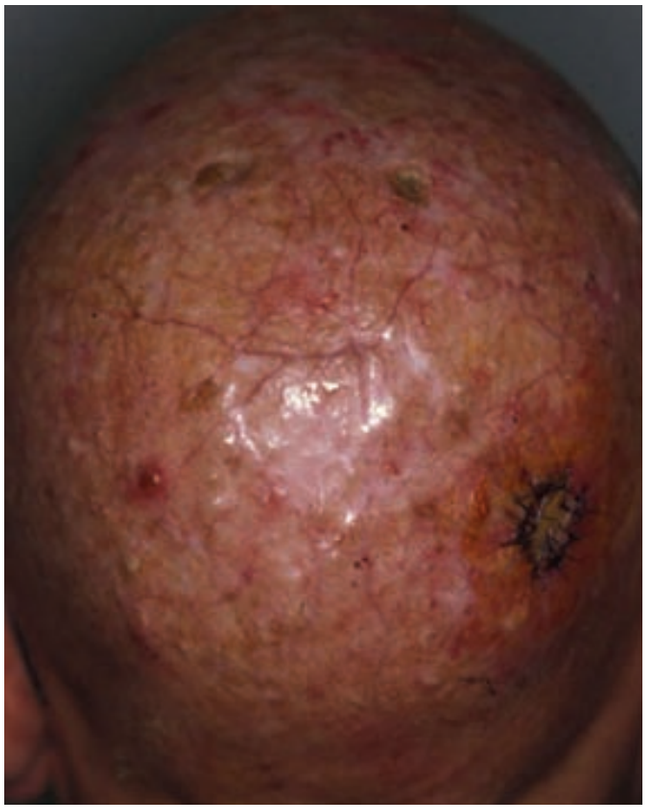


Figure 14.39
Chronic radiodermatitis of the scalp.

14.37, 14.38). X-ray alopecia is most commonly seen in patients undergoing X-ray therapy for brain tumors. Single short-term exposure to 300–600 Gy of irradiation produces temporary epilation (see page 66).

Basal and squamous cell carcinoma are common in chronic radiodermitis (Figure 14.39).

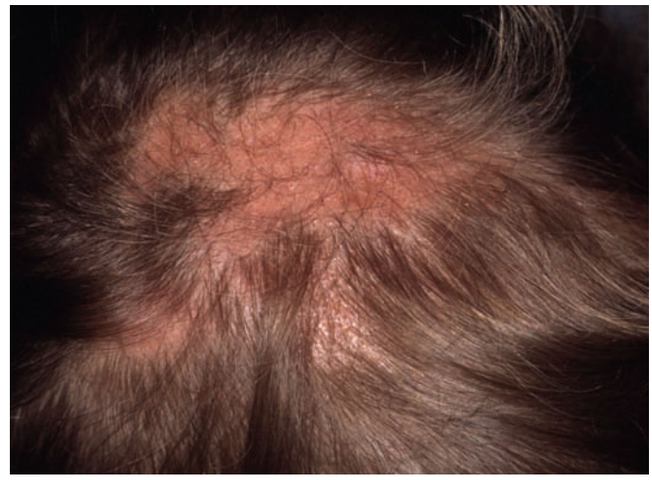
Scleroderma/morfea

Morfea can involve the scalp with one or a few patches of alopecia where the scalp is atrophic and yellow-white in color (Figures 14.40–14.42). Erythema at the periphery of the patch indicates active disease.

Frontoparietal scleroderma 'en coupe de sabre' presents as an enlarging band of linear scleroderma involving the forehead and the frontal scalp (Figures 14.43, 14.44). Bone atrophy and cicatricial alopecia are typical.

Treatment

- Calcitriol 0.50–0.75 μg/day (E).



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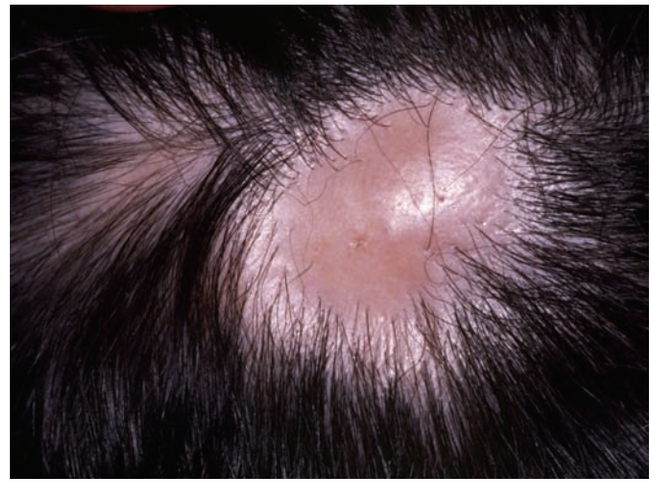

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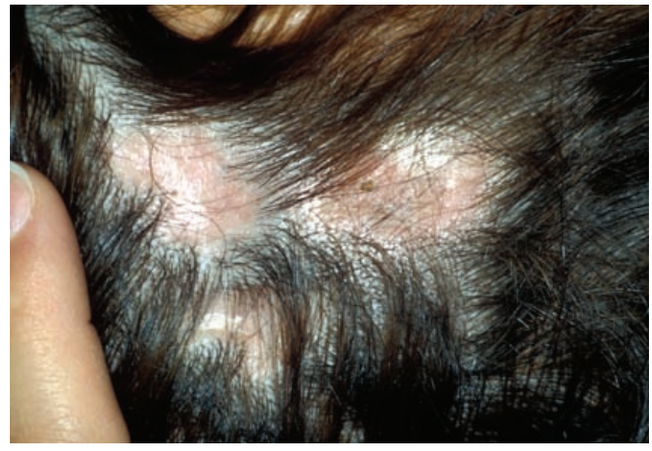

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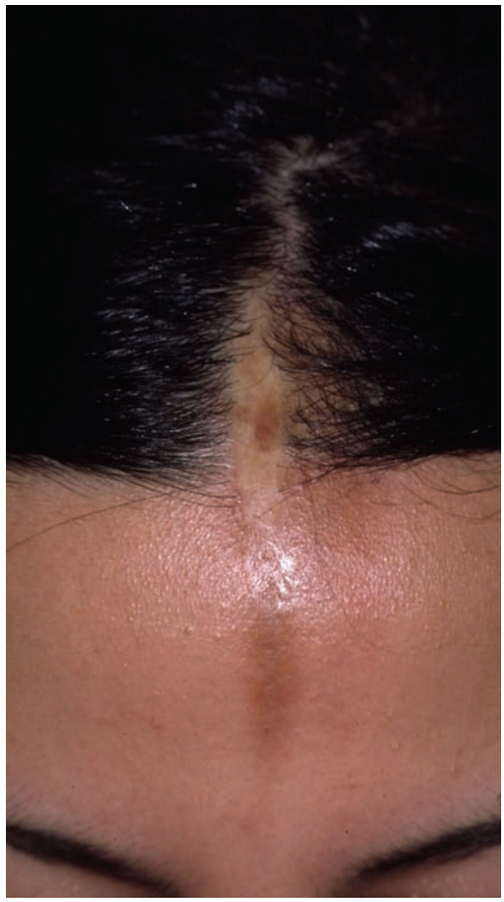
Figures 14.40–14.42
Morfea: cicatricial alopecia with scalp atrophy and yellow color of the scalp.



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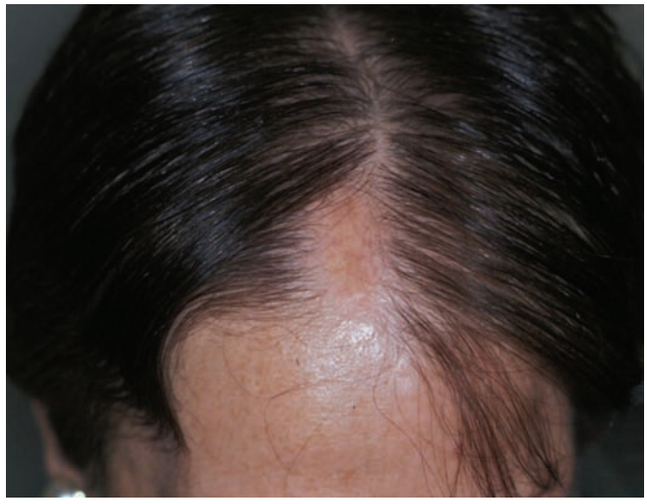

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Figures 14.43, 14.44

Frontoparietal scleroderma 'en coupe de sabre': cicatricial alopecia of the frontal area in a linear pattern.

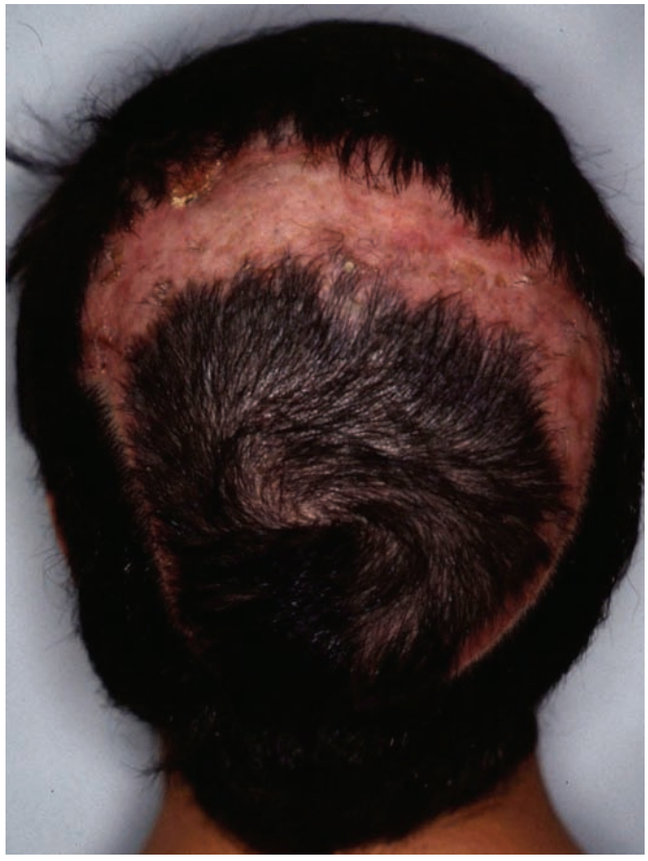

Figure 14.45

Scarring alopecia due to scalp burn.

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15 Infections and infestations

Tinea capitis

Hair invasion by dermatophytes has a different epidemiology depending on the causative agents. *Trichophyton* species are responsible for most infections in North America (*Trichophyton tonsurans*) and in eastern and southern Europe and North Africa (*Trichophyton violaceum*), where they affect both children and adults. Transmission is interhuman and asymptomatic carriers are frequent. In western Europe *Microsporum canis* is the most common cause of the condition and the infection affects almost exclusively children. Transmission occurs through a symptomatic or asymptomatic animal, usually a cat.

Clinical features

- Black dots: tinea capitis due to *Trichophyton* species produces multiple small patches of alopecia due to breakage of the hair shafts at the scalp surface (endothrix invasion) (Figure 15.1). Broken hairs appear as typical black dots in black individuals.
- Scalp scaling: it is a prominent feature in all types of tinea capitis (Figures 15.2–15.5). Scaling is associated with mild or moderate inflammation in *Microsporum canis* infections (Figure 15.6).
- Alopecia: alopecia results from hair shaft breakage and occurs at scalp level in endothrix infections (Figure 15.7) or at 1–3 mm from scalp in ectothrix

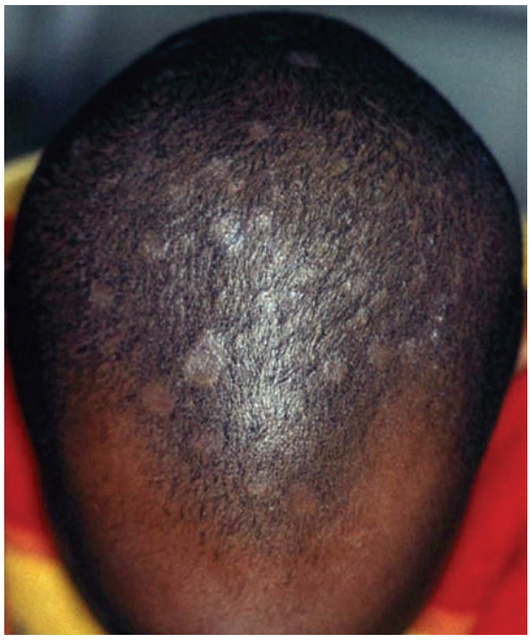


Figure 15.1
Endothrix tinea capitis due to *Trichophyton violaceum*: multiple small patches with hair broken at the scalp emergency (black dots).

Figure 15.2
Patchy scalp scaling and alopecia not associated with inflammation in *Trichophyton violaceum* tinea capitis.

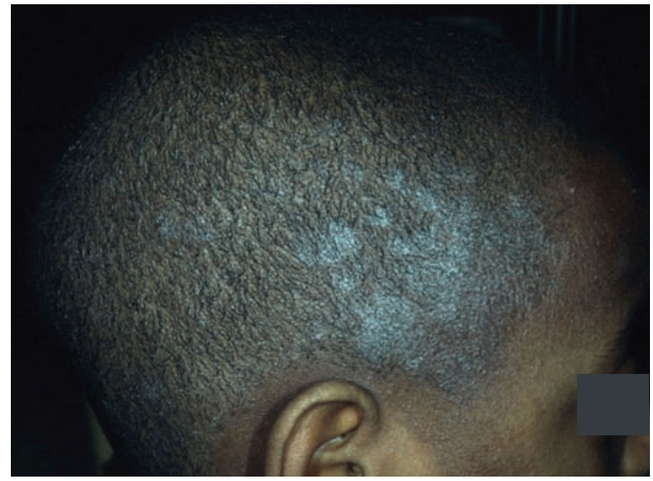
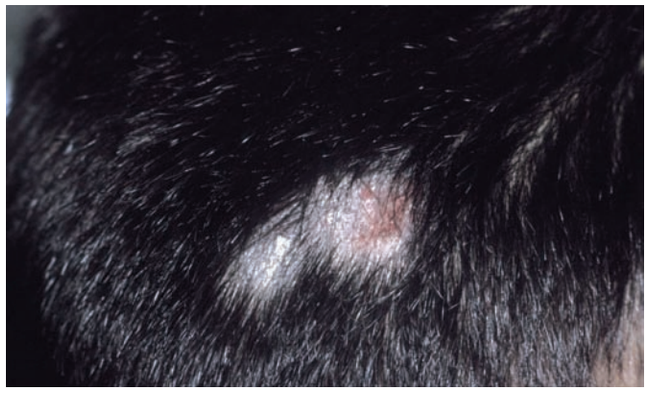


Figure 15.3
Tinea capitis due to *Trichophyton soudanense*.



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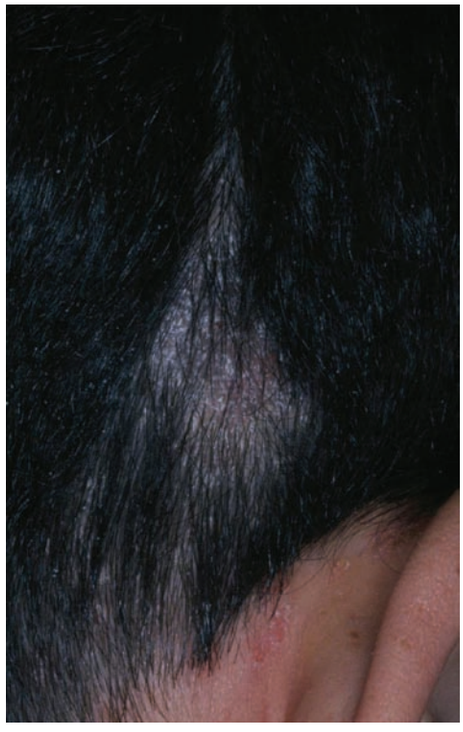


15.5

Figures 15.4, 15.5
Tinea capitis due to *Microsporum canis*.

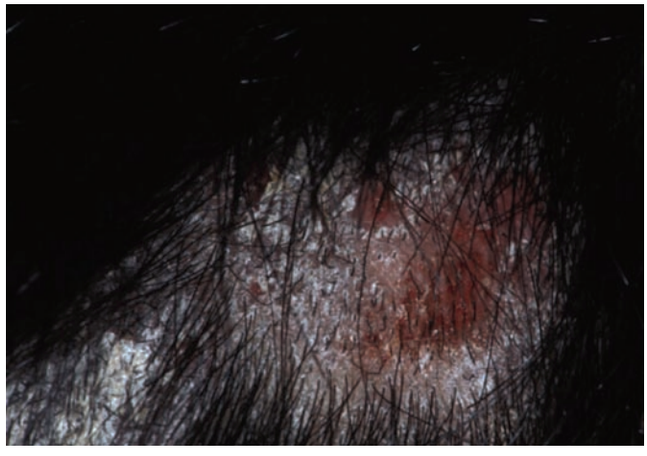


Figure 15.6
Scalp scaling and inflammation in *Microsporum canis* infection.

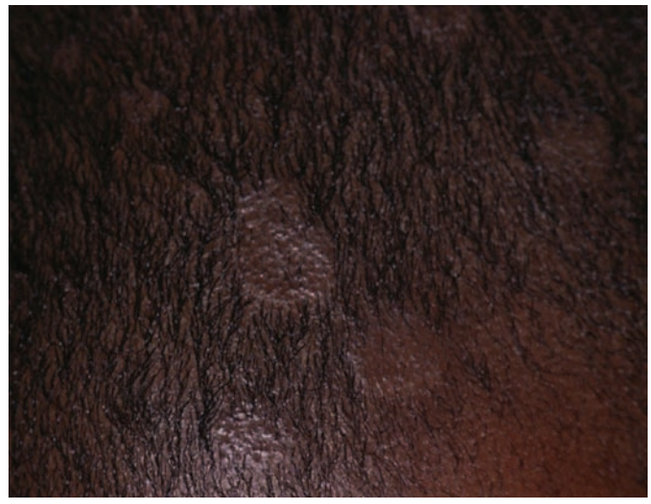


Figure 15.7
Alopecia due to *Trichophyton soudanense*: endothrix infection causes hair breakage at scalp level.

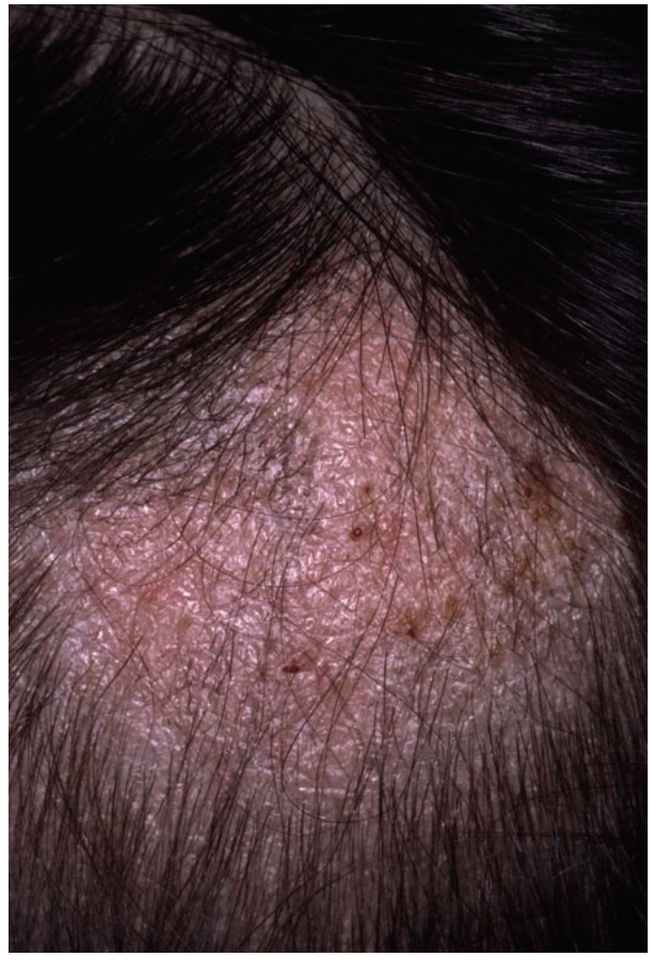


Figure 15.8
Alopecia due to hair breakage a few millimeters after scalp emergency in *Microsporum canis* infection.

infections (Figure 15.8). Pustular lesions may be observed in some cases (Figure 15.9). Cicatricial alopecia occurs in tinea favosa (Figure 15.10) and may complicate kerion (Figure 15.11).

- Lymph node swelling: cervical nodes are often enlarged.

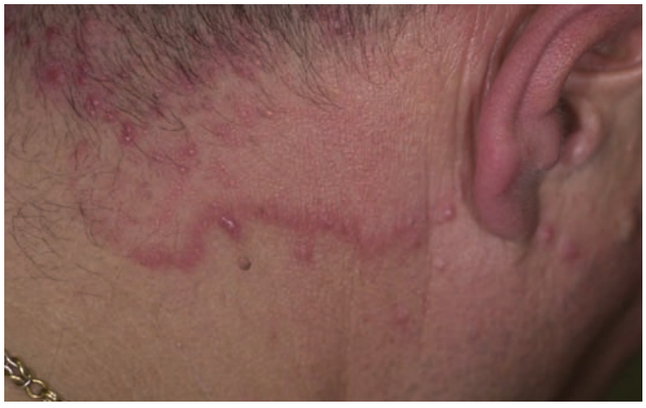


Figure 15.9

Cutaneous pustulae in tinea capitis due to *Trichophyton verrucosus*.

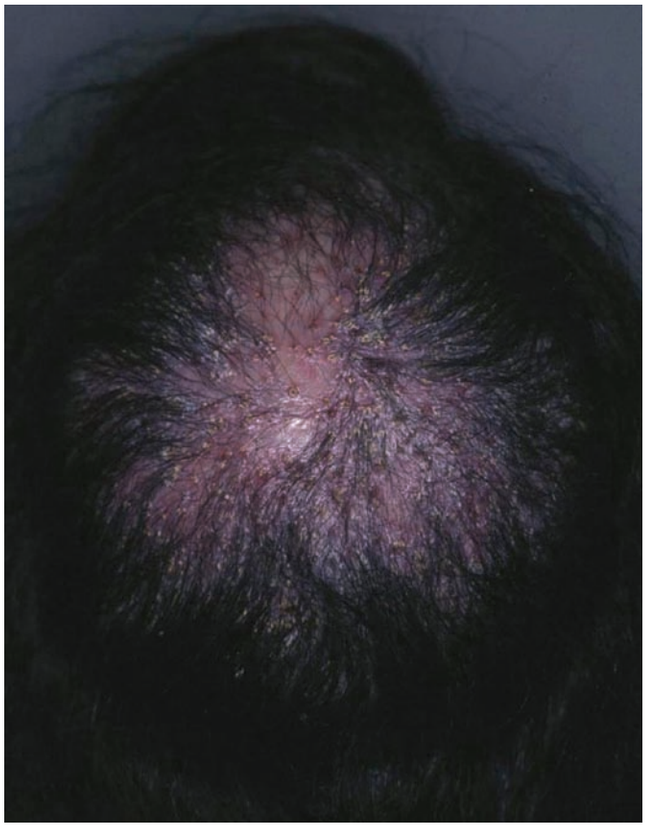


Figure 15.10

Cicatricial alopecia in tinea favosa.

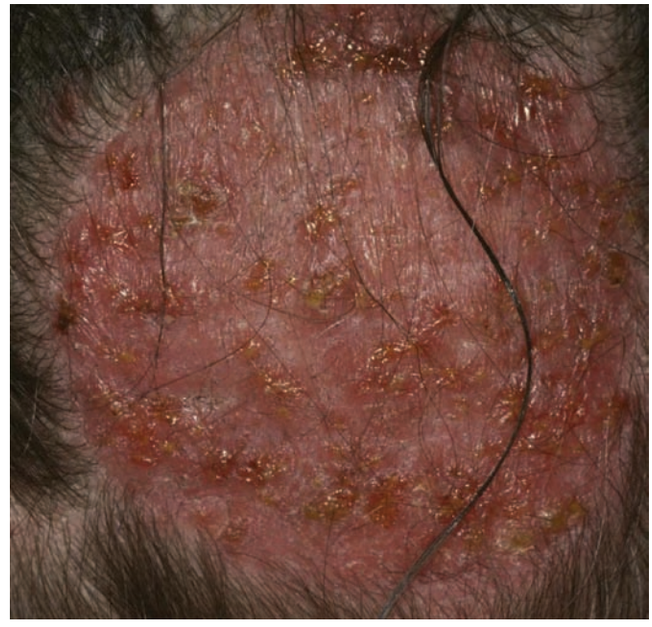


Figure 15.11

Cicatricial alopecia due to kerion in tinea capitis due to *Microsporum canis*.

- Kerion: severe inflammation with abscess formation (Figures 15.12, 15.13) is more frequent in tinea capitis due to zoophylic dermatophytes.
- Scutulum: scutulum is typical of tinea favosa, a rare disease due to *T. shoenleinii* infection. The scalp shows multiple yellow opaque round concave masses that correspond to concretions of fungi and scales (Figures 15.14–15.17). They have a typical urine smell. Erythema and alopecia are prominent features.

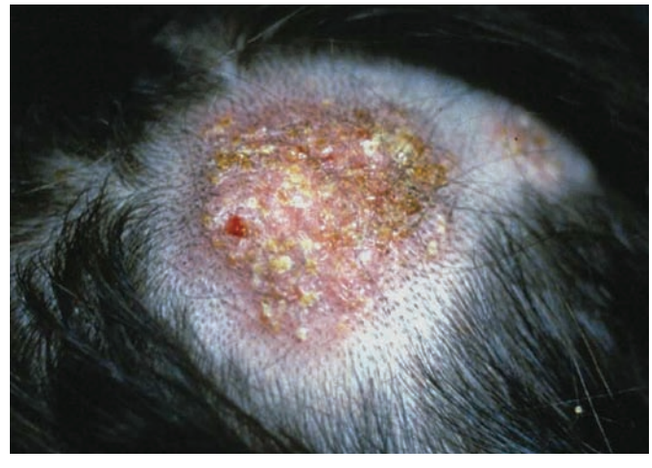


Figure 15.12

Tinea capitis due to zoophilic dermatophytes: kerion.

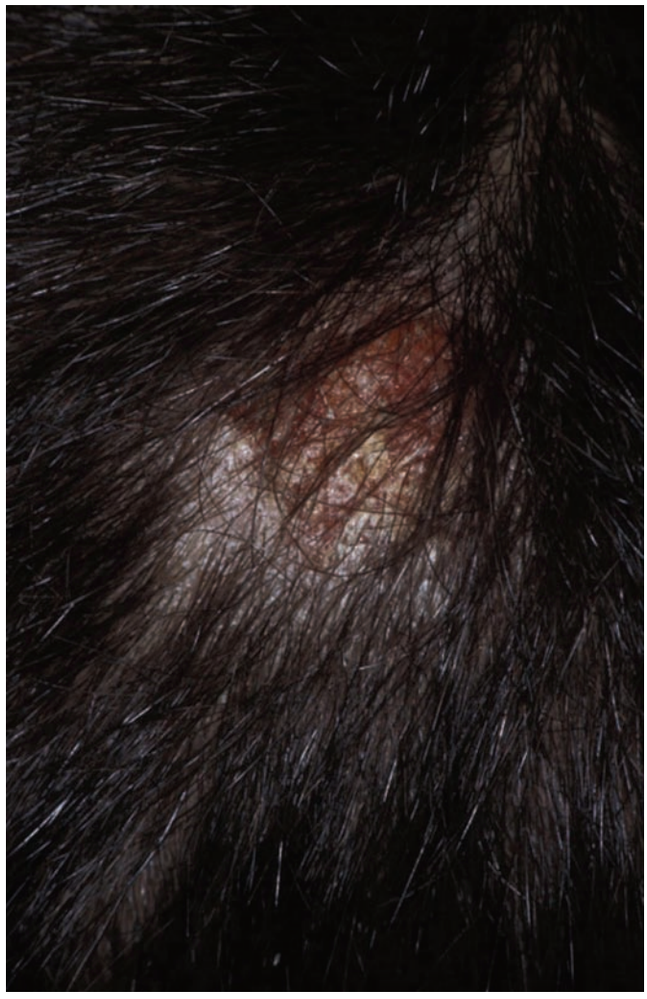


Figure 15.13
Tinea capitis due to zoophilic dermatophytes: kerion.

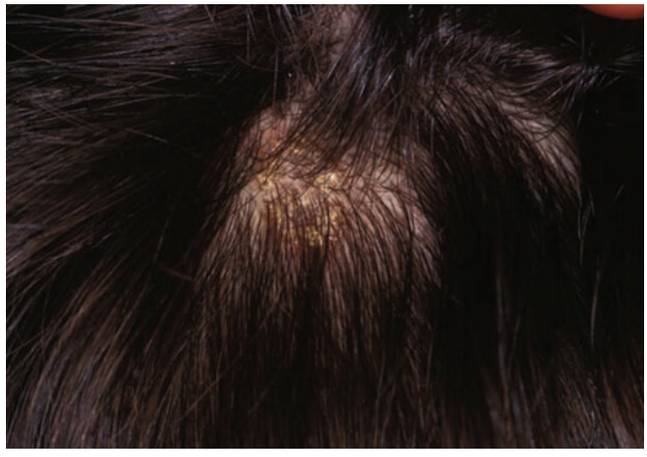
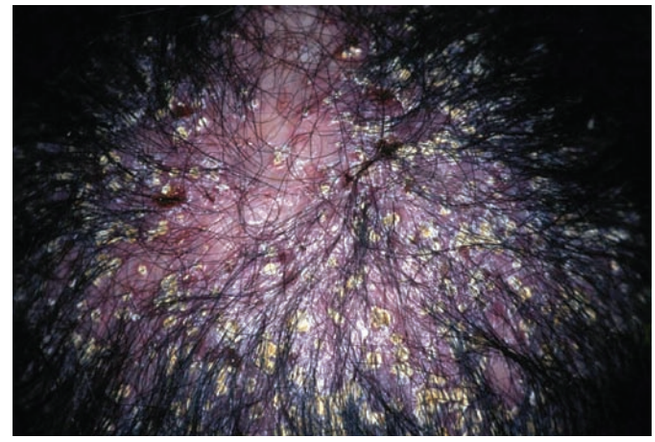


Figure 15.14
Tinea favosa due to *Trichophyton schoenleinii*: scutula.



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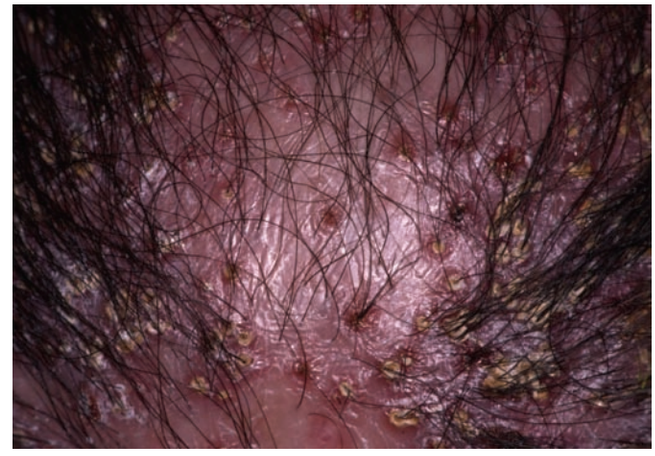


15.16

15.17

Figures 15.15–15.17
Erythema, cicatricial alopecia and scutula in tinea favosa.

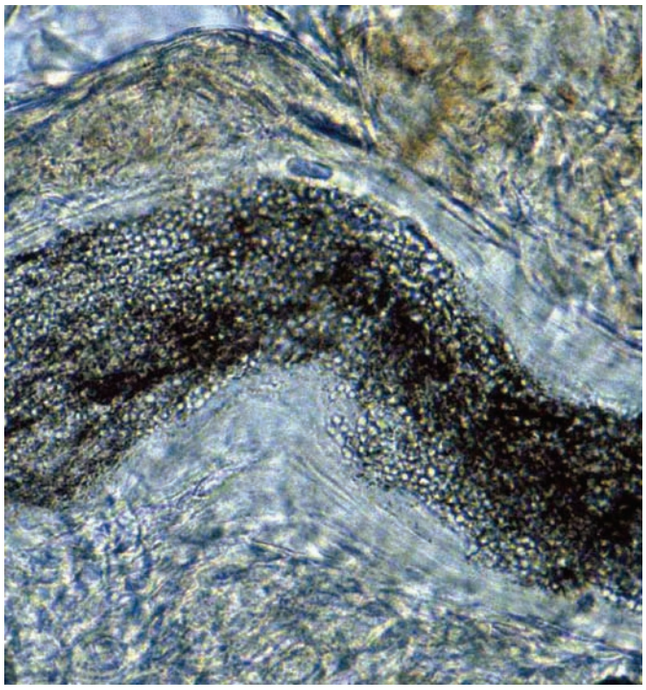


Figure 15.18
Microscopic examination of the broken hair in KOH: endothrix invasion.

Figure 15.19
Schematic drawing of ectothrix hair invasion.

Figure 15.20
Schematic drawing of endothrix hair invasion.

Diagnosis

- Wood's light examination reveals a typical green fluorescence in *M. canis* infections.
- KOH (potassium hydroxide) examination of the scales and broken hairs, which can be obtained by rubbing the scalp with a moistened gauze, confirms the diagnosis (Figures 15.18–15.20).
- Cultures are important to identify the responsible dermatophyte.

Treatment

- Oral antifungals (A) (Table 15.1) should be given until KOH and cultures are negative (usually 6–8 weeks). Griseofulvin is still the gold standard treatment for tinea capitis in children
- Ketoconazole shampoo to decrease spore shedding (B)
- Sterilize clothes and fomites
- Look for asymptomatic human (Figure 15.21) and animal carriers
- Systemic steroids and oral antibiotics improve inflammatory symptoms of kerion (A).

Table 15.1 Treatment of tinea capitis

Microsporum sp.	*Trichophyton sp.*
Griseofulvin 20–25 mg/kg/day	Griseofulvin 20–25 mg/kg/day
Itraconazole 3–5 mg/kg/day	Terbinafine 62.5 mg/day for weight <20 kg 125 mg/day for weight 20–40 kg 250 mg/day for weight >40 kg
Terbinafine 62.5 mg/day for weight <20 kg 125 mg/day for weight 20–40 kg 250 mg/day for weight >40 kg	Itraconazole 3–5 mg/kg/day
Fluconazole 5–7.5 mg/kg/day	Fluconazole 5–7.5 mg/kg/day

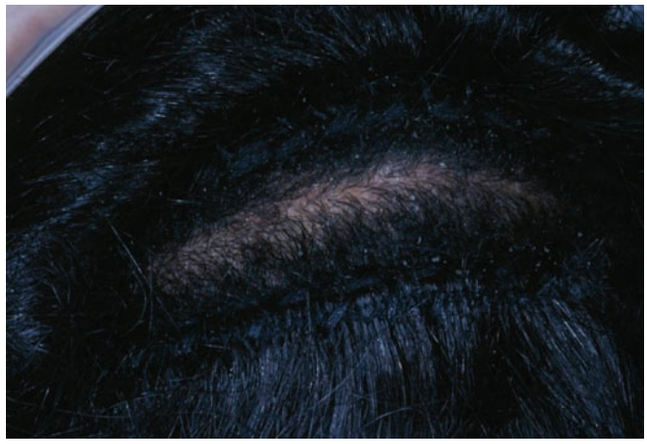


Figure 15.21
Scalp scaling in the mother of a child with *Trichophyton violaceum* tinea capitis: cultures were positive for the same dermatophyte.

Figure 15.22
Pediculus capitis on the hair shaft.

Pediculosis capitis (head lice)

It is a very common condition in school-aged children affecting up to 30% of children between the ages of 3–9 years. Incidence of pediculosis is much lower in blacks, probably because of the daily application of styling greasy products that prevent infestation. Spreading of the infestation is mostly through head to head contact, but transmission through fomite can also occur.

- Pruritus is the most common symptom, scalp erosions due to scratching and lymphoadenopathy may also be present.
- Clinical examination only rarely permits to visualize the lice (Figure 15.22), but it shows the presence of multiple nits, which are strongly cemented to the most proximal portion of the hair shaft (Figures 15.23, 15.24). Nits, which are especially evident in the retro-auricolar area and on the nape, appear as tear shaped gray to brownish 2.3 mm long formations that do not move along the hair shaft with traction (Figure 15.25). Nits, which correspond to lice eggs, may be viable (brownish) (Figures 15.26–15.30) or dead (white) (Figures 15.31, 15.32).

15.23

15.24

Figures 15.23, 15.24
Scalp pediculosis: nits.

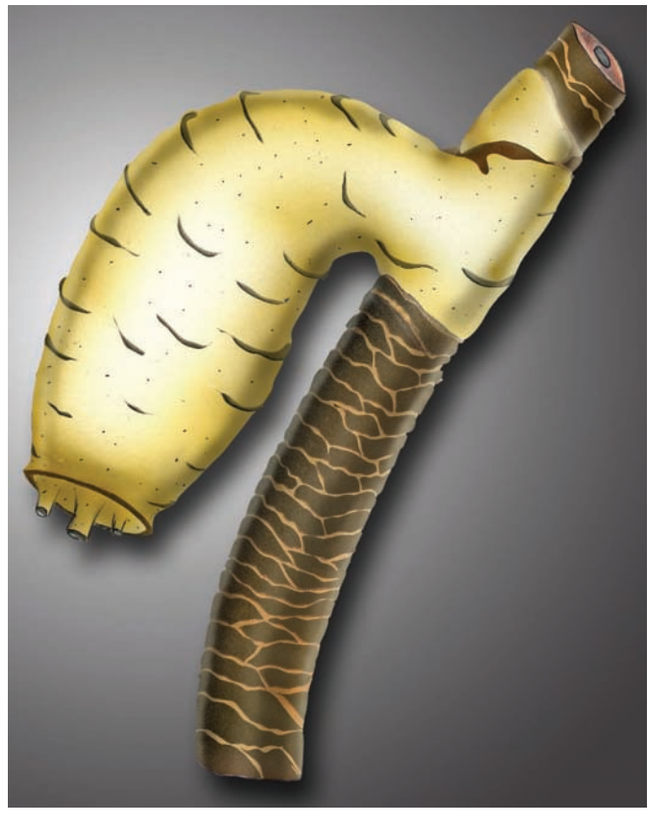


Figure 15.25
Nit on the hair shaft.

15.26

15.27

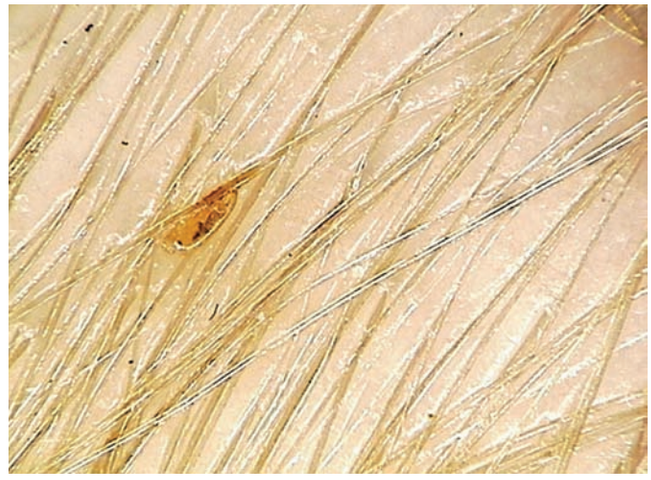
Figures 15.26, 15.27
Viable nits.

Figure 15.28
Viable nits.

Figure 15.29
Viable nits at the dermatoscope.

Figure 15.30
Viable nits at the dermatoscope.

15.31



15.32

Figures 15.31, 15.32
Dead nits.

Treatment (Table 15.2)

- Prevent community spreading by examination of all strict contacts
- Kill lices by insecticides or viscous petrolatum products (alternative)
- Kill and remove nits by manual removal and regular combing
- Sterilize clothes and fomites.

Table 15.2 Treatment of head lices

Malathion 0.5% lotion/gel (A)
Permethrin 1% (A)
Synthetic pyretroids (phenothrin) (B)
Oral invermectin 200–250 µg/kg (D)
Natural pyrethrins (E)
Oral clotrimoxazole (E)
Topical petrolatum (E)

Insecticides utilized for the treatment of pediculosis capitis act though a neurotoxic mechanism and do not kill eggs, which do not possess a nervous system. Insecticide treatment should therefore be repeated after 7 days to kill the newborn lices. Insecticide creams, lotions or foams are applied on the scalp and hair for several hours and then rinsed. Use of insecticide-containing shampoos as a preventive measure is to be avoided as it increases diffusion of insecticide-resistant lice.

Tick infestation

Temporary alopecia is a common sequela of tick bites (Figure 15.33) due to the effects of anticoagulants injected by the tick to suck blood.

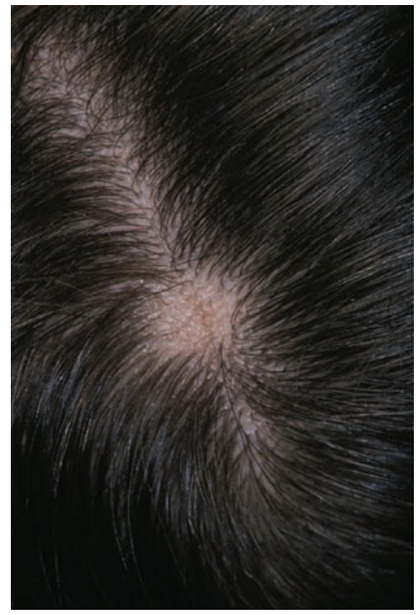


Figure 15.33
Temporary alopecia following tick bite.

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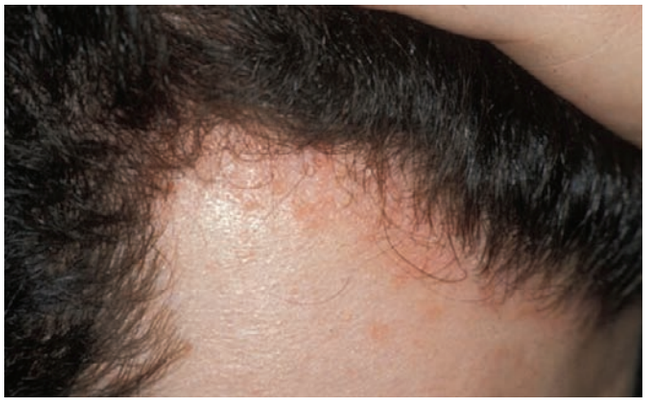
16 Seborrheic dermatitis

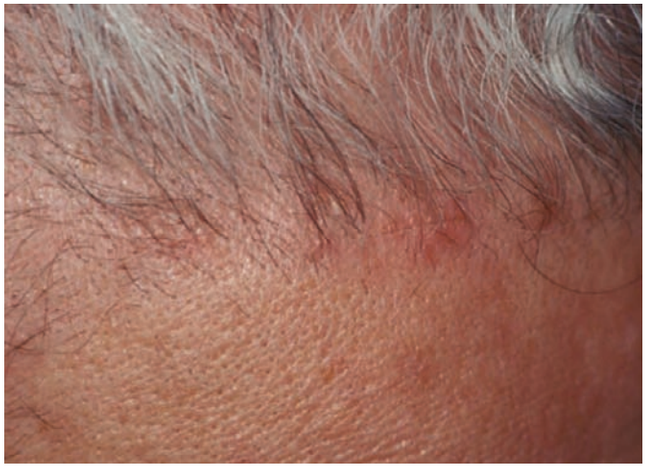
Seborrheic dermatitis affects 1–3% of the general population with up to 95% of patients having scalp involvement.

Clinical features

- The skin along the frontal hairline is most affected with mild erythema (Figures 16.1, 16.2) and scaling and the skin lesions often follow hairline recession in patients with pattern alopecia (Figure 16.3). Itching is common and scalp excoriations can be observed (Figure 16.4). Small follicular pustules may



16.1



16.2

Figures 16.1, 16.2
Seborrheic dermatitis of the hairline.

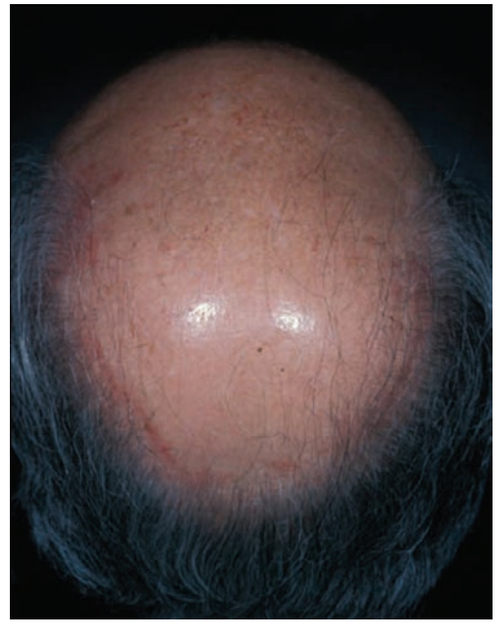


Figure 16.3
Seborrheic dermatitis of the hairline.

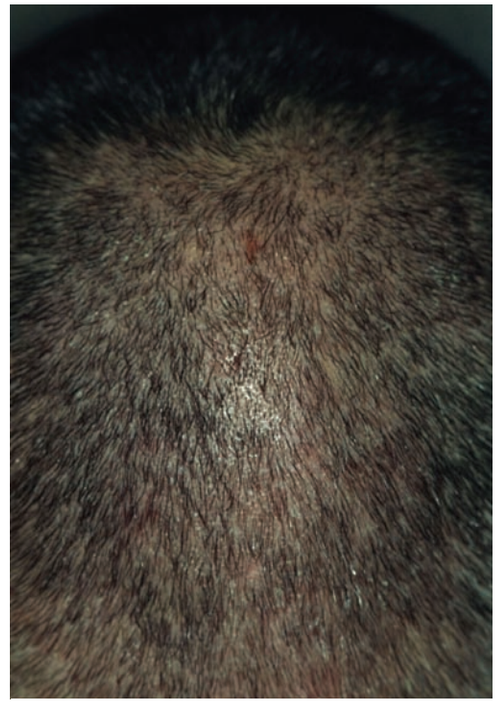


Figure 16.4
Scalp erythema and greasy scales in seborrheic dermatitis.

occur. Other hairy regions, such as the beard, the moustache, eyelashes (Figure 16.5) and eyebrows are also commonly affected. In newborns, seborrheic dermatitis typically produces cradle cap (Figure 16.6).

- Perspiration plus sebum contribute to give the hair a greasy, 'dirty' appearance and an unpleasant smell.
- Dandruff is considered a mild variant of seborrheic dermatitis characterized by non-erythematous subclinical inflammation with moderate to severe flakes (Figure 16.7, Table 16.1).

Figure 16.5
Seborrheic blepharitis.

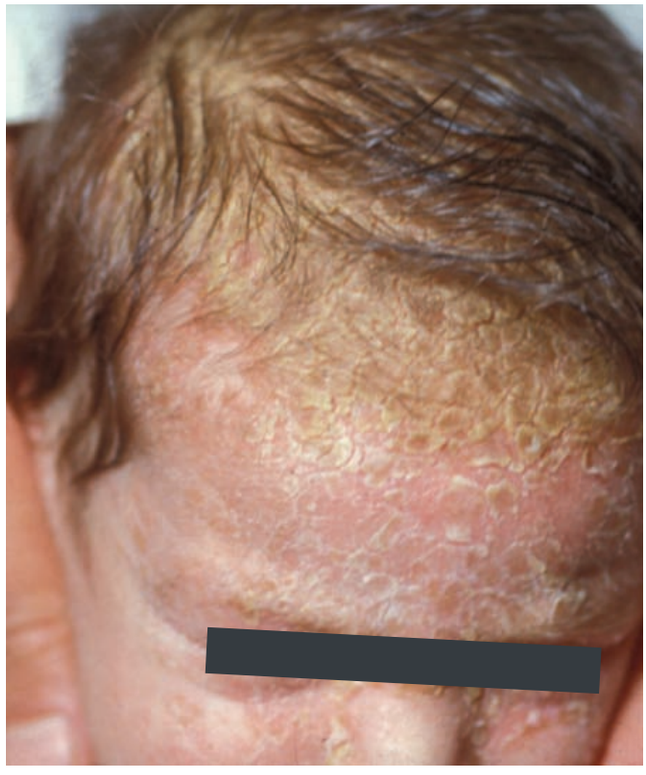


Figure 16.6
Cradle cap.

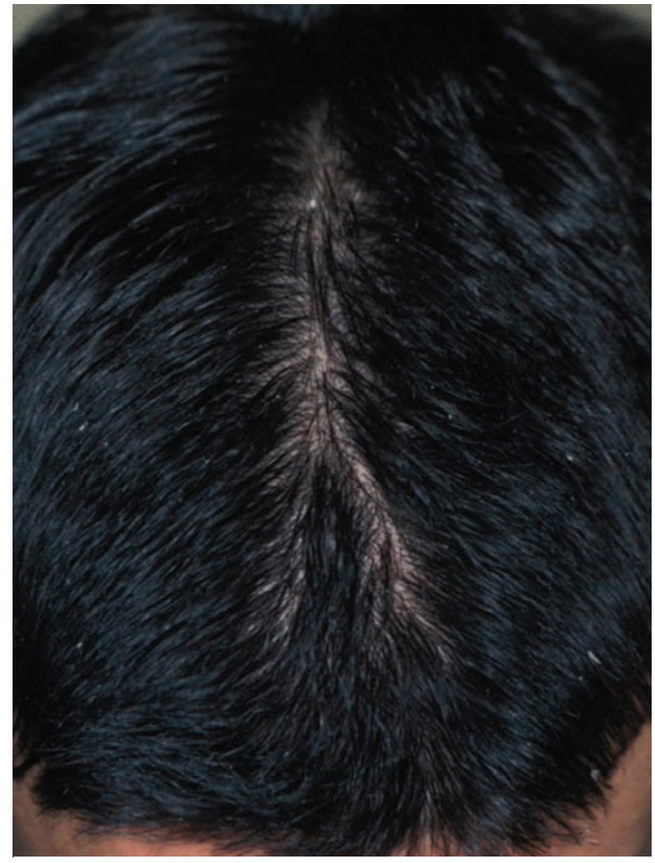


Figure 16.7
Dandruff.

Table 16.1 Dandruff

Affects mostly young males (up to 50% of 20-year-old males)
Exacerbates in winter
Is frequently accompanied by pruritus
May or not be associated with seborrhea
May be exacerbated by dietetic factors and psychological stress

Two varieties of dandruff are distinguished according to the presence or absence of seborrhea:

- Pityriasis sicca, characterized by small, dry and farinaceous scales (Figure 16.8)
- Pityriasis steatoid, characterized by large, thick and oily scales (Figures 16.9, 16.10).

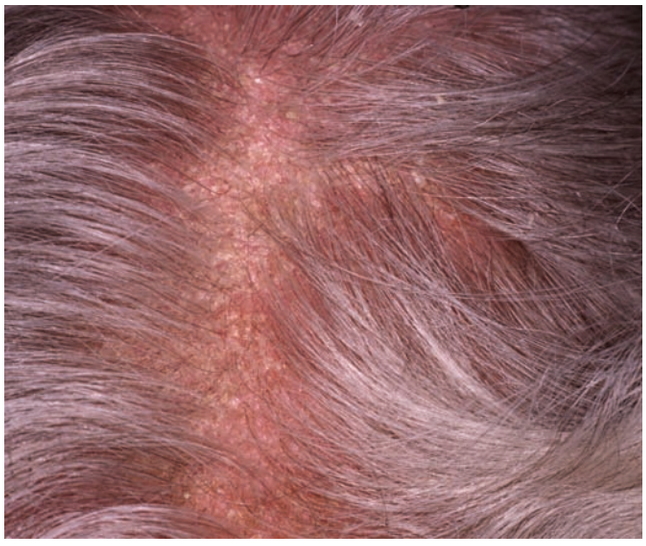
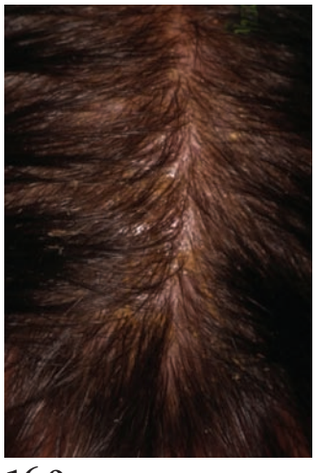
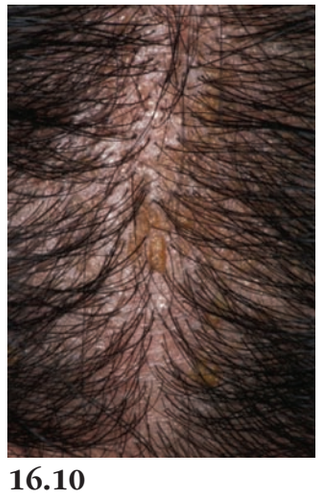


Figure 16.8
Pityriasis sicca.



16.9



16.10

Figures 16.9, 16.10
Pityriasis steatoid.

Pathogenesis

The pathogenesis of seborrheic dermatitis is summarized in Table 16.2.

Table 16.2 Pathogenesis of seborrheic dermatitis

Abnormal epidermal proliferation
Malassezia sp. colonization
Variation in the scalp content of lipids

Diagnosis

Differential diagnosis with psoriasis can be difficult since face and scalp psoriasis can cause greasy scales in seborrheic patients (sebopsoriasis). In Darier's disease scalp involvement may produce erythema, papules and greasy scales (Figure 16.11).

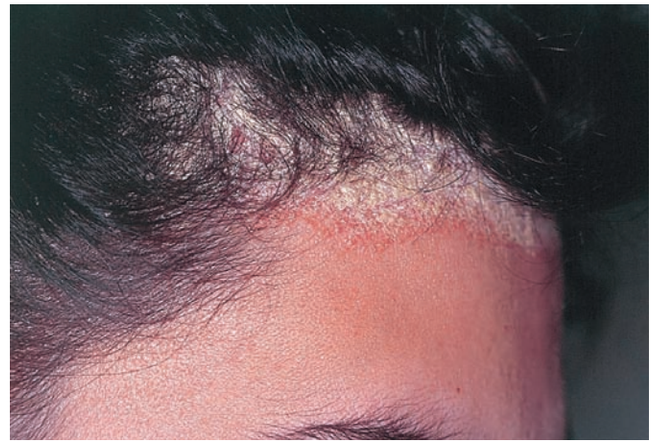


Figure 16.11
Erythematous papules and greasy scales of the scalp in Darier's disease.

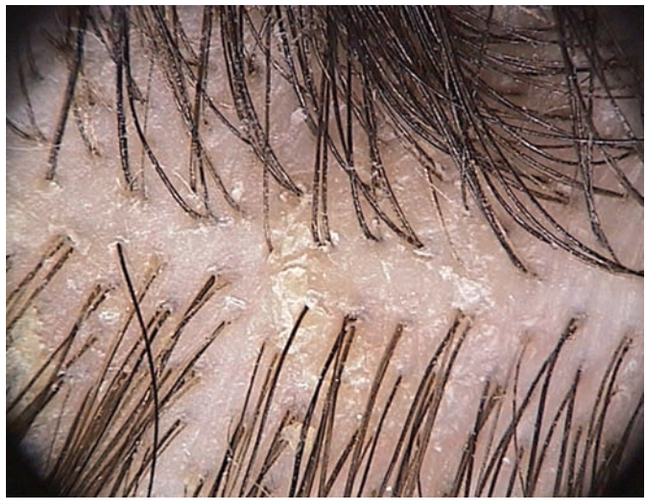
Scalp dermoscopy is useful (see Table 16.3, Figures 16.12–16.15).

Table 16.3 Scalp dermoscopy

Psoriasis	*Seborrheic dermatitis*
Scales	Scales
Hair casts	Hair casts
Dilated capillary loops	

Treatment

- Dandruff is a chronic recurrent condition that requires continuous treatment (Table 16.4).
- Frequent shampooing is itself a good treatment: it is important to instruct the patient to have a shampoo at least every other day because dandruff scales reform 4–7 days after the last shampoo, parallel to *Malassezia* colonization of the scalp.



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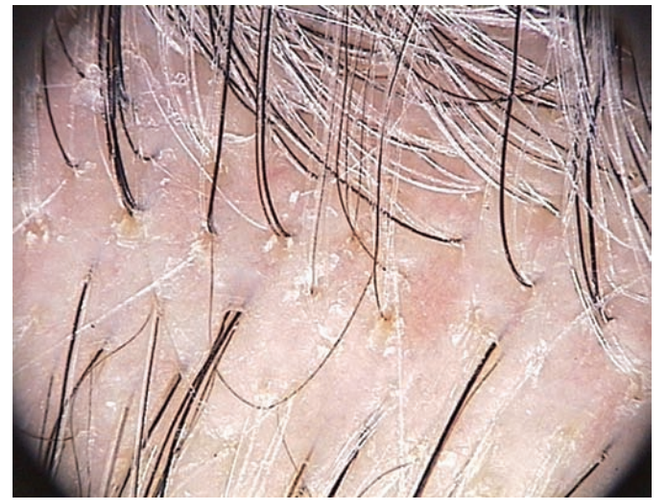

16.14

16.13

16.15

Figures 16.12–16.15
Scalp dermoscopy of seborrheic dermatitis.

Table 16.4 Factors influencing seborrheic dermatitis

Beneficial	*Exacerbating*
Mild UV exposure Frequent shampooing	Strong UV exposure Alcohol Diet Tobacco smoking Psychological distress

- Treatment objectives include:

decrease sebum → production	oral contraceptives in women (A) isotretinoin 0.3–0.5 mg/kg/day in both sexes (B)
reduce *Malassezia* → colonization	antifungal shampoos (A) 5% ketoconazole shampoo 0.75% pyroctone olamine shampoo 1–2.5% selenium sulfide shampoo 1% zinc pyrithione shampoo

Shampoos should be left on the scalp for 3–5 minutes before rinsing to allow the antimicrobial effect.

Oral antifungals can be → an option in severe cases	itraconazole 200 mg/day for 7 days (B) terbinafine 250 mg/day for 4 weeks (A)
reduce inflammation →	anti-inflammatories and corticosteroids (B)

Topical corticosteroids in foams and lotions are useful when itching and inflammation are prominent symptoms.

scale removal → keratolytics (E)

Keratolytics however deplete barrier lipids and contribute to impair the scalp skin barrier.

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17 Scalp psoriasis

Although the scalp is a typical localization of psoriasis (up to 50% of patients), hair loss is not common. Combing may contribute to the frequency of scalp psoriasis through Koebner phenomenon.

Clinical features

The scalp may be the only localization of the disease. The severity of scalp psoriasis is extremely variable, ranging from small scattered patches to diffuse scaling of the scalp.

- Mild forms are often misdiagnosed as dandruff (Figures 17.1–17.4). Itching is not uncommon.

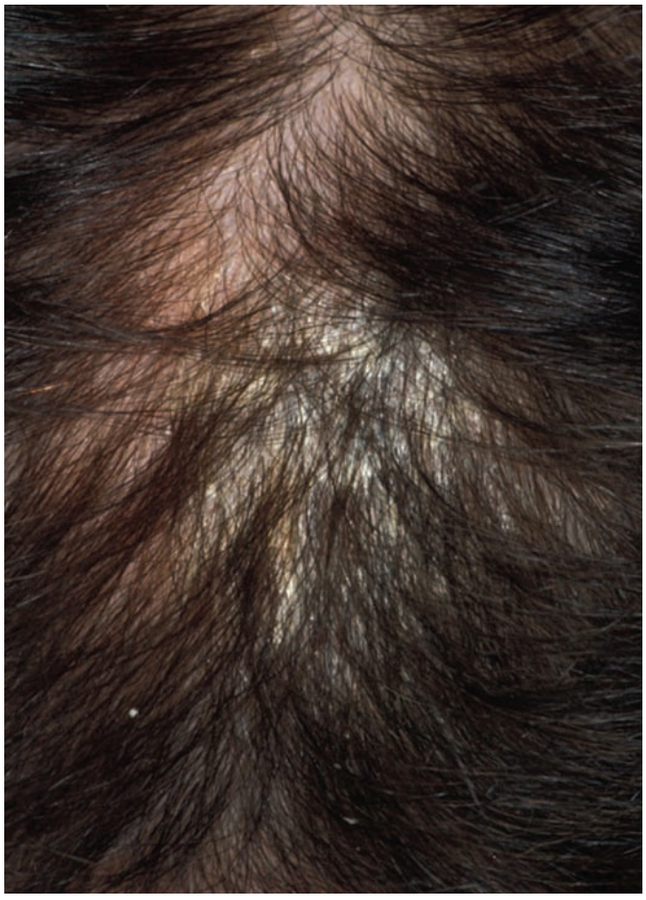
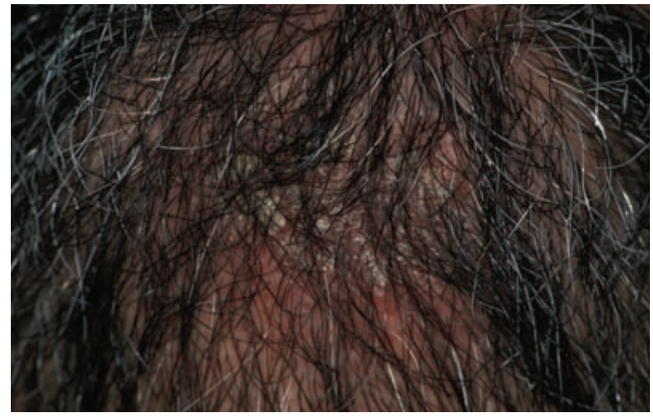


Figure 17.1
Mild scalp psoriasis.

17.2



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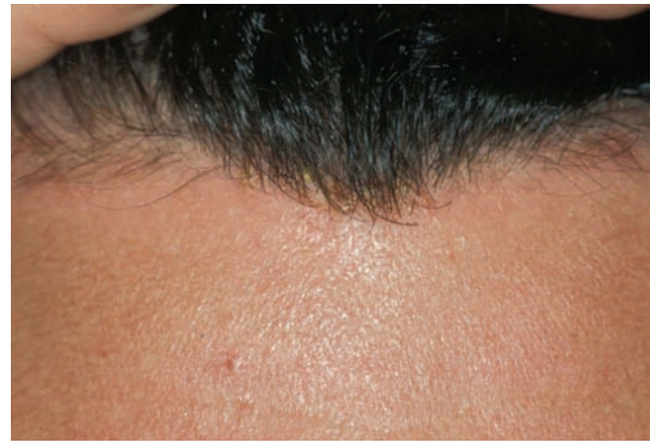

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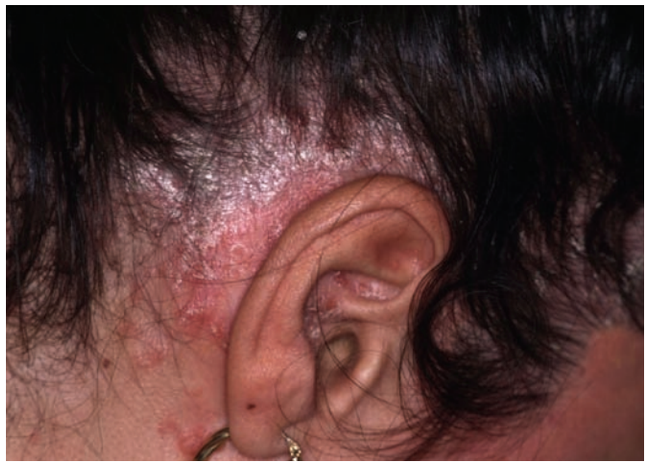
Figures 17.2–17.4
Mild scalp psoriasis resembling seborrheic dermatitis.



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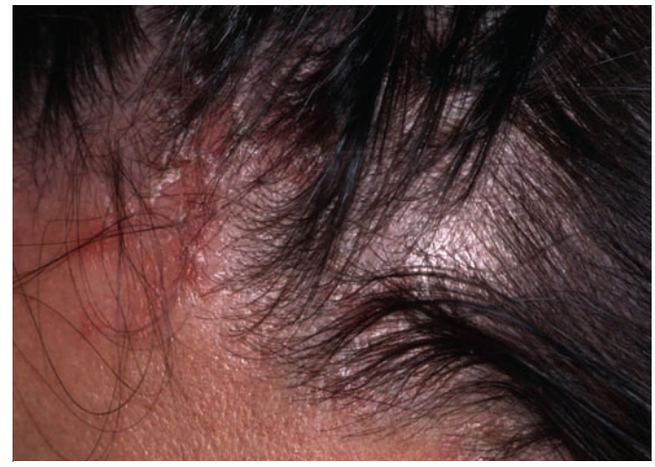

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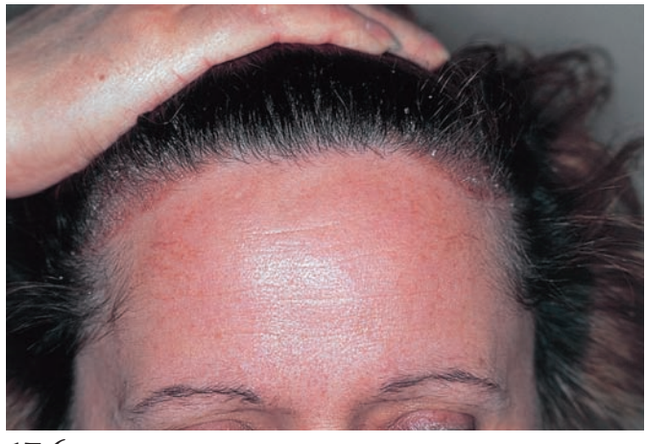

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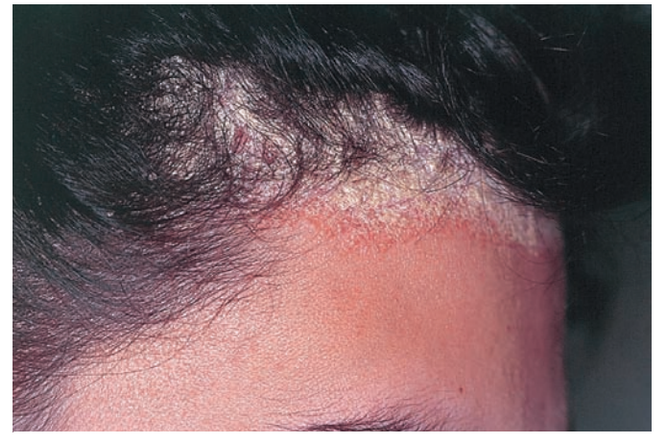

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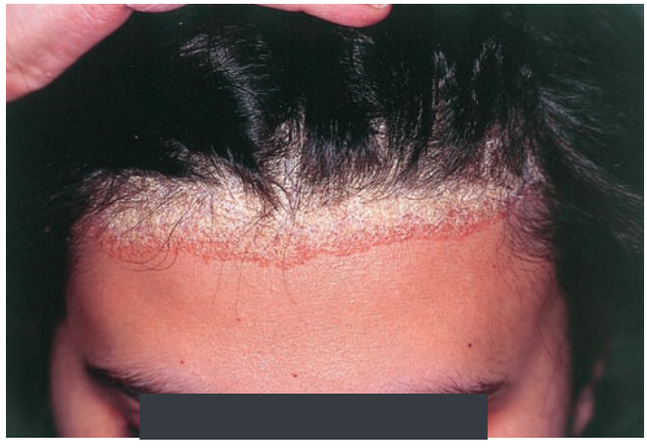


17.7

Figures 17.5–17.9
Scalp psoriasis: hairline involvement.

- The hair line may be affected by typical erythematous-squamous psoriatic lesions (Figures 17.5–17.9).
- In patients with oily skin differential diagnosis with seborrheic dermatitis may be difficult due to the greasy appearance of the scales (sebopsoriasis) (Figure 17.10).
- A diffuse silvery-white cap of scales may cover the whole scalp (Figures 17.11–17.14).
- Scalp psoriasis may cause hair loss which may be moderate or massive and often occurs in tufts. It may be caused by cytokine release due to the inflammatory infiltrate or can be a consequence of delayed

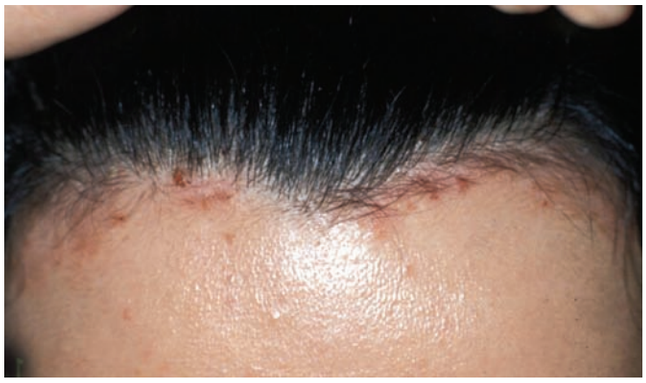
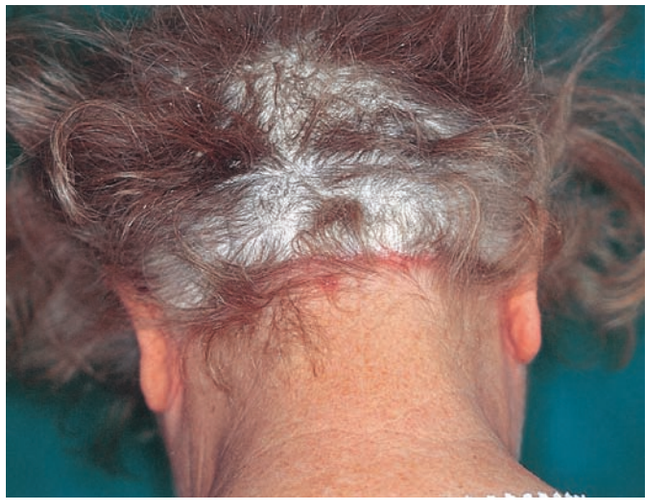


Figure 17.10
Sebopsoriasis.



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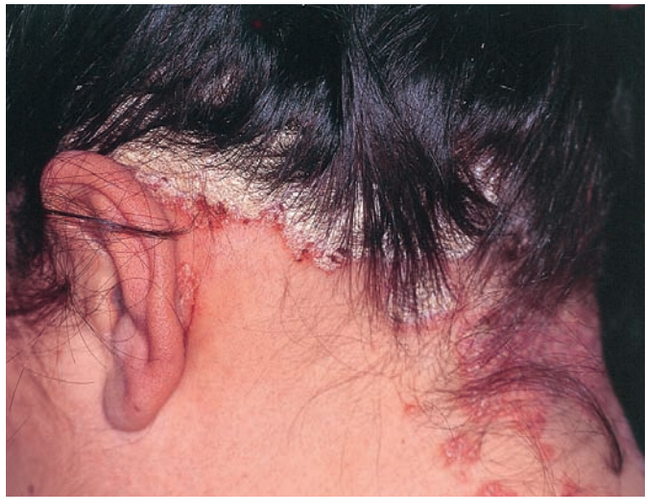

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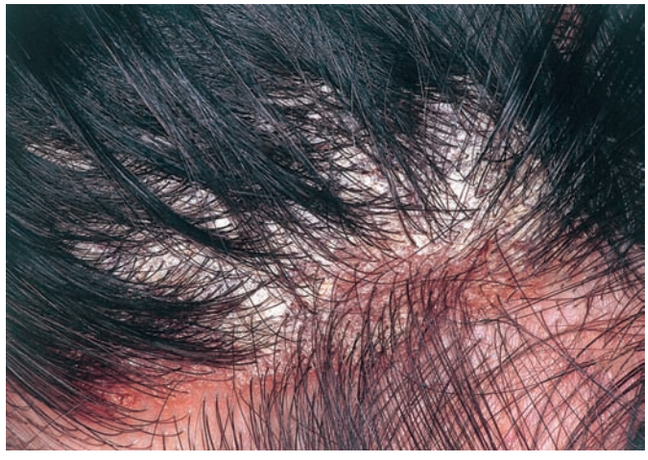

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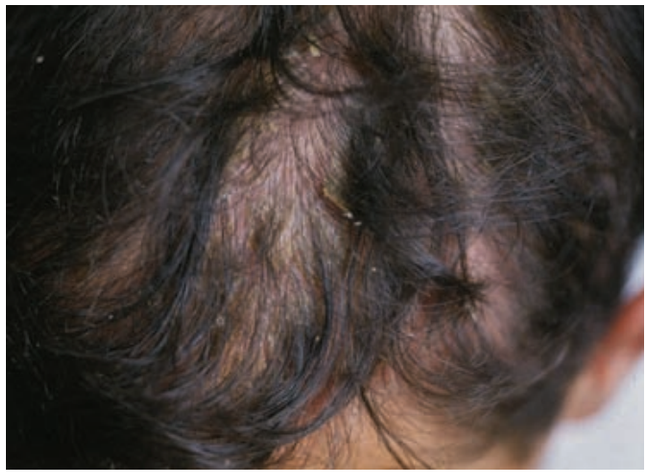
Figures 17.11–17.14
Psoriatic cap.



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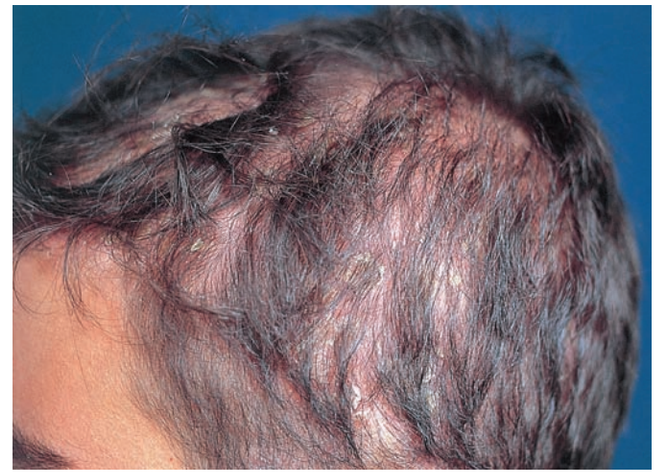

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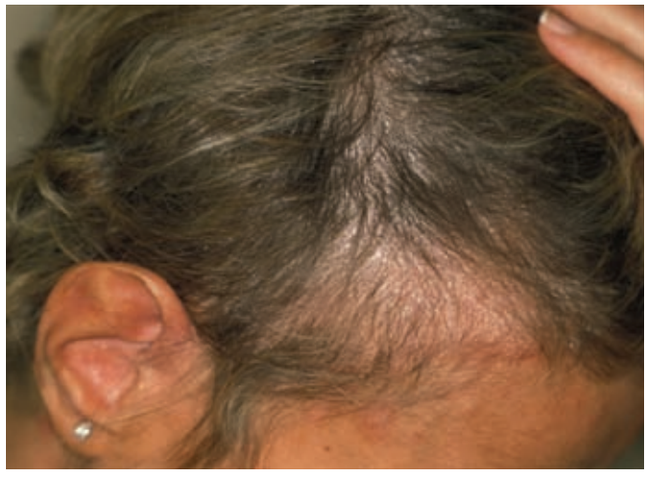

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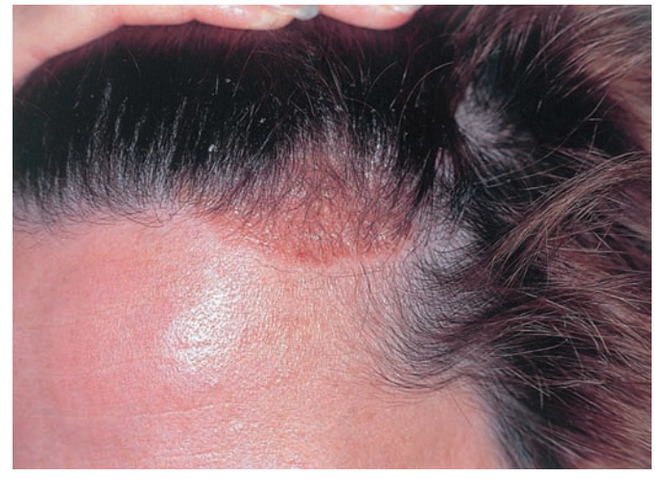


17.18

Figures 17.15–17.18
Mild alopecia in scalp psoriasis.

Figure 17.19
Nail psoriasis: irregular pitting.

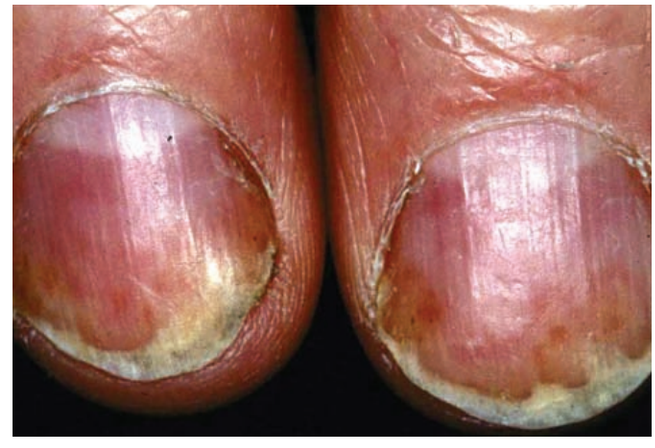
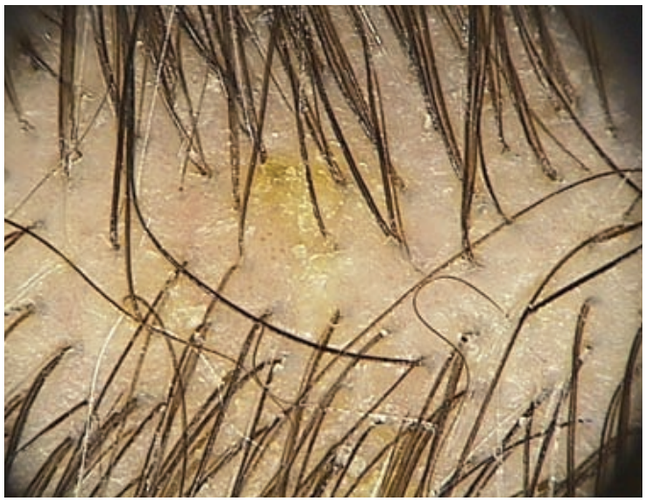


Figure 17.20
Nail psoriasis: salmon patches of the nail bed.



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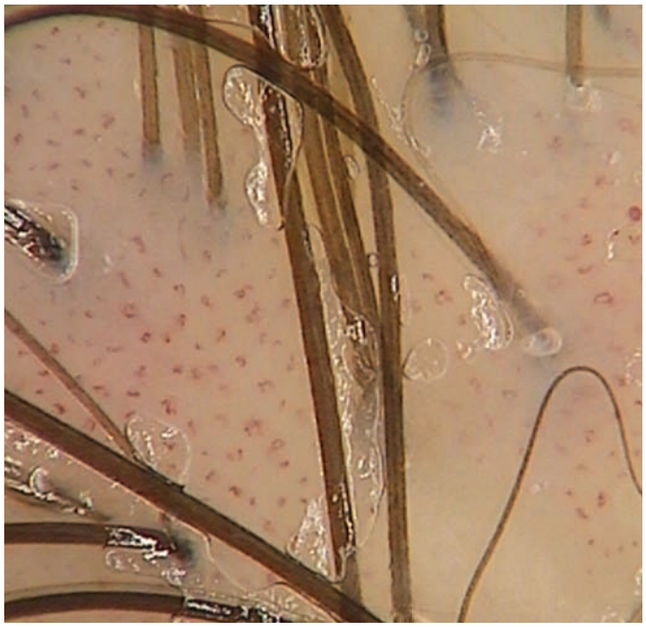

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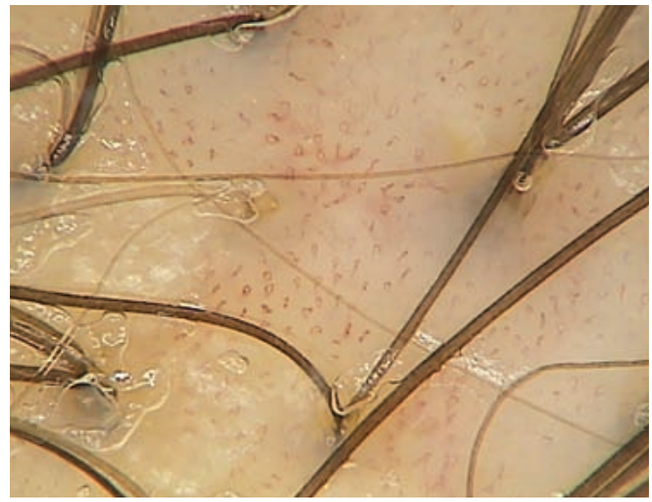

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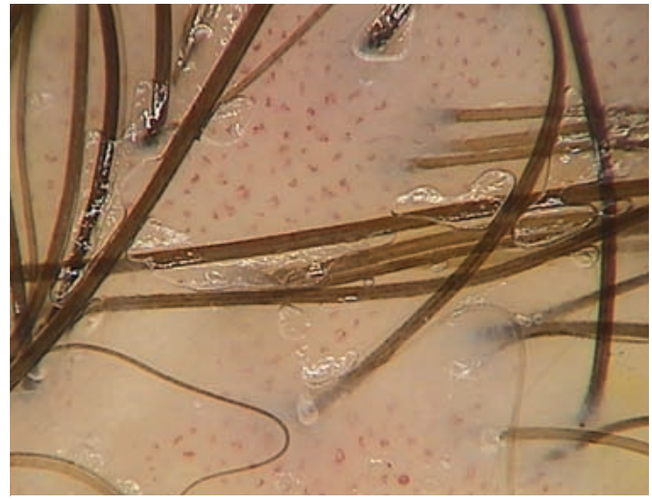

17.25

Figures 17.21–17.25
Yellow scales and coiled capillary loops.

teloptosis. Alopecia may be evident (Figures 17.15–17.18). Scarring may be a long-term sequela.

Diagnosis

- Look for typical nail lesions (Figures 17.19, 17.20).
- Dermoscopy (Figures 17.21–17.25): the presence of uniformly distributed tightly coiled capillary loops is typical.

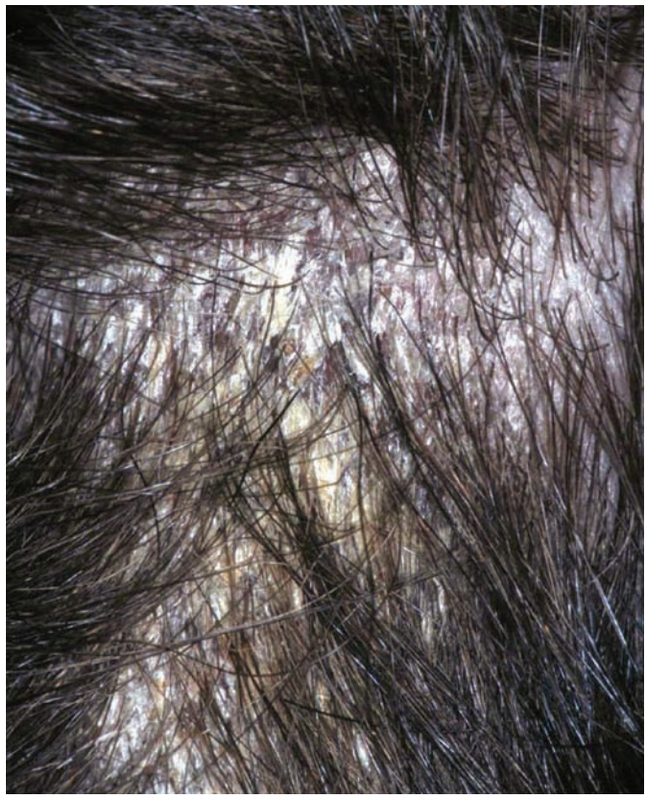

Figure 17.26
Pseudotinea amiantacea.

Treatment

- Calcipotriol/calcipotriene solution (A)
- Topical steroids (A): betamethasone valerate foam, momethasone/betamethasone dipropionate lotion, clobetasone propionate 0.05% solution/foam
- UVB comb (C)
- Tar shampoos (D)
- Keratolytics (salicylic acid 5–10% in ointment) (E).

Pseudo tinea amiantacea

This is probably a reaction pattern of the scalp to various inflammatory scalp conditions, including psoriasis, seborrheic dermatitis, eczema.

It is more common in children, in young adults and in females. The scalp presents one or a few patches of thick, silvery adherent scales that engulf and bind down tufts of hair (Figure 17.26).

Pseudotinea amiantacea may be associated with hair loss which is usually temporary. Scarring alopecia due to pseudotinea amiantacea has been reported.

Diagnosis

Mycological examination to exclude tinea capitis.

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18 Scalp contact dermatitis

Allergic contact dermatitis of the scalp is not common (only 4% of cases of head eczema involve the scalp) and is most frequently caused by hair dyes or topical drugs. Shampoos are an uncommon cause of scalp contact dermatitis due to dilution and short contact time with the skin.

Clinical features

- Most patients complain of itching. Clinical examination reveals mild scaling and excoriations (Figure 18.1).

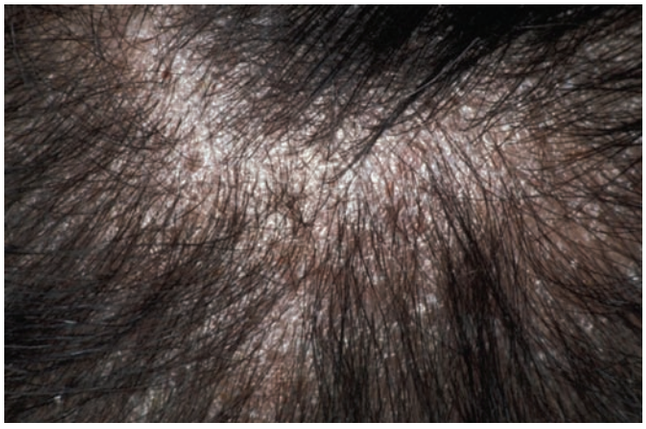


Figure 18.1
Contact dermatitis: mild scalp scaling.

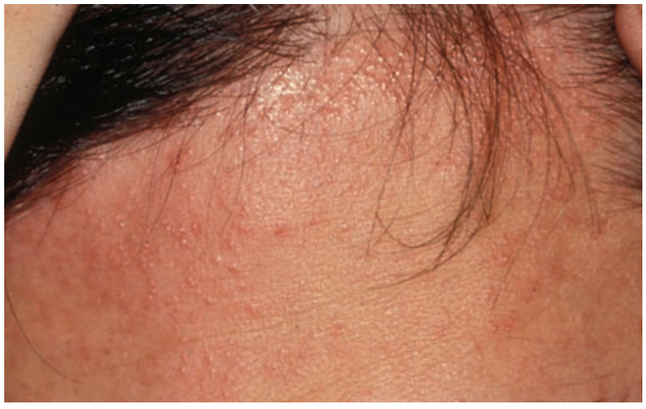
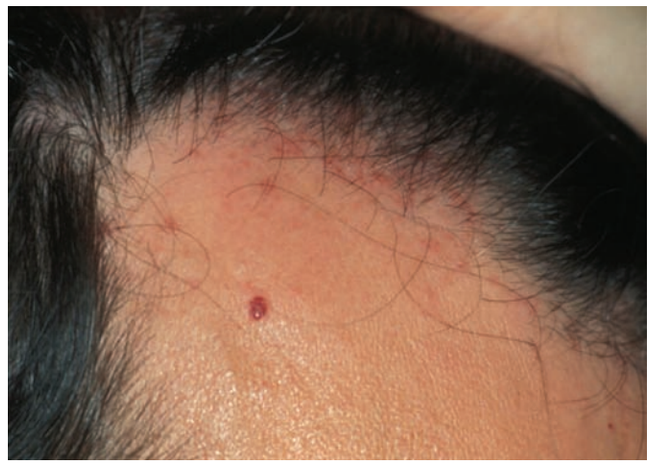


Figure 18.2
Contact dermatitis from topical minoxidil: note typical hairline and ear involvement.



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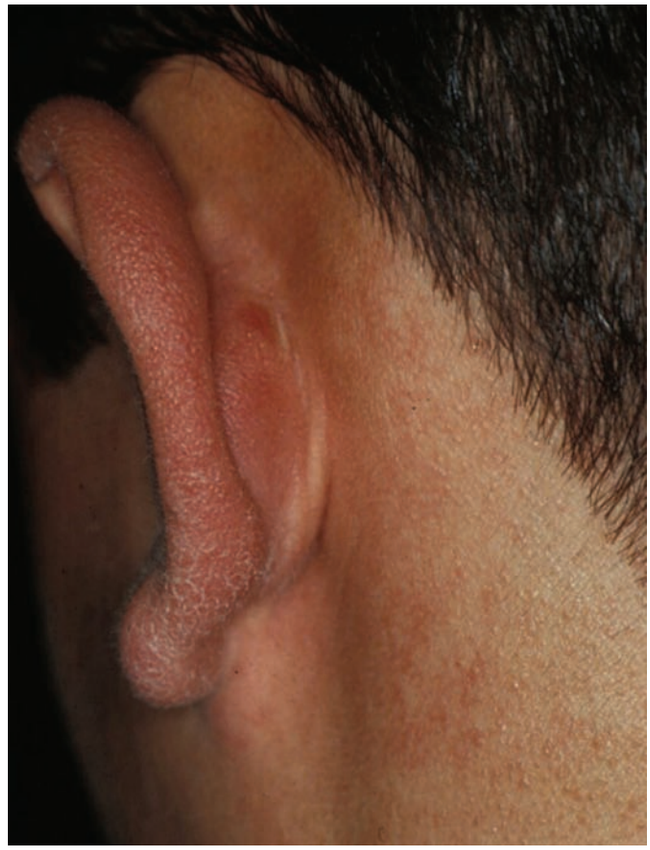
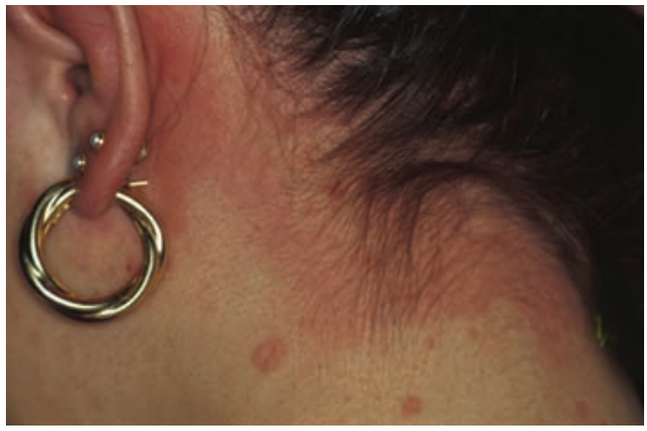


18.4

Figures 18.3, 18.4
Contact dermatitis from topical minoxidil: note typical hairline and ear involvement.



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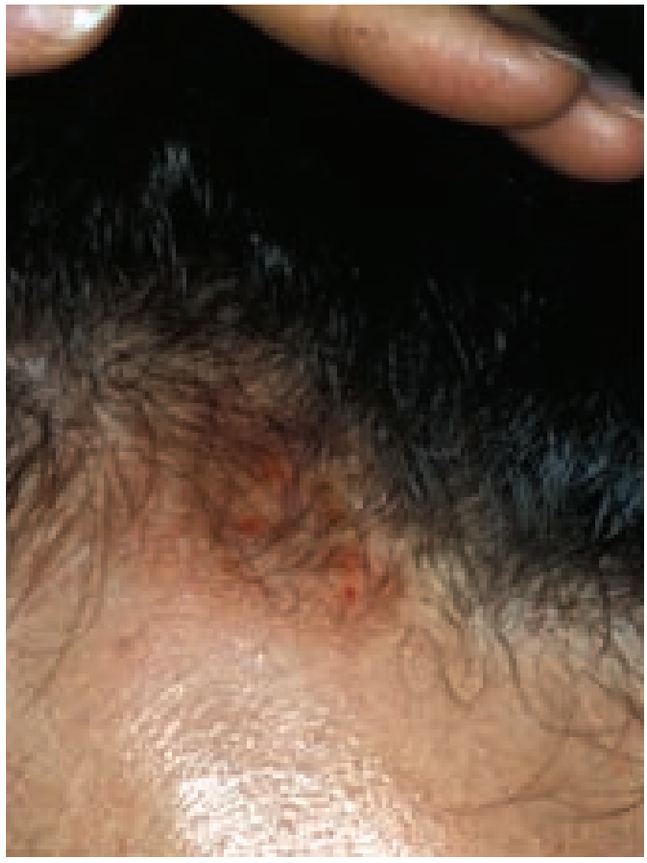

18.6

Figures 18.5, 18.6

Contact dermatitis from topical minoxidil: note typical hairline and ear involvement.

- The non-hairy skin of the hairline and ears may show typical eczematous symptoms (Figures 18.2–18.7). Severe edema of the face, scalp and ears is not uncommon with the use of permanent hair dyes (Figure 18.8). The same occurs in patients with

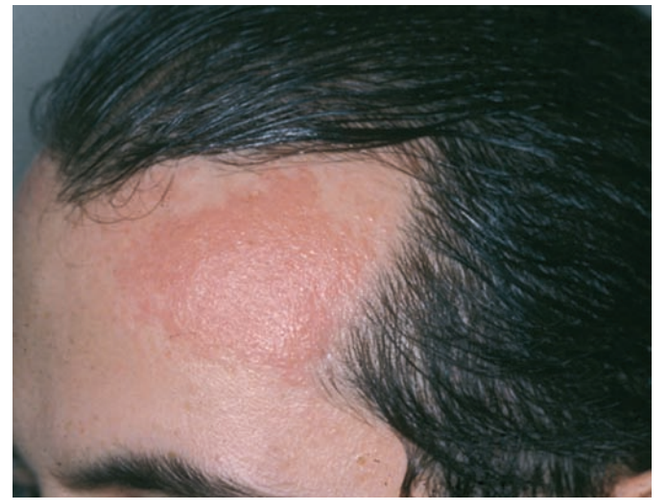

Figure 18.7

Contact dermatitis from hair gel.

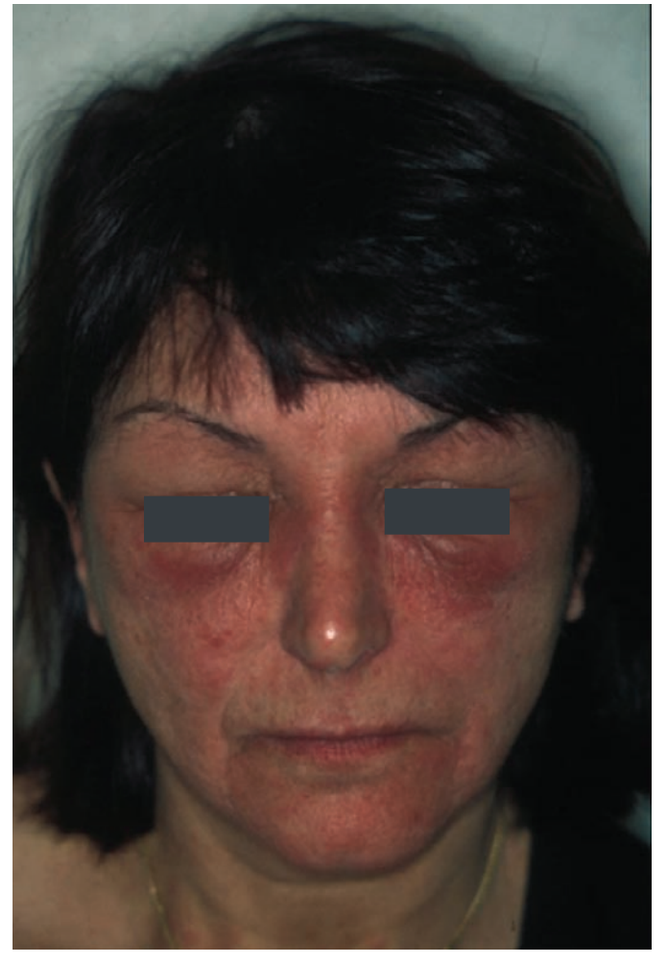

Figure 18.8

Edema of the face due to contact dermatitis to hair dyes.

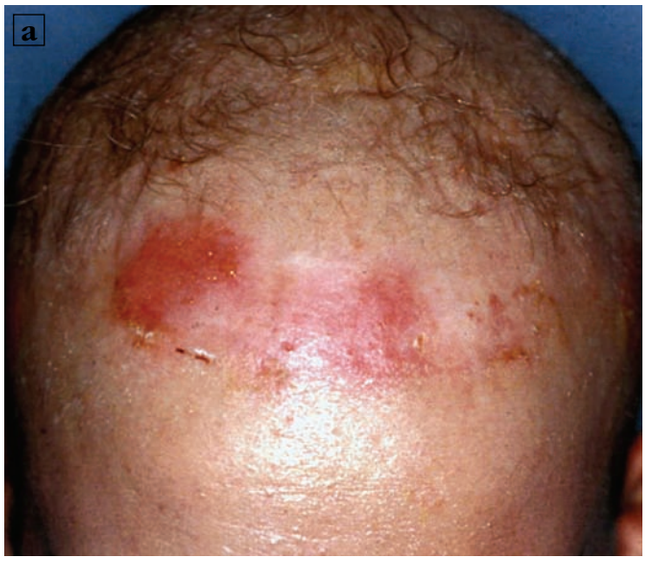

Figure 18.9a,b
Contact dermatitis (a) due to wig-adhesive (b) in a patient with alopecia areata.

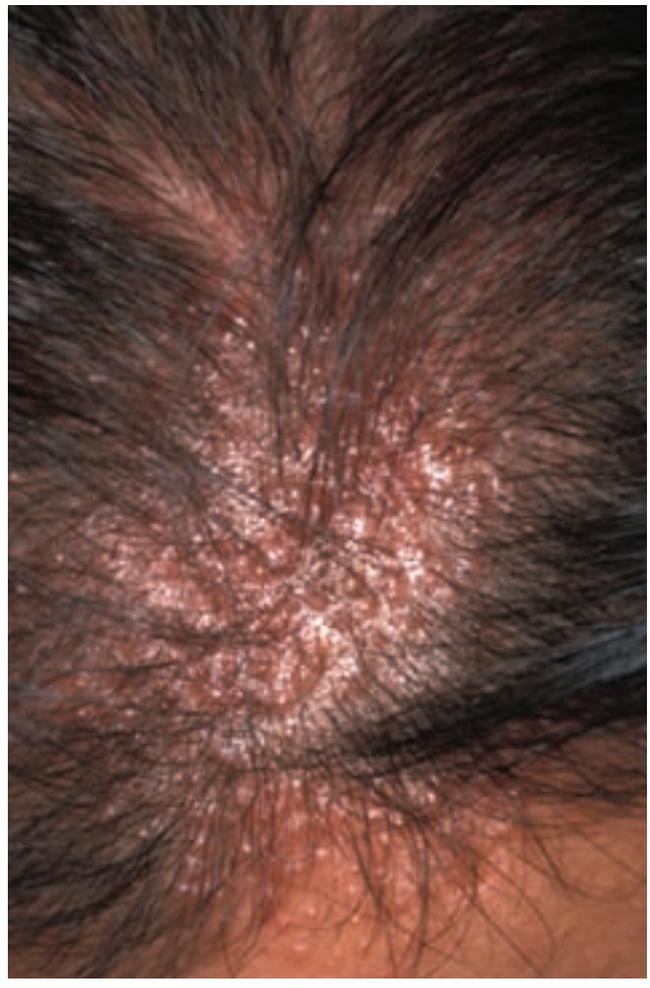

Figure 18.10
Contact dermatitis of the scalp due to hair dyes.

alopecia areata where eczematous changes may occur in the scalp (Figure 18.9a,b).
- Diffuse alopecia associated with a telogen effluvium may occur 2–4 months after an episode of acute scalp contact dermatitis.

Causes

Hair dyes

- *Paraphenylendiamine (PPD), textile dyes.* Most cases of scalp reactions are related to the use of permanent hair dyes and most cases are due to home treatments (Figure 18.10). Beard dermatitis due to PPD is not uncommon in Arabic and Japanese men who dye their beards. Cross-reaction to other related amines found in the permanent and semipermanent dyes are common. Polymorphous-like reactions and severe facial edema are possible. Type I hypersensitivity with anaphylaxis may exceptionally occur. The diagnosis is confirmed by patch testing.
- *Henna* is a very rare sensitizer. Henna is a vegetable coloring that is used alone or in combination with other coloring agents, such as paraphenylendiamine, oil of lemon or beet juice in order to induce more intense coloration. The active component of henna is lawsone, a naphtoquinone.
- *Permanent wave solutions (thioglycolates).* Thioglycolates are strong irritants but uncommon sensitizers in the scalp. Glyceryl mono-thioglycolate is a common cause of hand dermatitis in hairdressers.

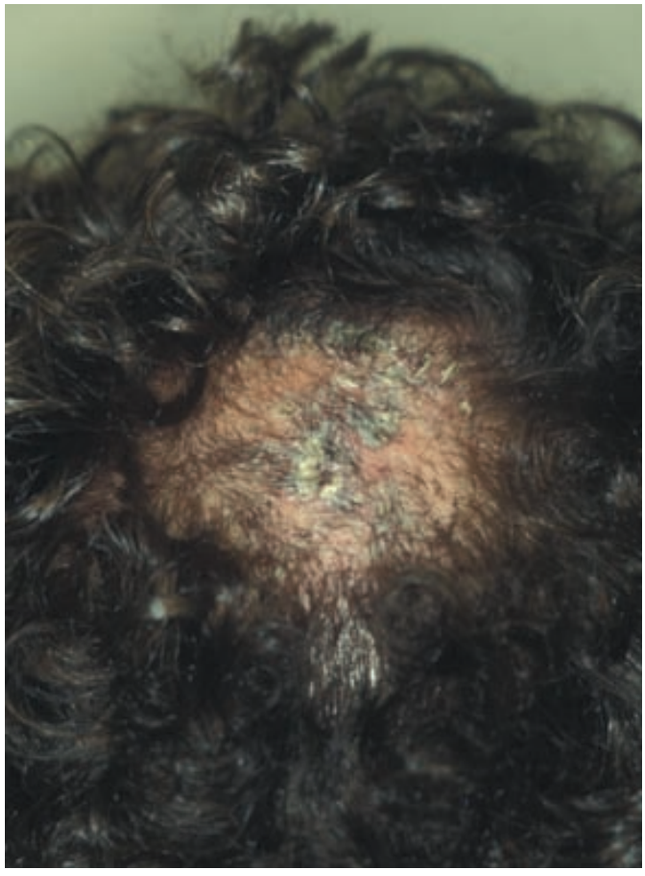

Figure 18.11
Acute contact dermatitis of the scalp due to topical minoxidil.

Fragrances

Sensitization may rarely occur from leave-on hair products such as hair gel, mousses and spray.

Topical drugs

Topical minoxidil. Although sensitization to minoxidil is not rare in our experience (up to 10% of patients), it goes frequently undetected without patch testing. Regular patch testing shows that most irritative reactions (as described by the literature) are actually contact allergy. Symptoms are usually minimal, except for scalp itching. Acute dermatitis is exceptional (Figure 18.11). Patients with scalp itching and scaling during topical minoxidil treatment should undergo patch testing with the minoxidil lotion (2% or 5% in propylene glycole, ethanol and water) as is and propylene glycole 5% in pets. Testing minoxidil in other vehicles yields false-negative results. Although sensitization to propylene glycole is exceptional, PEG400 can be used as an alternative vehicle for minoxidil in these cases.

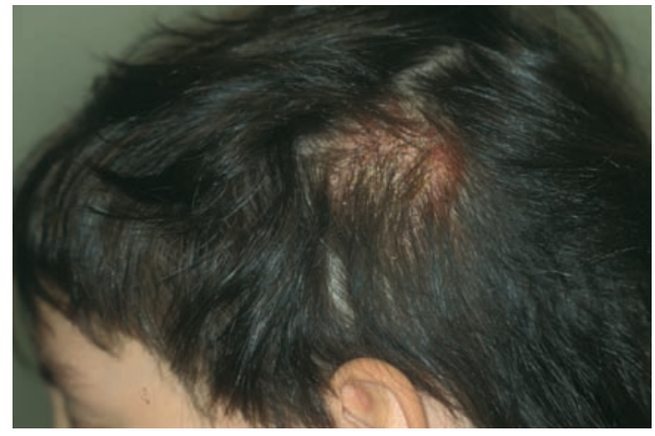

Figure 18.12
Severe contact dermatitis from SADBE.

SADBE. A mild contact dermatitis of the scalp is a goal of topical immunotherapy for alopecia areata. Severe reactions are rarely observed (Figure 18.12).

Other drugs that may rarely cause scalp contact dermatitis include imydazole derivatives, topical steroids, zinc pyrithione.

Nickel

Hairpins and curlers may induce localized scalp dermatitis.

Adhesives

Patients wearing wigs may become sensitized to the glues utilized to fix the prosthesis (Figure 18.9a,b).

Shampoo ingredients

Surfactants

Cocamidopropyl betaine is the most common cause of scalp dermatitis due to shampoos. Sensitization is due to betaine contaminants and has become less frequent in the last few years due to better purification of the raw ingredients.

Preservatives

These include formaldehyde releasers, methyldibromoglytaronitrile, isothyazolinones and parabens. Sensitization is exceptional.

Fragrances

Fragrance sensitization is exceptional.

Diagnosis

Patch testing is necessary for diagnosis.

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19 Papulo-pustular disorders

Papulo-pustular disorders that most frequently affect the scalp are reported in Table 19.1.

Table 19.1 Papulo-pustular disorders of the scalp

Acnes keloidalis nucae
Artificial hair implantation
Chronic acneiform eruptions (acne necrotica)
Chronic non-scarring folliculitis
Dissecting folliculitis
Erosive pustular dermatosis of the scalp
Folliculitis decalvans
Impetigo
Herpetic folliculitis
Keratosis follicularis spinulosa decalvans
Pemphigus/pemphigoids
Pustular eosinophilic folliculitis

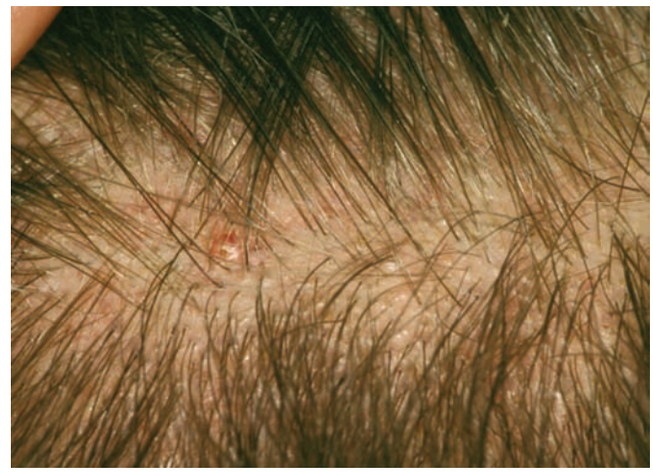
Chronic acneiform eruptions (acne necrotica)

This chronic condition mainly affects adults between 30 and 50 years of age and is probably maintained by self-excoriation.

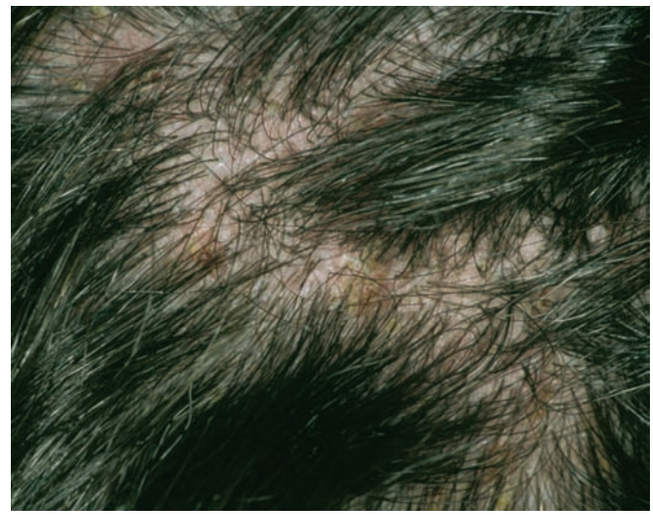
Acne necrotica has two clinical variants: a superficial non-scarring variety (acne necrotica miliaris) and a deep scarring variety (acne necrotica varioliformis).

In acne necrotica miliaris, the scalp presents several relapsing pruritic pinhead-sized vesciculo pustules which are rapidly escoriated to form crusted erosions (Figures 19.1, 19.2).

In acne necrotica varioliformis, lesions are larger, follicular-based and undergo central necrosis with formation of an adherent hemorrhagic crust. Healing produces a depressed varioliform scar.



19.1



19.2

Figures 19.1, 19.2
Acne necrotica miliaris.

Treatment

- Systemic retinoids (E).

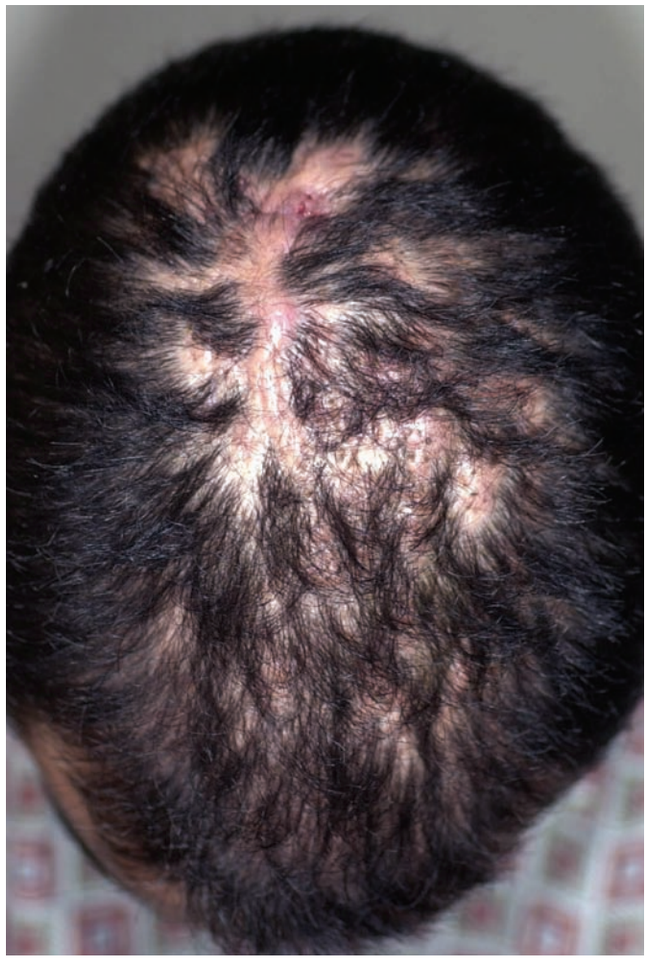
Dissecting folliculitis

This presents as follicular papules and nodules that coalesce into dermal abscesses and sinus tracts (Figures 19.3, 19.4). Scarring alopecia progressively occurs.

It is typical in Afro-American men and it is probably due to an abnormal host response to bacterial antigens. *Staphylococcus aureus* is the most commonly isolated organism.

Treatment

- Oral antibiotics (E)



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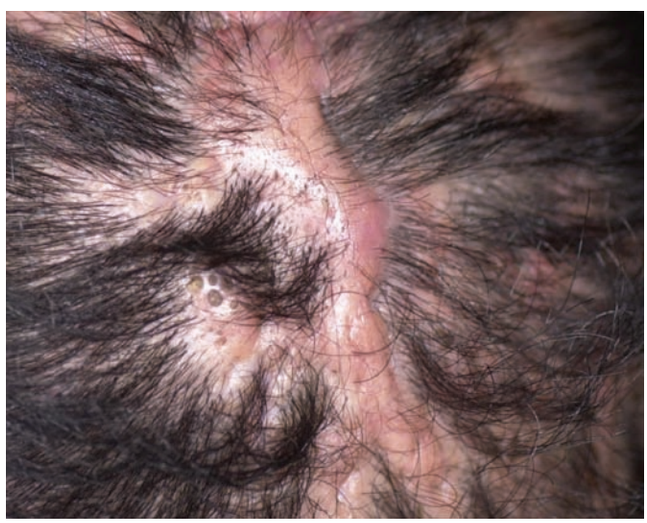


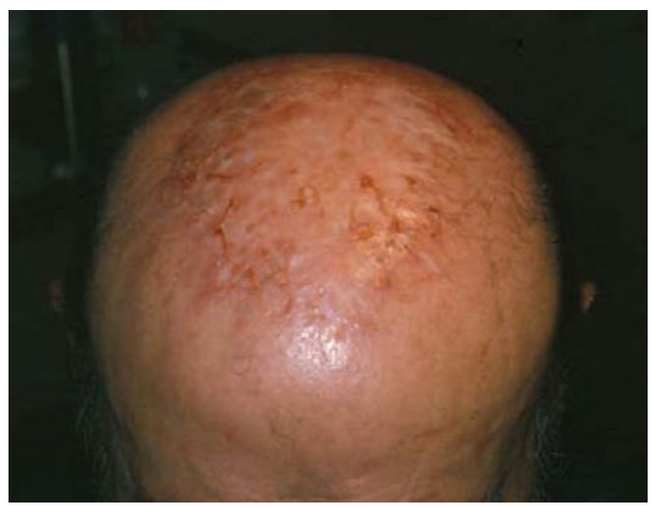
19.4

Figures 19.3, 19.4
Dissecting folliculitis: scalp nodules comedones and sinus tracts

- Corticosteroids: systemic/topical/intralesional (E)
- Systemic retinoids (E).

Erosive pustular dermatosis of the scalp

This condition is almost exclusively seen after the age of 60, and often develops after mechanical or physical trauma to the scalp. The bald scalp is affected with pustules, crusts and erosive lesions that do not heal and often leads to areas of scarring alopecia (Figures 19.5–19.8).



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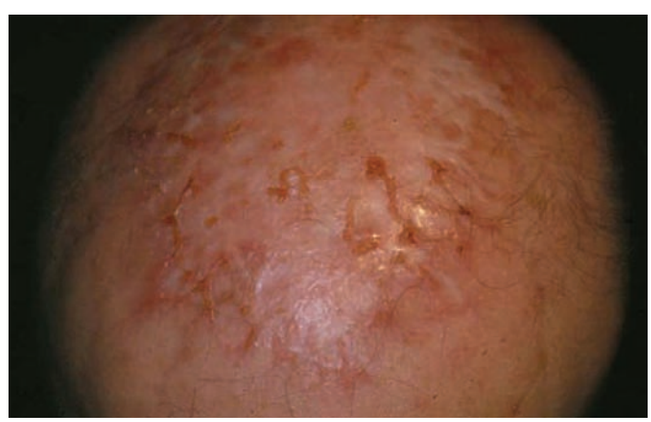
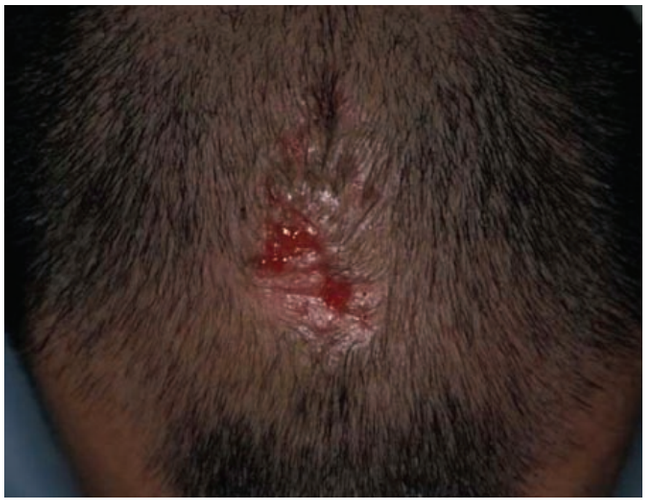


19.6

Figures 19.5, 19.6
Erosive pustular dermatosis of the scalp.



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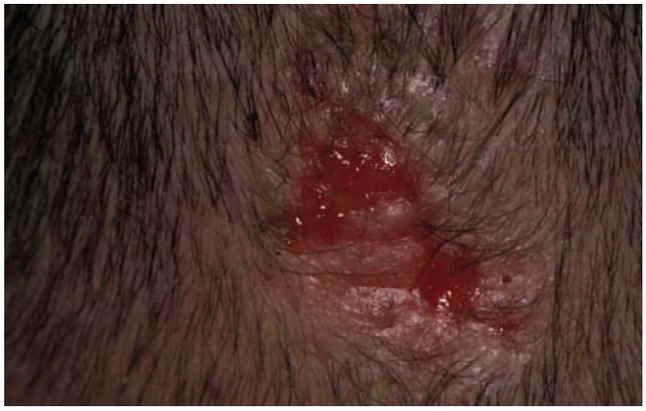


19.8

Figures 19.7, 19.8
Erosive pustular dermatosis of the scalp.

Treatment

- Topical steroids, high-potency (D)
- Topical tacrolimus (E)
- Topical calcipotriol (E).

Pemphigus vulgaris

The scalp may be severely affected in pemphigus vulgaris with erosions, exudative lesions, pustules and crusts (Figure 19.9). Anagen hairs with their sheaths can easily be pulled out from the perilesional scalp. The phenomenon is due to cleavage within the outer root sheath, which is a preferential target for pemphigus antibody as desmosomal proteins are overexpressed in follicular epithelium.

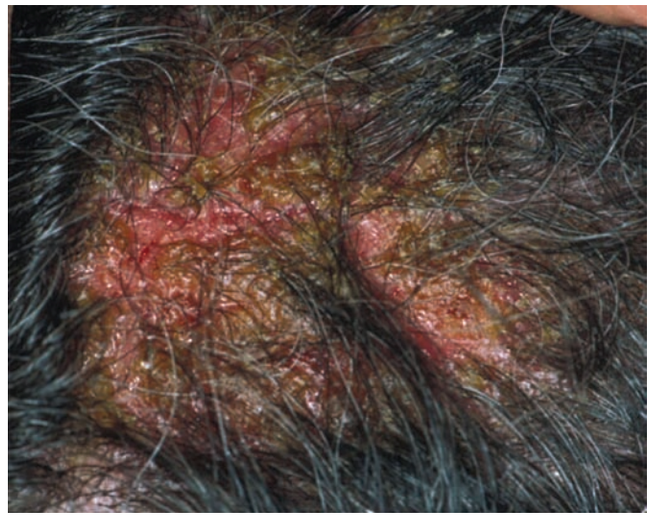
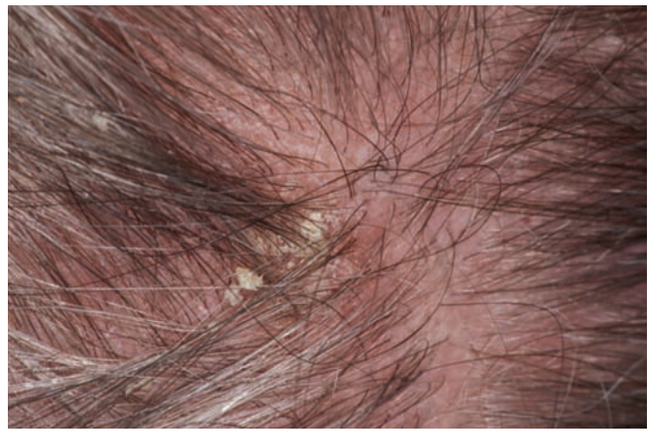


Figure 19.9
Scalp erosions, pustules and crusts in pemphigus vulgaris.



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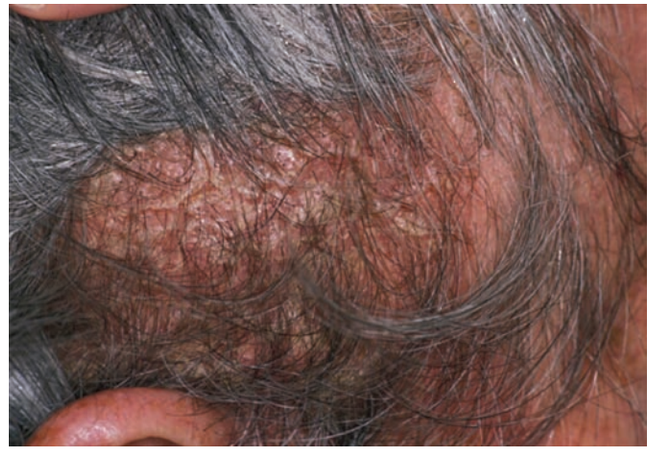


19.11

Figures 19.10, 19.11
Scalp involvement in seborrheic pemphigus.

Cicatricial alopecia is an uncommon outcome. Tufted hair folliculitis can rarely occur.

In seborrheic pemphigus scalp erosions and scales simulate seborrheic dermatitis (Figures 19.10, 19.11).

In bullous pemphigoid the scalp involvement is rare and produces erosions and crusts (Figure 19.12).

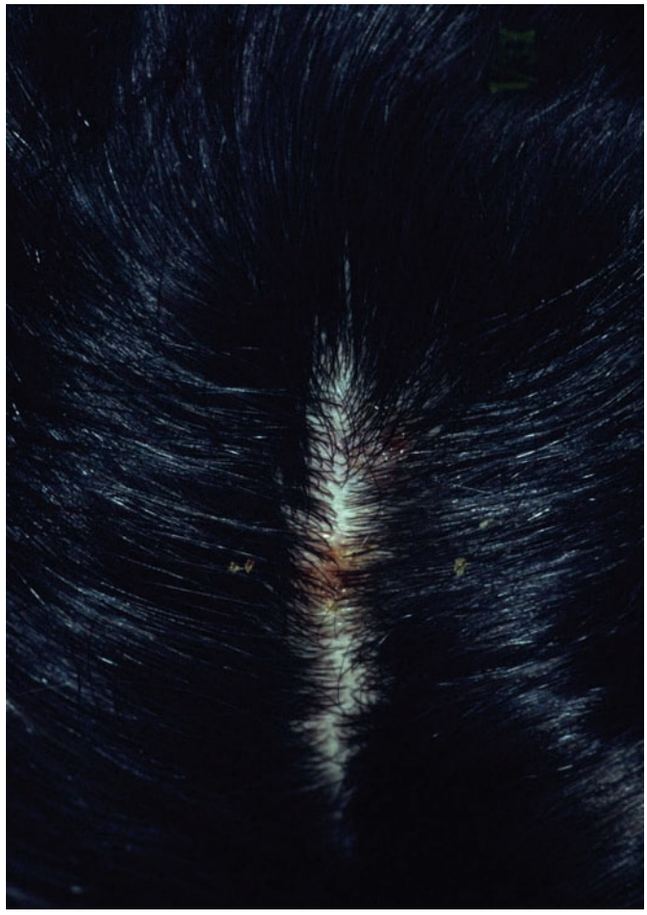
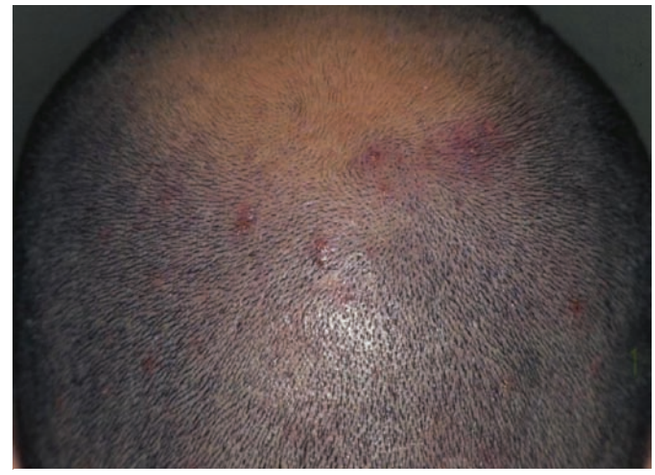


Figure 19.12
Scalp erosions and crusts in bullous pemphigoid.



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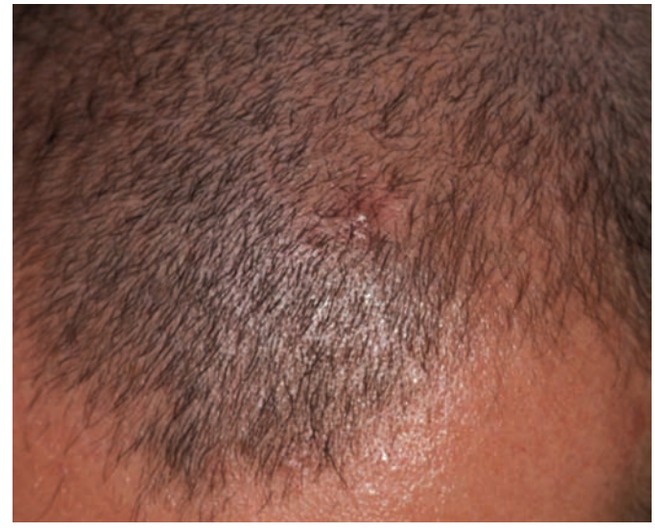


19.14

Figures 19.13, 19.14
Non-scarring folliculitis of the scalp.

Chronic non-scarring folliculitis

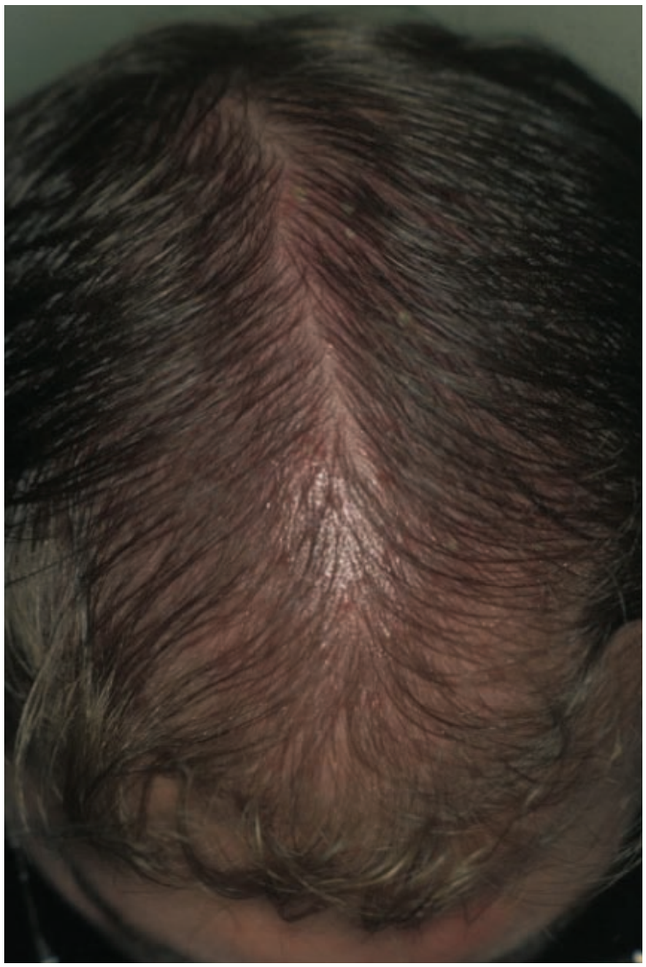
This condition is characterized by recurrent follicular pustules of the scalp without necrosis or residual scarring (Figures 19.13–19.15). Males aged from 20–40 years are most commonly affected. Histopathology reveals neutrophilic folliculitis. Bacteriological examination most commonly reveals *P. acnes*.

Treatment

- Tetracyclines (E)
- Oral retinoids (E).

Artificial hair implantation

Artificial hair implantation is frequently responsible for local post-operative complications, such as pustular eruptions and scarring alopecia (Figures 19.16–19.18). Edema, erythema and bacterial infections usually start a few days after surgery involving the entire treated area. Graft elevation or depression may also occur.



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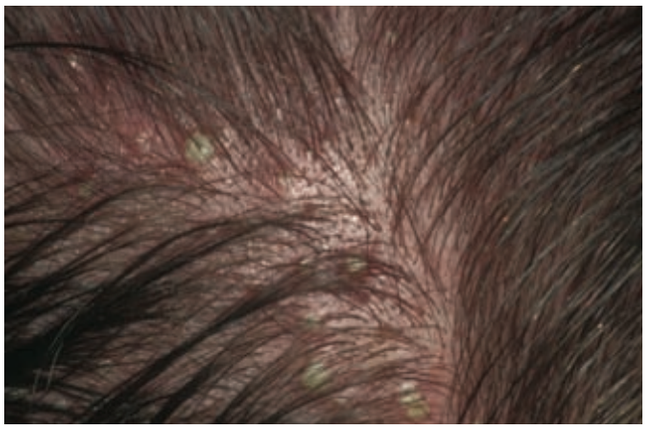

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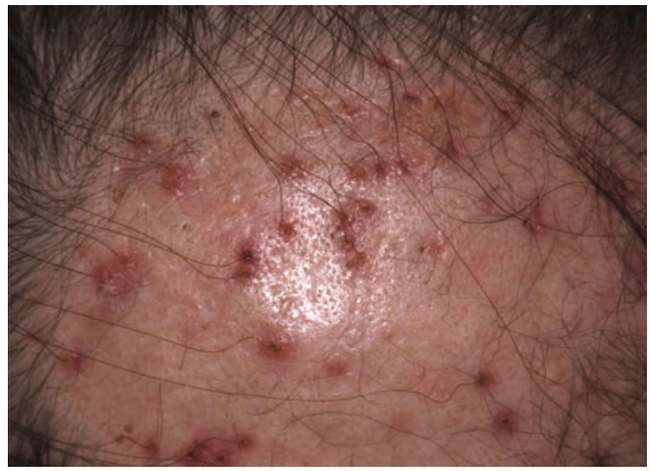
Figure 19.15a,b

Recurrent severe folliculitis of the scalp associated with severe seborrhea. The condition was successfully treated with oral isotretinoin.



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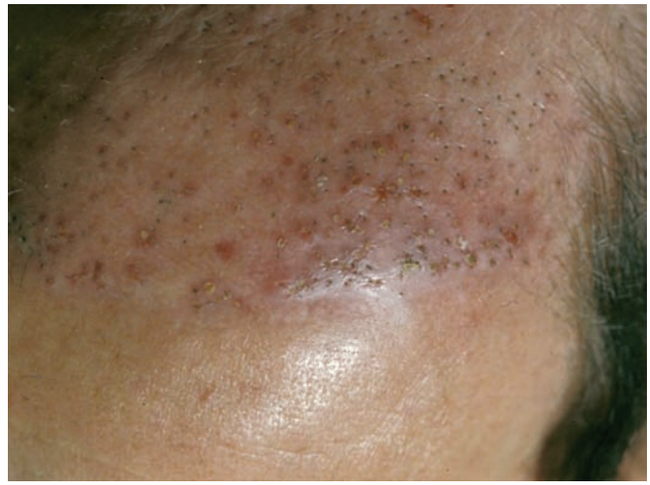

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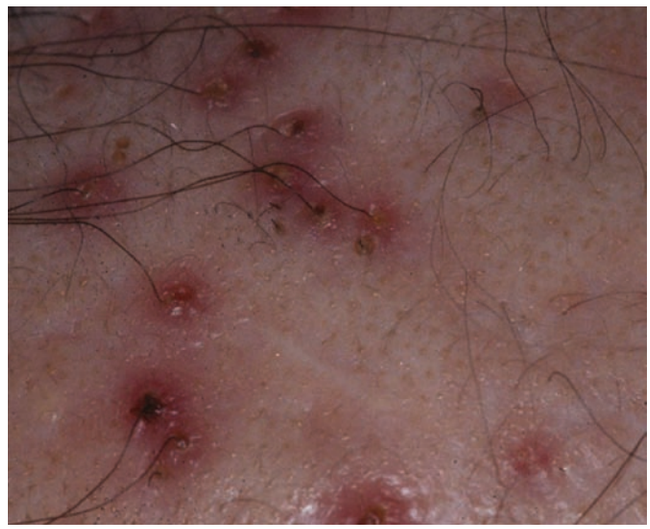

19.18

Figures 19.16–19.18

Scalp erosions, pustules and cicatricial alopecia in a patient with artificial hair implantation.

Clinical manifestations generally persist despite antibiotic therapy, until grafts are removed. Occasionally all the treated area has to be surgically removed.

Impetigo

Impetigo commonly occurs as a result of scratching in children infected by head lice.

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20 Beard disorders

Beard hairs are often lighter than scalp hair. Hair density is lower in Orientals as compared with Africans and Caucasians. The beard grows approximately 0.47 mm/day; hair growth is fastened by stress and slowed by alcohol abuse. White beard hair grows faster than black hair (Figure 20.1). Hair growth is faster in Caucasians as compared with other races.

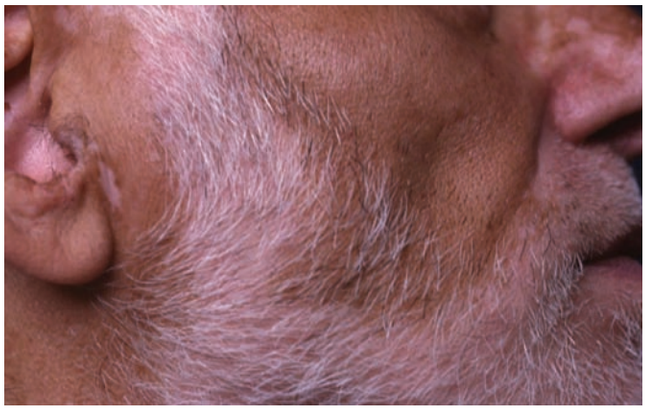


Figure 20.1
Vitiligo: white hair grows faster than pigmented hair.

Pseudofolliculitis of the beard

Pseudofolliculitis of the beard occurs in subjects with tightly curly beard hair that, after shaving, transect the wall of the hair follicle, or re-enter the epidermis 1–2 mm away from their emergency, producing a foreign body reaction (Figures 20.2–20.5). Severe forms occur almost exclusively in blacks, which are very commonly affected. Painful, pruritic and sometimes hyperpigmented papulo-pustular lesions are commonly observed under the chin and on the neck (Figure 20.6). Healing may result in hypertrophic scars and hyperpigmentation.

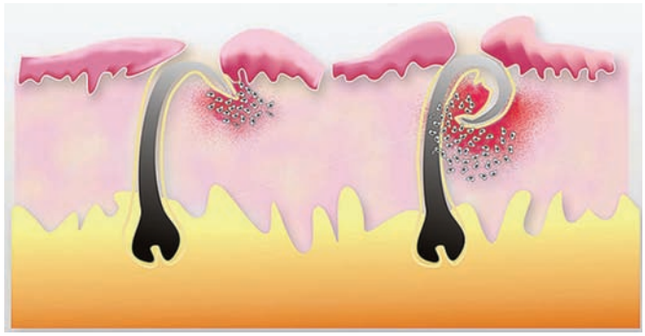


Figure 20.2
Pseudofolliculitis: trans-follicular penetration.

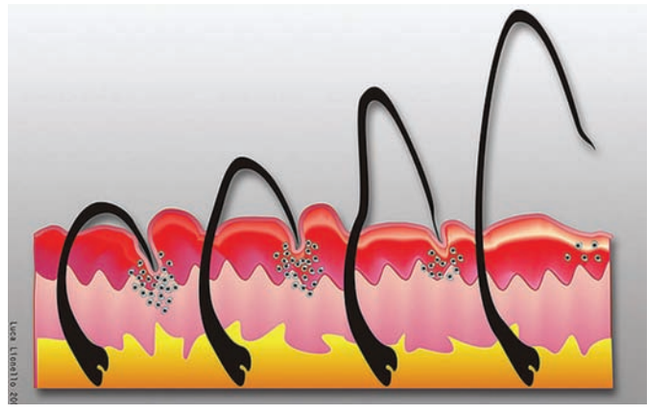


Figure 20.3
Pseudofolliculitis: epidermal re-entering of curly hair.

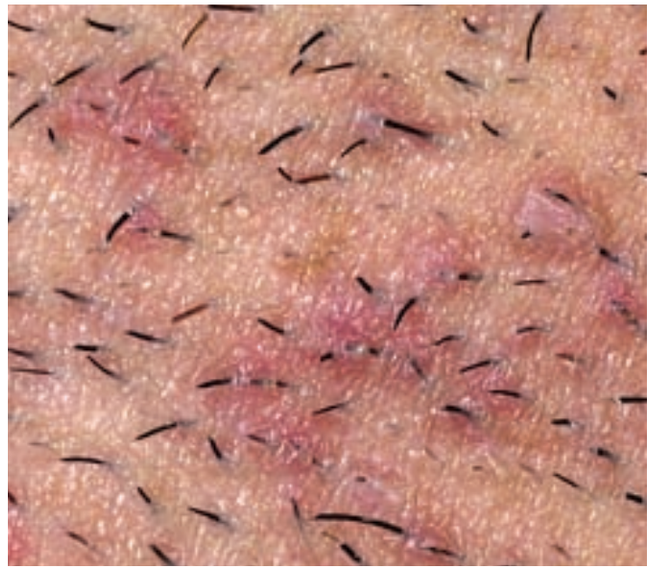


Figure 20.4
Pseudofolliculitis of the beard.

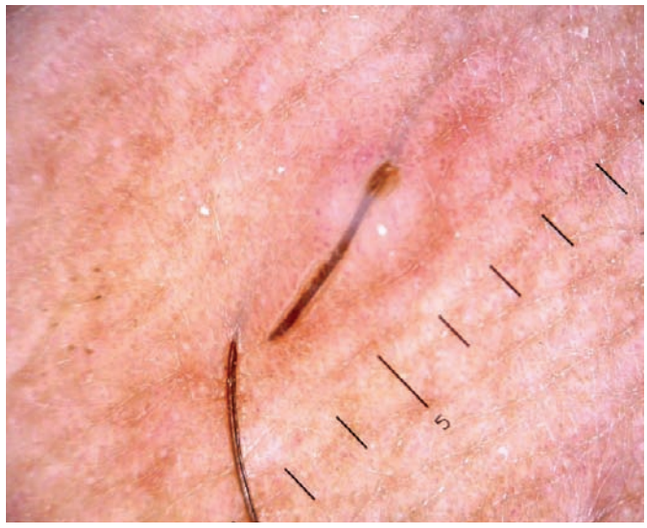
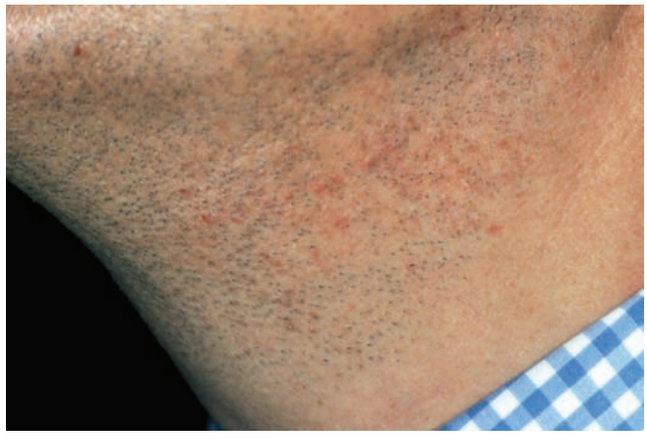


Figure 20.5
Transfollicular penetration.



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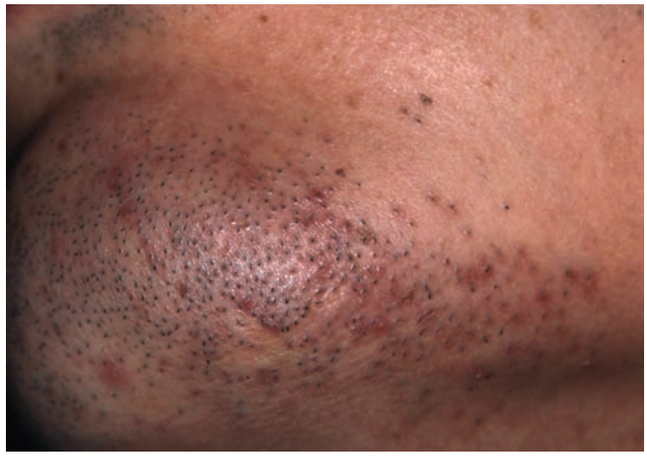


20.7

Figures 20.6, 20.7
Pseudofolliculitis of the beard.

In Caucasians a less severe, but more persistent variety of pseudofolliculitis is observed in subjects with curly and coarse beard hair who shave their beard very close and stretch the skin before shaving (Figure 20.7). Cutting of the hair shaft below the follicular ostium produces embedding of the hair shaft before its emergency from the skin (trans-follicular penetration) with foreign body reaction and inflammation. This variety of folliculitis is frequent in the bikini area and in the legs of women who depilate with techniques that produce rupture of the shaft within the follicle (waxing, tweezing, plucking).

Differential diagnosis of inflammatory lesions of the beard are reported in Table 20.1.

Table 20.1 Differential diagnosis of papulo-pustular lesions of the beard

Acne vulgaris
Bacterial folliculitis
Candida folliculitis
Impetigo
Perioral dermatitis
Pseudofolliculitis
Rosacea
Sarcoidosis
Sicosis
Tinea barbae
Traumatic folliculitis

Management

The condition resolves 2–6 weeks after interruption of shaving in most cases, but recurs when shaving is restarted.

Beard region:

- Soften the hair with warm water before shaving
- Avoid double- or triple-bladed razors
- Avoid close shaving
- Shave in the direction of hair growth
- Utilize electric razors that cut the shaft at a minimum distance of 1 mm from the skin
- Topical antiseptics/antibacterial soaps
- Topical retinoids and/or glycolic acid 5–10%
- Laser epilation.

Other regions:

- Change depilatory methods (wax)
- Laser epilation.

Traumatic folliculitis

This common condition is characterized by escoriations and small erythematous follicular papules without infection or evidence of follicular penetration. Traumatic folliculitis is caused by too close shaving and resolves spontaneously if shaving is stopped for a few days.

Contact dermatitis

The skin of the beard region is often affected by irritative dermatitis favored by the mechanical and chemical traumas associated with daily shaving. Allergic contact dermatitis is uncommon and most frequently caused by after-shaving products, especially fragrances. An exception is represented by Arabic men who are at risk of contact dermatitis due to hair dyeing.

Trichorrhexis nodosa

Trichorrhexis nodosa of the moustache may result by twirling and reverse combing the hair at the lateral edges of the moustache.

Tinea barbae

This is most commonly caused by zoofilic dermatophytes (*T. mentagrophytes* var. *mentagrophytes* and *T. verrucosum*) in agricultural workers who are occupationally exposed to infected animals (Figure 20.8).

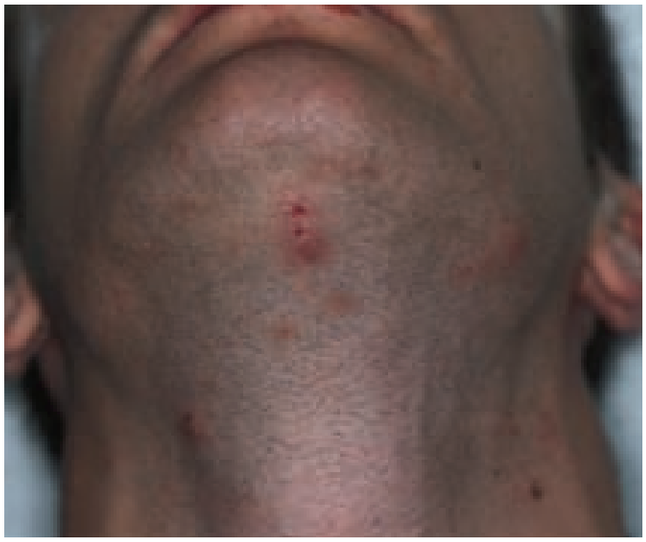
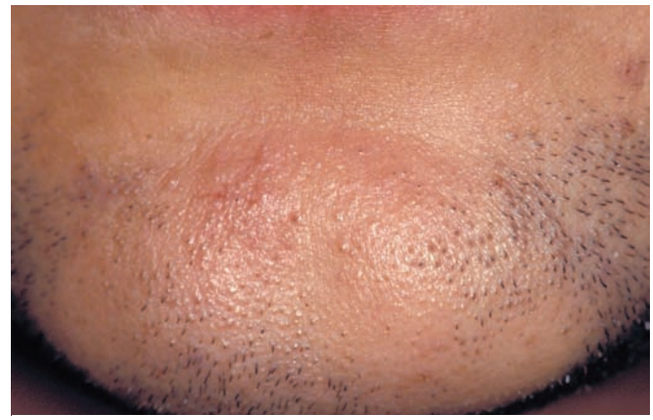


Figure 20.8
Tinea barbae.



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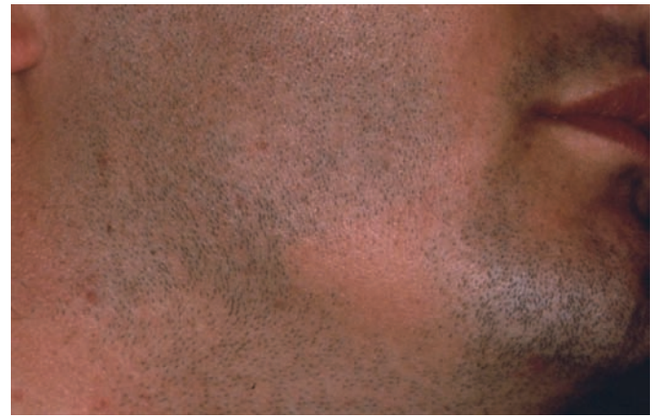

20.10

Figures 20.9, 20.10
Alopecia areata of the beard.

Differential diagnosis with other facial infections, especially from *Staphylococcus aureus*, is suggested by the absence of fever and leukocytosis.

Alopecia areata

Alopecia areata limited to the beard is frequent in young men. The prognosis is usually good, even though the condition tends to persist for many years (Figures 20.9, 20.10).

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21 Ethnic hair disorders

The frequency and clinical aspects of hair disorders vary in different races. Androgenetic alopecia, for instance, is more frequent in Caucasians than in Africans and Asians, whereas African and Asian hair is more susceptible to weathering and fragility.

Hair disorders that are influenced by chemical procedures and hair styling also considerably vary among races, Africans being the most commonly affected by hair damage from styling procedures.

The most important characteristics of African and Asian hair are reported in Table 21.1.

Figure 21.1 shows the shape of the hair shaft in different races.

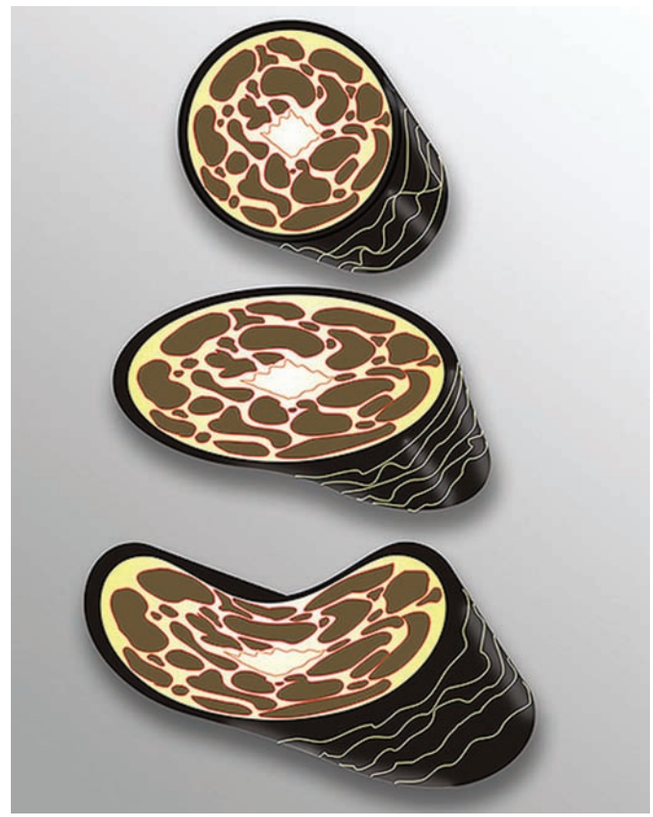


Figure 21.1
Different shapes of the hair shaft. Top: round, Asians. Middle: oval, Caucasians. Bottom: flat, Africans.

African hair

Hair density in Africans is lower than in Caucasians. African hair also has a slower growth rate and is more frequently found in the telogen phase. It tends to be dry since its moisture content may be 5% less than that of Asian or Caucasian hair. African hair is more fragile in both wet and dry conditions and is very prone to damage from weathering, frequently presenting longitudinal fissures, splitting and a high number of broken hairs or hair shafts with knots.

Dandruff is very common due to high sebum production, use of oily hair cosmetics and shampooing habits.

Table 21.1 Hair characteristics in the different races

	Caucasians	*Asians*	*Africans*
Graying	30 yrs	35 yrs	40 yrs
Hair density	227 ± 55 cm^2	174 ± 46 cm^2	190 ± 40 cm^2
	35.5/4 mm punch	–	21.4/4 mm punch
Average length	219 ± 75 mm	250 ± 21 mm	54 ± 22 mm
Hair shaft section	Oval	Round	Elliptic/flat
Hair diameter	40–90 microns	120 microns	40–90 microns, irregular with constrictions
Hair shaft shape	Cylindric	Cylindric	Twisted oval rod
Tensile strength	–	–	Low
Combability	–	–	Low
Hair moisture	–	–	Low
Sebum production	–	–	High
Growth rate	396 ± 55 μm/day	–	256 ± 44 μm/day

Table 21.2 Hair disorders due to hair styling

Women	
• Braiding	Traction alopecia, hair breakage
• Cornrow braiding	Traction alopecia, hair breakage
• Artificial braiding	Traction alopecia, hair breakage, scarring
• Hair lacing	Traction alopecia, hair breakage, scarring
• Straightening Hot combing Relaxing	Hair breakage, chemical burns
Men	
• Box (short sides with high square top)	
• Fade (back progressively shorter from top to bottom)	
Children	
• Ponytail	Traction alopecia, hair breakage
• Braiding	Traction alopecia, hair breakage
• Cornrow braiding	Traction alopecia, hair breakage

Hair disorders due to hair styling (Table 21.2)

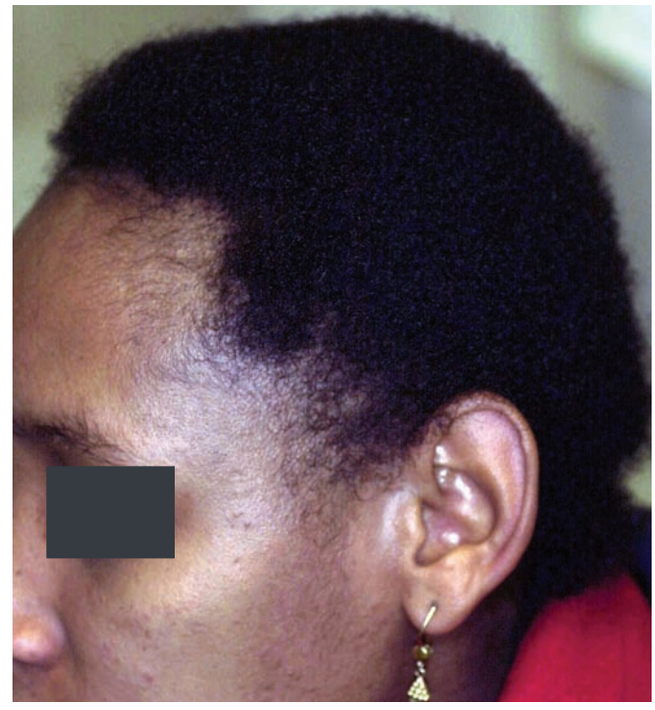
Traction alopecia (see page 103)

Traction alopecia is a common complication of tight hair style such as braiding and lacing and typically produces a frontal and lateral pattern of hair loss due to excessive traction (Figure 21.2). Permanent scarring is a possible sequela.

Hair breakage

Hair breakage may result from traction due to hair styling or to chemical damage during hair straightening or relaxing. Hair straightening can be obtained by heat or chemicals.

- Hot combing is a variety of hair setting acting on the hydrogen bonds and permitting temporary straightening of curled African hair. After application of an emollient (usually petrolatum) the tuft is combed with a hot metal comb that reaches very high temperatures. The hair remains straight until the next washing.
- Eighty per cent of African-American women use hair relaxers. African hair requires stronger chemicals than Caucasian hair and products for African hair often come in creams or gels in order to achieve a better contact with the hair shaft. Hair relaxers are highly alkaline products and are frequently responsible for hair or scalp damage. The breakage occurs



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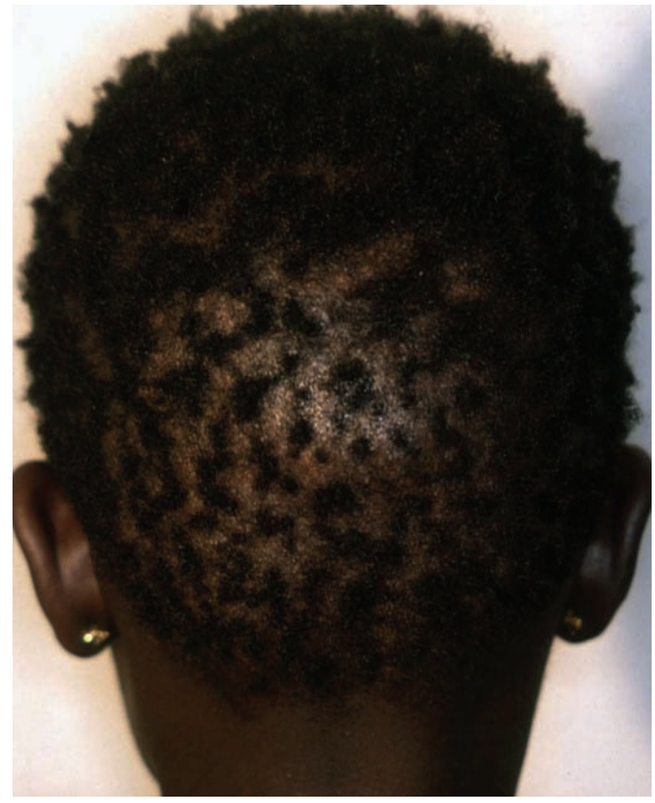

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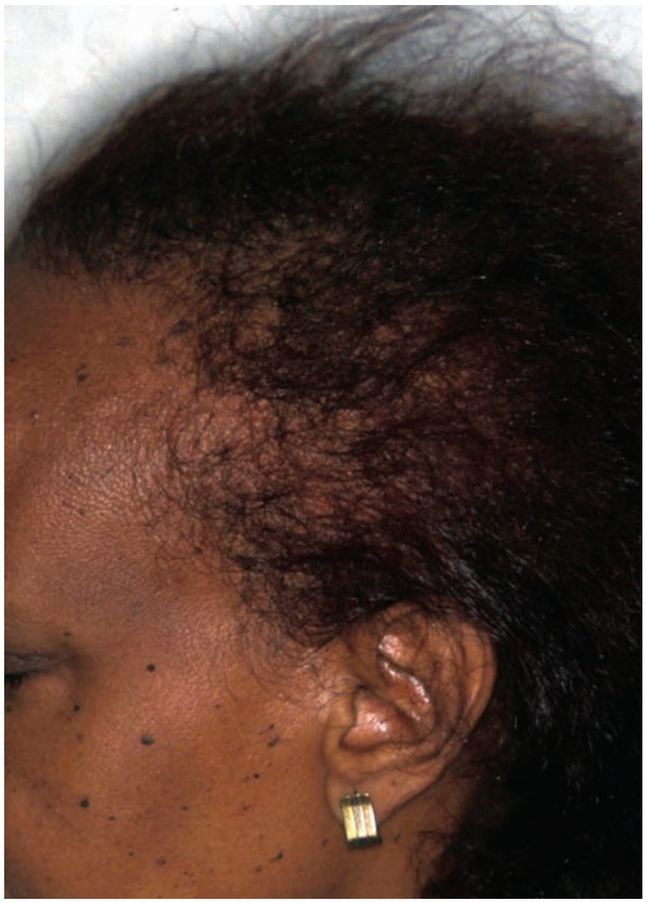
Figure 21.2a,b
Traction alopecia.



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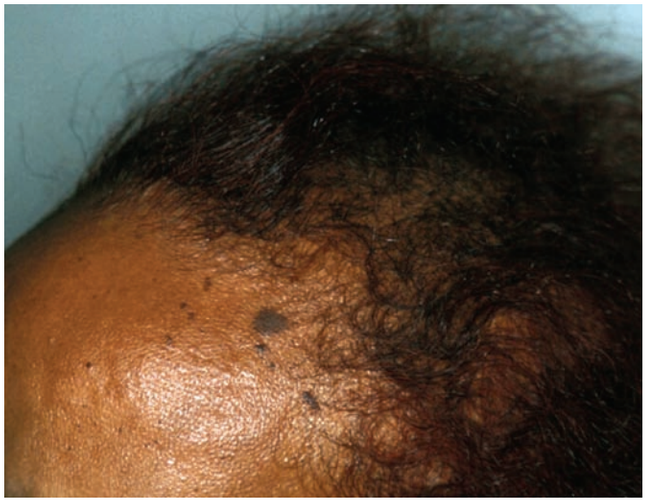

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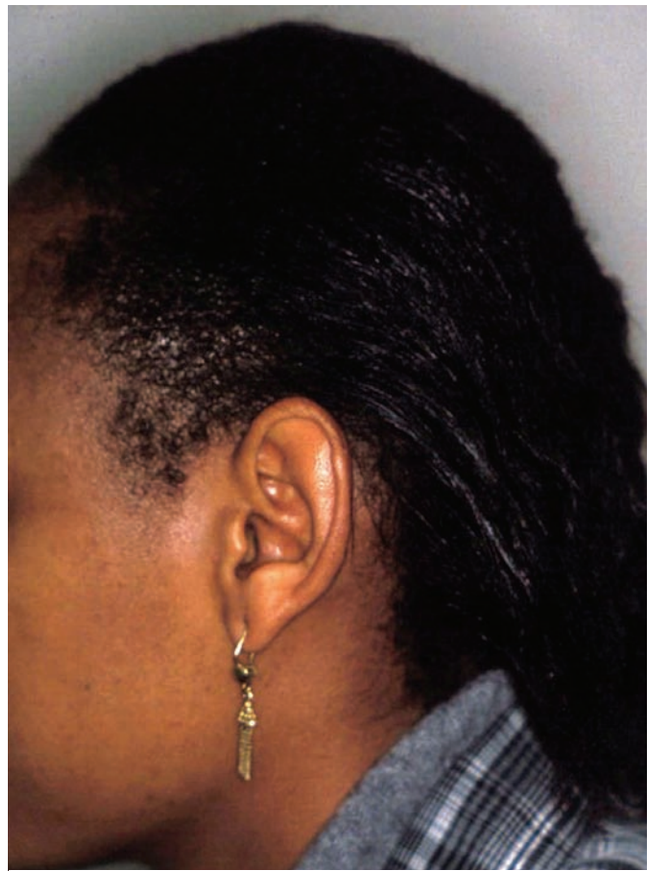

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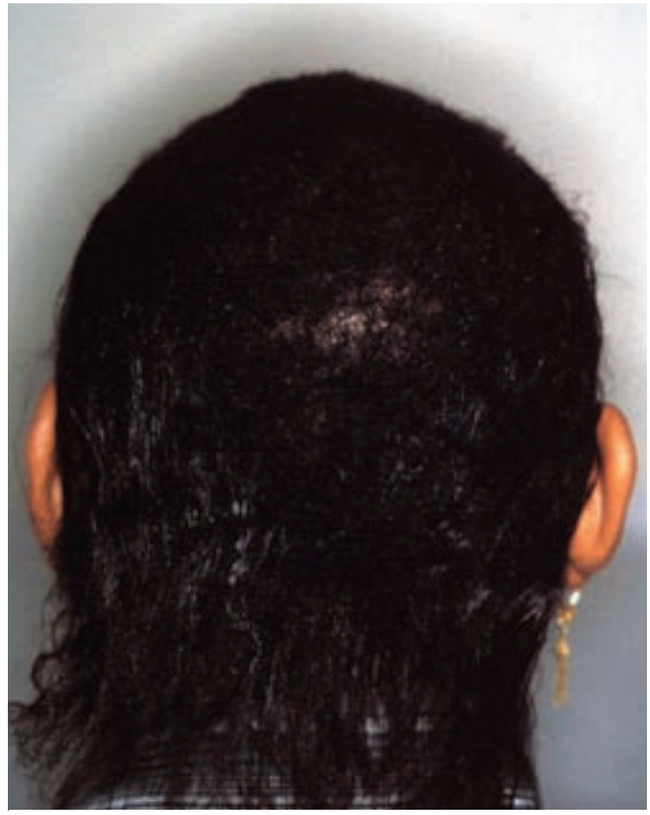
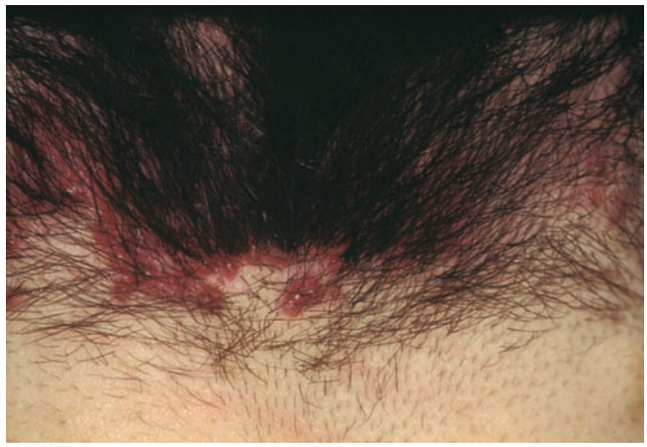


21.6

Figures 21.3–21.6
Alopecia due to hair breakage after repetitive hair relaxing.



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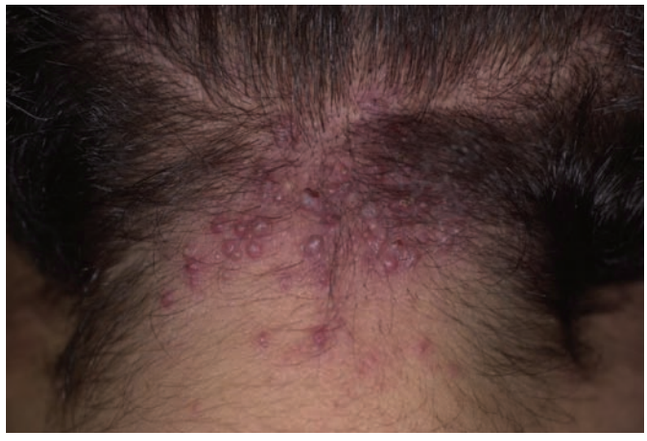

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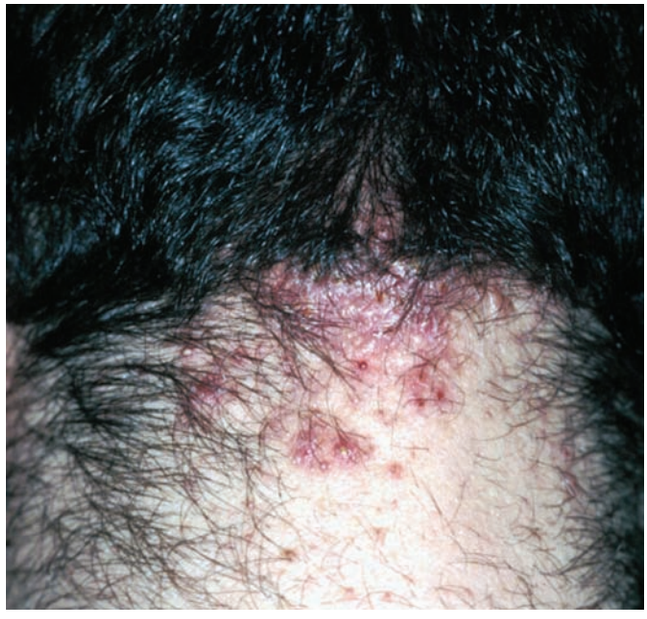

21.9

Figures 21.7–21.9
Acnes keloidalis nucae.

at the junction of the new growth and the previously relaxed hair, usually 1 week after the procedure. Hair breakage is usually seen in the sub-occipital and neck area or along the frontal hairline (Figures 21.3–21.6).

- Africans often re-perm, wave or curl their relaxed hair in order to obtain a more fashionable hair style. Hair damage by curling tends to occur more distally along the shaft.

Acnes keloidalis nucae

This condition commonly affects the occipital scalp and posterior neck and produces scarring alopecia. The affected region shows relapsing painful papular and pustular lesions that produce hairless keloidal plaques (Figures 21.7–21.9). Treatment is difficult and the condition typically causes severe cosmetic damage.

Hot comb alopecia – follicular degeneration syndrome – central centrifugal scarring alopecia

This condition typically affects middle-aged African women and was originally thought to be caused by heat and chemicals utilized for hot combing. This is now discussed since the condition may also occur without history of hot comb usage. Scarring alopecia starts from the vertex and spreads symmetrically in a centrifugal direction, similar to the progression of female androgenetic alopecia.

Pseudofolliculitis barbae (see page 156)

This is the most common dermatological complaint in Africans, affecting up to 50% of men who shave regularly.

Management

- Establish styling habits and cosmetic procedures taken in the last year.
- Since African hair is very dry and fragile, shampooing should not be too frequent.
- Utilize shampoos that contain humectants and mild cleansing agents.
- Suggest conditioner products containing moisturizing and coating agents, such as cationic polymers (polyquaternium 10) and silicone oils (dimethicone).

Asian hair

The hair density of Asians is lower than that of Caucasians. The Asian hair shaft is round in shape, thick, robust and straight. Asian hair is very difficult to perm and to dye and for these reasons is often damaged when submitted to these procedures due to the necessity to use high concentrations or longer exposure to chemicals. Styling may be difficult when the hair is kept short since the hair does not curl and tends to have a spikey appearance.

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22 Hair cosmetics

Combing/brushing

- Combing and brushing damage hair. Metal or wood combs and combs with irregular teeth may be particularly hazardous, the best instruments being soft plastic combs which bend when caught in tangled hair. Caucasian hair is more easily damaged when combed wet. The opposite applies to African hair.
- Back-combing considerably damages the hair shaft since it produces uplifting of the cuticular scales that becomes fragile with exposure of the cortex.
- Excessive brushing can induce trichorrexis nodosa and trichoschisis. Aggressive brushing may also induce premature loss of telogen hair characterized by roots with an attached epithelial tail indicating they were still attached to the scalp (premature teloptosis, see page 57).
- Hair pins, clasps and rubber bands break the hair and may cause hair fracturing alopecia.

Hair cosmetics can be distinguished into two main groups:

1. Cosmetics that produce only temporary changes to the hair, such as shampoos, conditioners, hair sprays, temporary colors.
2. Cosmetics that produce a permanent change to the hair shaft such as permanent waves, relaxers, bleaches, permanent colors.

Shampoos (see Table 22.1)

Commercially available shampoos have a pH of around 5. Shampoos should remove dirty particles and sebum from the hair shaft and at the same time make the hair shiny, soft and easy combable.

Typical shampoo ingredients are foaming agents and sequestering agents, which prevent the formation of insoluble soaps (scum) on the hair and scalp.

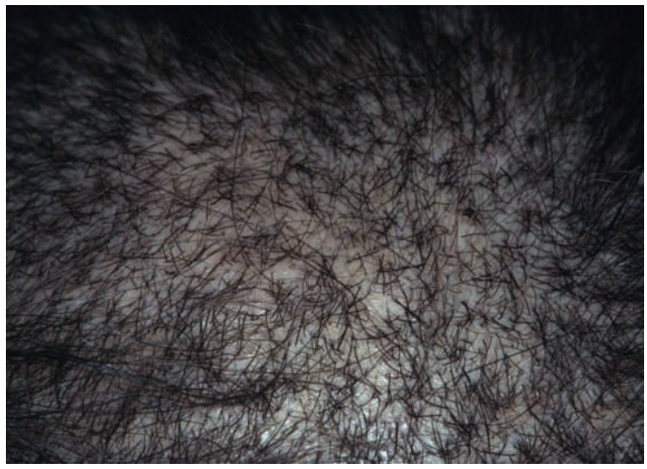
Shampoos can also deliver selected ingredients such as antifungals or keratolytic agents to the scalp and the hair follicle. Shampoos by themselves are very safe products. Skin and eye irritation may occur, but contact dermatitis is rare, due to water dilution and short contact time.

Conditioners

Conditioners improve manageability, gloss and antistatic properties of the hair. Common ingredients of conditioners include dimethicone, simethicone, glycerin, propylene-glycol, quaternized cationic derivatives, cationic polymers, polyvinyl-pyrrolidone, stearalkonium-chloride and hydrolyzed animal proteins, which are attracted to keratin and can temporarily improve split ends.

Table 22.1 Shampoo indications and compositions

Shampoo	*Indication*	*Composition*
Detergent shampoo	Everyday shampoo	Mild detergents, no conditioner
Baby shampoo	Children	Detergents non-irritating to the eyes (betaines)
'2 in 1'	Dry-damaged hair	Mild detergents, conditioner
Soap shampoo	Oily hair	Strong detergents, minimal conditioner
Deep cleaning shampoo	Remove hair styling products	Strong detergents
Medicated shampoo	Itching, scaling scalp	Tar derivatives, salicylic acid, sulfur, selenium sulfide, zinc pyrithione, poly-vinyl-pyrrolidone, iodine complex, menthol, antifungals
Professional shampoo	Before/after chemical process	*Bleaching*, anionic to neutralize alkalinity after bleaching *Dyeing*, cationic to reduce cuticular swelling and prevent color fading



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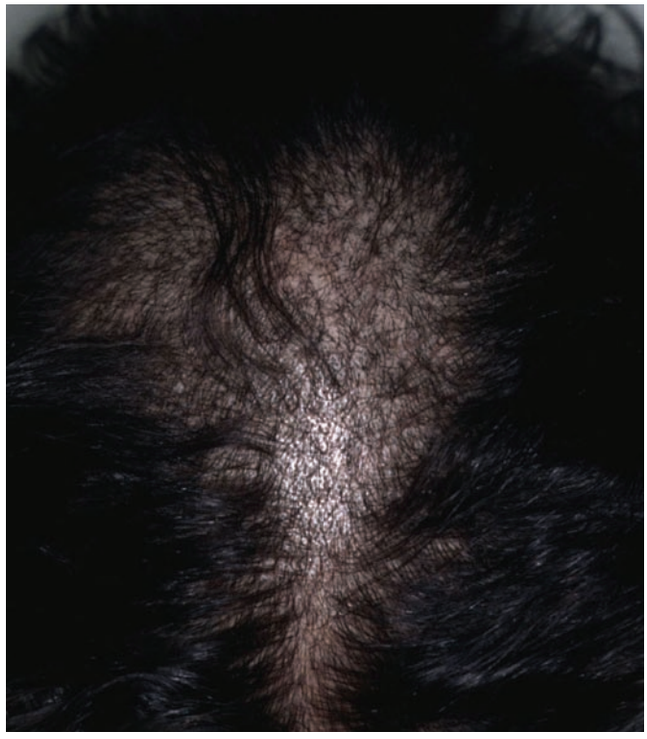

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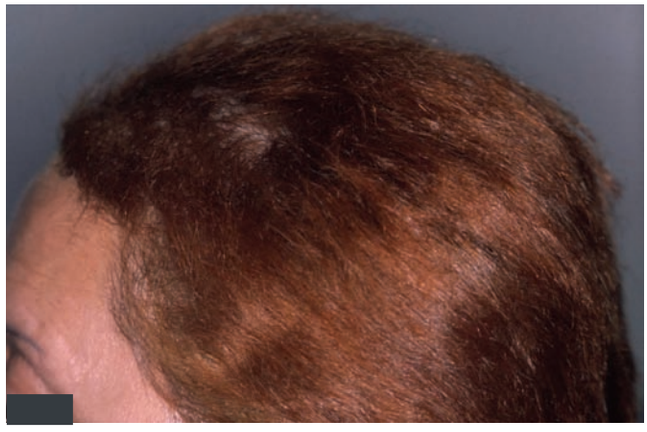

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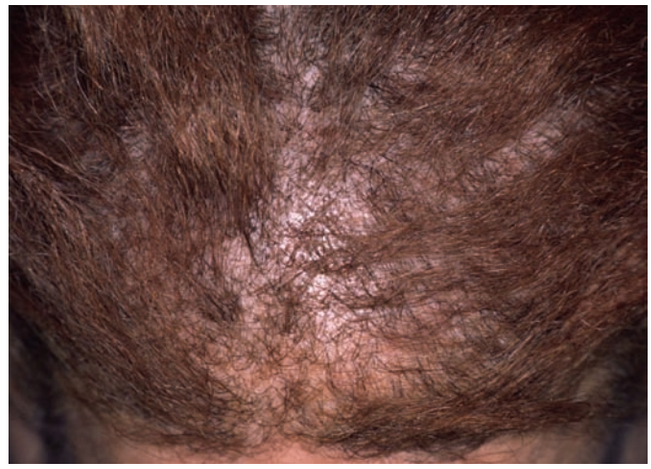

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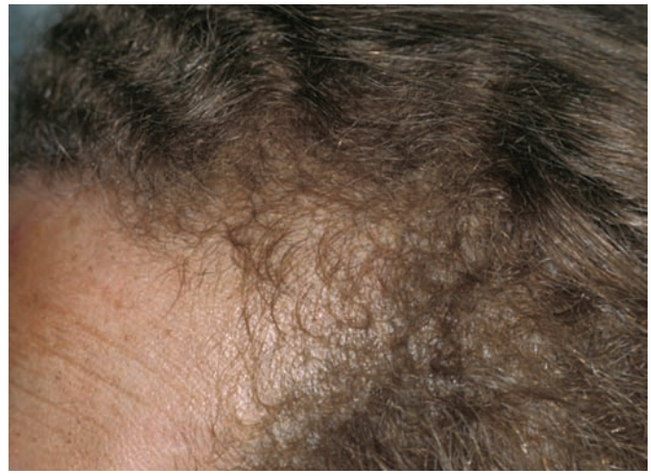


22.5

Figures 22.1–22.5
Severe hair breakage after permanent waving.

Hair lotions

Cosmetic lotions are often claimed to promote hair growth, even though their ingredients do not have any proven efficacy. Massage of the scalp can induce kenogen follicles to re-enter anagen and produce temporary hair regrowth.

Over-the-counter hair cosmetics containing human bovine placenta constituents have been associated with premature sexual development in Afro-American girls.

Permanent waving

This procedure manipulates the hair shape by breaking the disulfide bonds between the keratin filaments and

then reforming new bonds in different positions. A large number of bonds are, however, lost with the procedure, resulting in permanent reduction of the hair strength.

- An alkalyn perming lotion is first applied to the hairs that are wrapped around rollers.
- When the keratin linkages are broken, a neutralizing lotion containing an oxidizing agent is applied in order to remake new disulfide bonds in the new shape.

The waving procedure is a very delicate process since while the disulfide bonds are broken, the hair shaft is vulnerable and can be easily broken. Factors that may produce hair breakage include sudden changes in the temperature and mechanical trauma. Hair damage is usually caused by overperming, poor neutralization or formation of new incomplete bonds due to shampooing the night or a few days after the procedure. Hair breakage occurs close to the scalp and usually happens a few days after perming (Figures 22.1–22.5).

Hair relaxing

This procedure is utilized to straighten curly hair. The process is similar to the waving process. Complications are more frequent due to the fact that hair is manipulated when the bonds are broken. African hair is more fragile than Caucasian hair and is difficult to straighten without damage.

Hair coloring

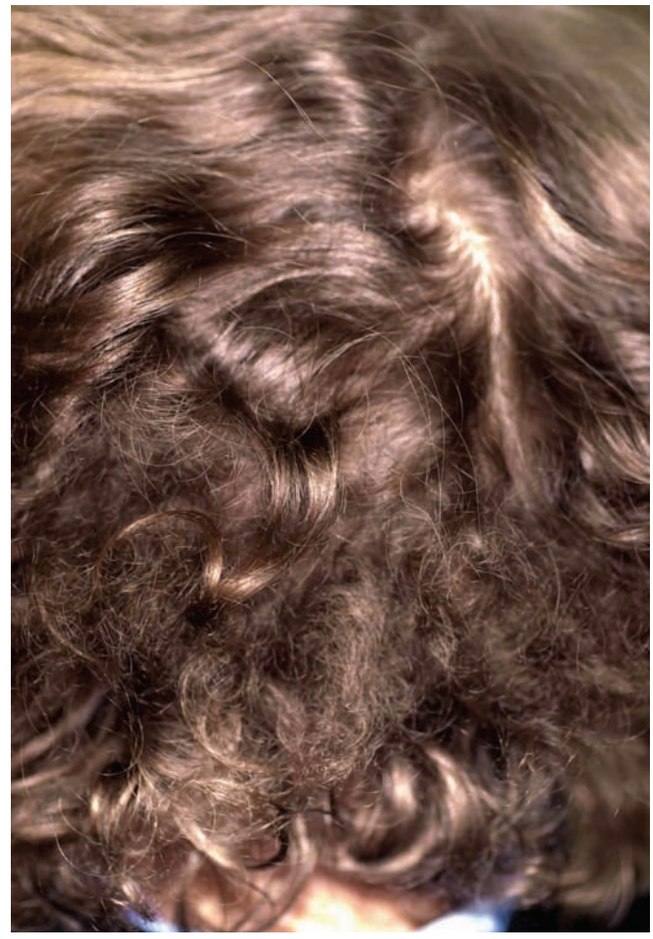
Although hair coloring may contain mutagenic and carcinogenic compounds, epidemiological studies have not shown an increased risk of cancer, including breast cancer, in hair coloring users.

Contact dermatitis is a well-known side effect of hair coloring products which contain strong sensitizers such as PPD and other aromatic amines. Scalp contact dermatitis may be followed by telogen effluvium (see page 145).

Hair abnormalities due to improper hair cosmetic procedures

Hair knotting (trichonodosis)

In trichonodosis the hair shaft presents a single or double knot. The hair tends to splinter and fracture. The condition, which is more common in patients with curly hair, usually involves one or a few hair shafts and is caused by trauma such as combing, brushing and scratching.



22.6

22.7

Figures 22.6, 22.7
Reversible matting of the occipital scalp.

22.8

22.9

Figures 22.8, 22.9
Irreversible hair matting.

Matting of hair

Matting describes reversible or irreversible tangling of locks of hair, which stick together.

Self-induced matting characterizes the rasta hair style, where tufts of matted hair are deliberately kept long.

- Reversible matting. The occipital hair of subjects with hair weathering is frequently affected by matting when friction is applied, such as during prolonged bed rest (Figures 22.6, 22.7). Although tangling can be solved with conditioning and gentle combing, it often tends to recur at the same site.
- Irreversible matting. A mild degree of irreversible tangling is common in individuals with long, damaged hair (Figure 22.8) and may require cutting of small hair tufts (Figure 22.9). Severe hair matting is, on the other hand, a very rare event which usually occurs suddenly and may involve most of the scalp hair. The etiology remains unclear in most cases and cutting the tangled hair is usually necessary.

Factors that may concur in producing hair matting include:

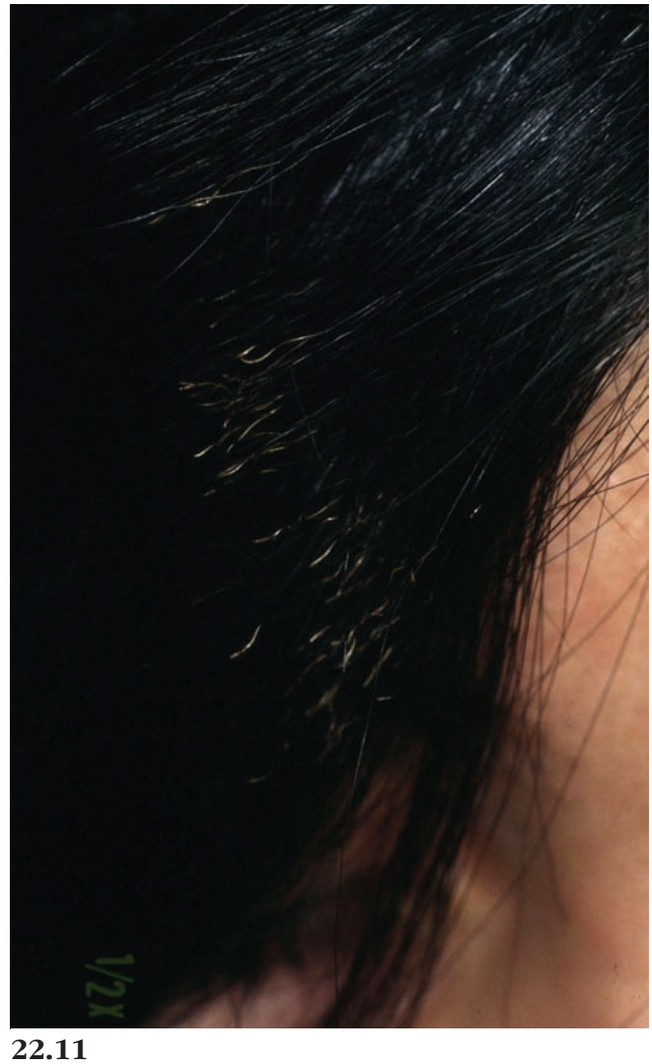
- Mechanical trauma (behavior problems, friction during shampooing, scratching)
- Humidity
- Hair weathering.

Most cases of acute matting occur immediately after shampooing with cationic surfactants.

Bubble hair

When wet hair is exposed to temperatures higher than boiling water temperature, sudden evaporation of water may occur with formation of cavities filled with steam within the shaft. The boiling water softens the keratins and makes the hair easily breakable. Common causes of bubble hair include: hair dryers that have not been properly cleaned, hot curling irons, electric rollers.

22.10



22.11

Figures 22.10, 22.11
Bubble hair.

Hair breakage is usually more evident in the occipital and parietal region with hair broken 1–4 cm from the scalp. Affected hair is often lighter in color (Figures 22.10, 22.11). Microscopic examination of the hair shaft reveals bubbles of various size, giving a 'Swiss cheese' appearance to the hair.

Hair weathering

See page 32.

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23 The hair in systemic disorders

Hair in forensic medicine

DNA typing can be performed on hair samples and dandruff scales

Drugs, chemical and biological substances are stored in hair where they can be detected and dosed (Table 23.1).

Table 23.1 Substances that can be detected and dosed in the hair shaft

Amfetamine/metamfetamine
Anticonvulsants
Benzodiazepines
Cannabinoides
Chloroquine
Cocaine/benzoylecigonine
DNA
Doping substances (clenbuterol, corticosteroids, efedrine, methenolone, nandrolone, salbutamol, stanazolol, testosterone)
Indinavir
Opiates/morphine/dextromoramide
Neuroleptics
Nicotine/cotinine
Poisons (arsenic, lead, mercury, thallium)

The hair represents a unique substrate for forensic purposes because the hair shaft is not influenced by changes in the blood chemistry or by exposure to chemicals which occurred after hair formation. Advantages of hair samples also include their easy and non-invasive collection, the small sample size required for analysis and the easy storage at room temperature. The reliability of hair analysis is reduced in cosmetically treated hair where the content of drugs and toxics is decreased.

Hair analysis is useful to identify doping practices and drug abuse

The dosage of nandrolone and its metabolites in the hair permit to discriminate between intake of doping agents and intake of other 19-norsteroids, such as norandrostenedione and norandrostenediol, which are available in over-the-counter vitamin supplementations.

In case of drug abuse, the detection of the drug together with its metabolites (cocaine/benzoylecigonine; amfetamine/metamfetamine) confirms intake of the incriminate substance followed by its metabolism and rule out contamination of hair from the environment.

Hair analysis can also be utilized for monitoring treatment compliance in psychiatric, epileptic and HIV patients.

Poisoning can also be diagnosed from hair analysis

Poison from heavy metals is characteristically associated with hair loss (Table 23.2). Hair analysis can confirm diagnosis even in patients without scalp involvement.

Table 23.2 Heavy metals causing alopecia

Arsenic	Gold
Bismuth	Lithium
Cadmium	Mercury
Copper	Thallium

Arsenic and thallium

Arsenic and thallium produce hair loss and poisoning may be diagnosed by dosing the metals in hair sampling. Thallium poisoning causes a very acute and severe anagen effluvium with involvement of scalp, eyelashes, the lateral portion of eyebrows and body hairs. Hair loss starts 2 weeks after poisoning.

Mercury

Hair loss is a diagnostic sign of mercury poisoning. Other symptoms include stomatitis, neurological and mood disturbances, nail pigmentation.

Dosage of androgens in terminal scalp hair

This may provide a basis for predicting baldness: the ratio of testosterone–epitestosterone being significantly

greater in the hair of balding fathers and their sons than in the hair of non-balding control subjects.

Hair in systemic disorders

Hair loss is a symptom of a large number of systemic disorders, including infective diseases, metabolic and nutritional disorders and any condition that produces fever, weight loss and anemia.

In most cases the hair density is not severely reduced and alopecia is not the symptom that leads to the diagnosis.

HIV infection (Table 23.3)

Hair loss and seborrheic dermatitis are very common in HIV patients. Hair loss may be particularly evident in patients taking indinavir which also produces loss of body hair and changes in the hair texture and shape with dry and curly hair.

Straightening of the hair has also been associated with HIV infection, especially in blacks. Hair straightening is often associated with increased fragility.

HIV-infected women have a significant prevalence of fine hair, especially when their viral loads are high. SEM examination reveals variation in the hair diameter, cuticle abnormalities and trichoschisis. Hypertrichosis of the eyelashes is another possible feature as well as hypertrichosis of the eyebrows and of the helix.

Table 23.3 Hair changes in HIV-positive patients

Alopecia areata*
Dandruff
Fragility (trichorrexis nodosa)
Graying
Hypertrichosis of the eyelashes
Loose anagen hair*
Straightening
Telogen effluvium
Thinning

*Anedoctal.

Connective tissue diseases

Systemic lupus erythematosus (SLE) produces telogen effluvium in up to 50% of patients.

Hair loss may be associated with scalp erythema. Hair dryness and fragility can also occur and hair at the frontal margin may be broken and unruly. Discoid lesions of the scalp with scarring alopecia are rarely seen in SLE. Lupus panniculitis may produce non-scarring patchy hair loss resembling alopecia areata.

Syphilis

The scalp may be involved in all stages of syphilis. In primary and tertiary syphilis, however, scalp involvement is rare and due to the presence of specific lesions.

In secondary and latent syphilis hair loss occurs in up to 7% of patients and is most commonly an isolated finding (essential syphilitic alopecia).

Essential syphilitic alopecia is reversible and may have three different clinical presentations:

1. Patches of hair loss with typical moth-eaten appearance
2. Diffuse hair thinning
3. Combination of patchy and diffuse hair loss.

Patchy alopecia of the beard and loss of body hairs may also occur. Eyebrow alopecia (omnibus sign) is a typical feature (Figure 23.1).

Syphilitic alopecia may be worsened by initiation of therapy (Herxheimer reaction).

Syringomyelia

Asymmetrical growth of scalp hair has been reported.

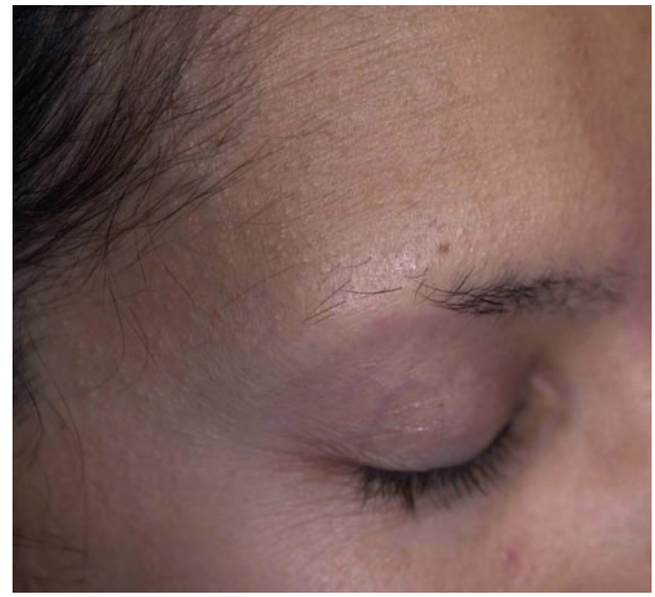


Figure 23.1
Eyebrow alopecia in syphilis.

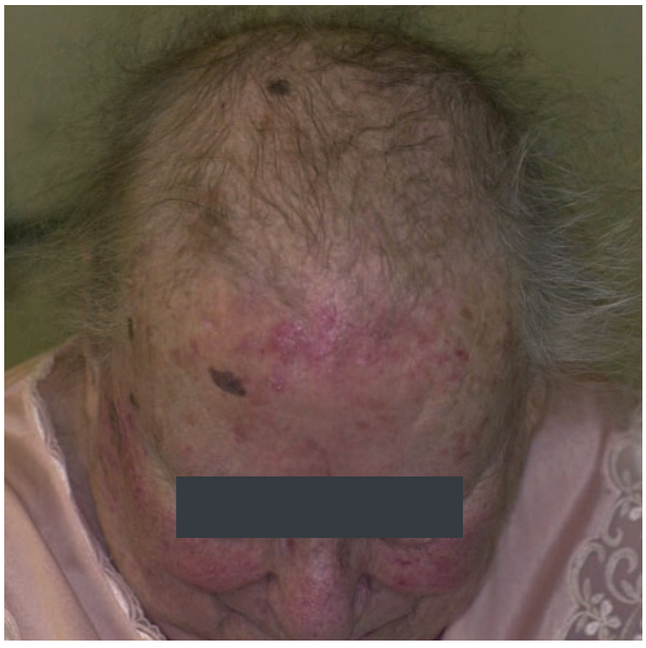


Figure 23.2
Diffuse hair loss in dermatomyositis.

Dermatomyositis

Scalp erythema and scaling may occur in patients with dermatomyositis.

Reversible alopecia occurs in 15–20% of cases (Figure 23.2). Hypertrichosis of the face and limbs is a feature of juvenile dermatomyositis.

Letterer–Siwe disease

Scalp involvement is common with scaly and crusted papules that resemble seborrheic dermatitis. Coalescence of lesions may result in alopecia (Figure 23.3).

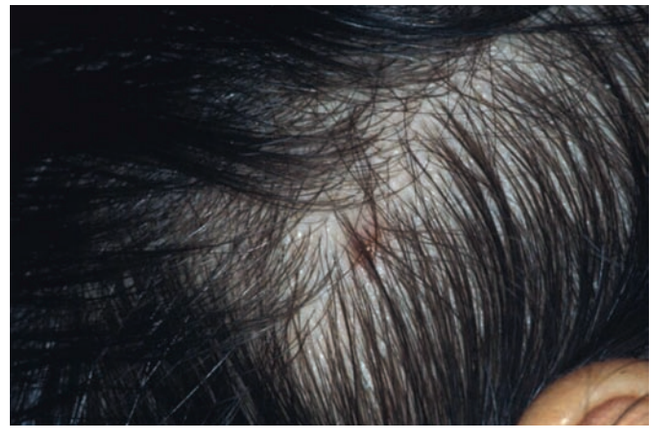


Figure 23.3
Crusted papule of the scalp in histiocytosis X.

Nutritional deficiencies

See Table 23.4.

Anorexia nervosa

Hypertrichosis of the face, back and arms is a typical sign of anorexia nervosa, where it occurs in up to 50% of patients (Figure 23.4). Telogen effluvium and hair weathering are also frequent.

Mycosis fungoides

Hair loss in mycosis fungoides is most commonly, but not exclusively, caused by follicular mucinosis. The alopecia may closely resemble alopecia areata and involve the scalp or other body areas (Figure 23.5a,b).

Table 23.4 Nutritional deficiencies and hair growth

Ascorbic acid (vitamin C)	Corkscrew body hair, follicular hyperkeratosis, hair loss, perifollicular hemorrhages, perifollicular pigmentation
Pantotenic acid (vitamin B5) Riboflavin (vitamin B2)	Hair loss, hypopigmentation, hair curling
Biotin vitamin H/B8)	Hair loss
Zinc	Hair loss
Iron	Hair loss
Protein deficiency (anorexia nervosa, malnutrition)	Hair loss, hair depigmentation diffuse or banded (flag sign, see Table 23.5), hair fragility, slow hair growth rate, body hair hypertrichosis

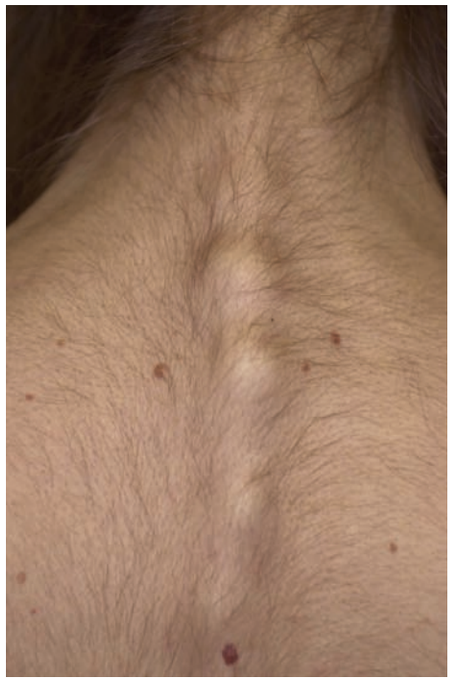
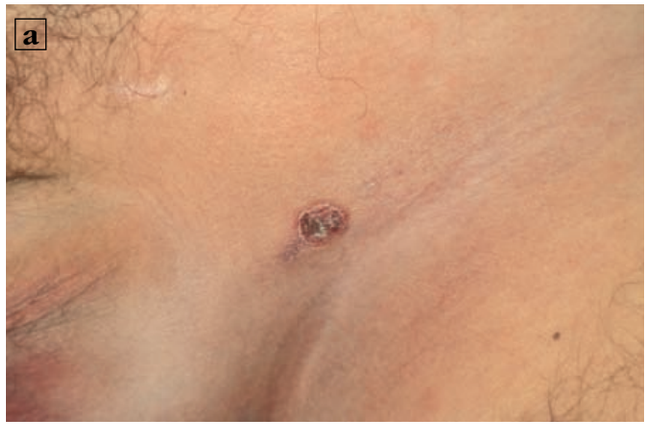


Figure 23.4
Posterior neck hypertrichosis in anorexia.



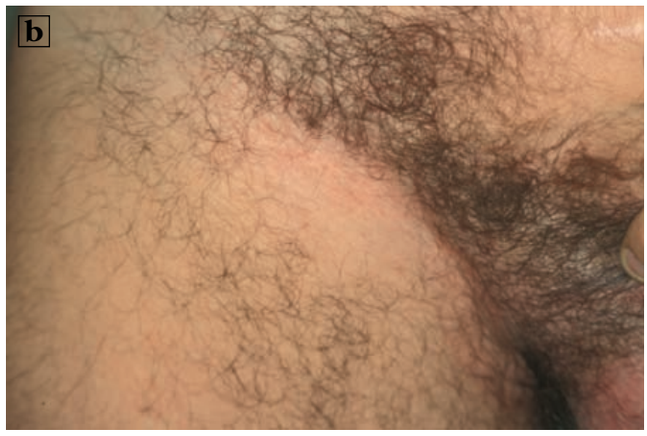


Figure 23.5a,b
Pubic alopecia in mycosis fungoides. Note in (a) a lesion typical for the disease.

Table 23.5 Causes of 'flag sign'

Alcoholism
Anorexia nervosa
Bowel resection
Chemotherapy
Kwashiorkor
Parenteral nutrition
Ulcerative colitis

Follicular mycosis fungoides is a rare variant of mycosis fungoides characterized by a poor prognosis. The disease produces patchy alopecia, resembling alopecia areata, comedo-like lesions and cysts (Figures 23.6, 23.7).

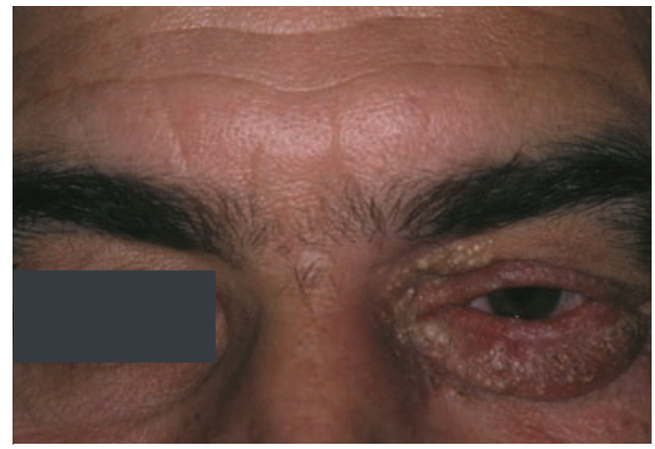


Figure 23.6
Alopecia and comedo-like lesions of the eyelashes in follicular mycosis fungoides.

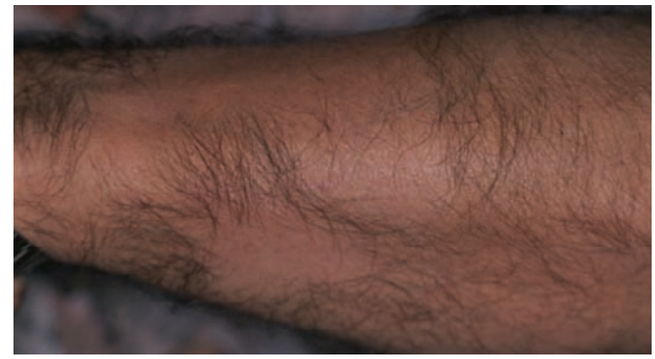


Figure 23.7
Alopecia areata-like patches of hair loss in follicular mycosis fungoides.

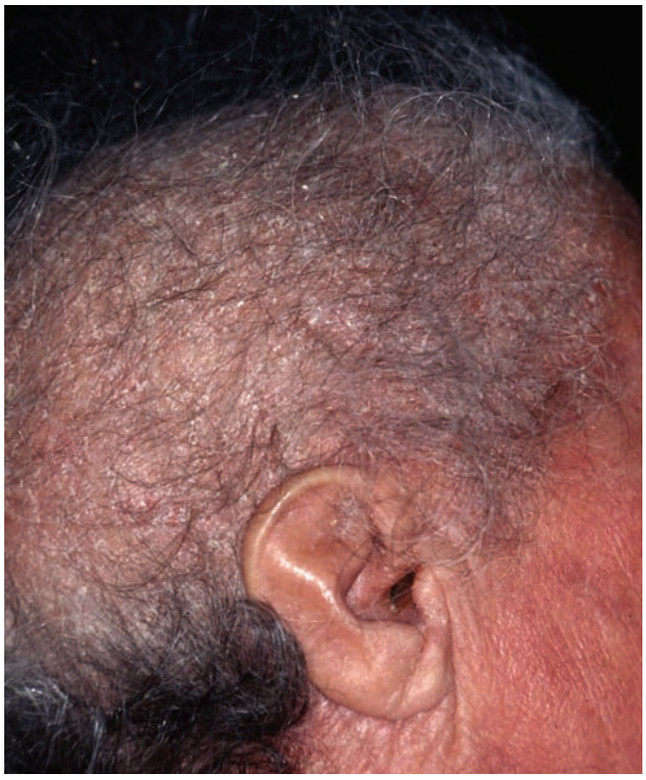


Figure 23.8
Diffuse alopecia in Sézary syndrome.

Diffuse alopecia is a common feature in Sézary syndrome (Figure 23.8).

Follicular mucinosis

Two types of follicular mucinosis are presently recognized: follicular mucinosis associated with mycosis fungoides or Sézary syndrome and idiopathic follicular mucinosis, which probably also represents a non-aggressive localized variant of mycosis fungoides with excellent prognosis.

In follicular mucinosis associated with mycosis fungoides the scalp, the beard or the eyebrows show erythematous indurated bald patches (Figure 23.9). The follicular orifices are often prominent.

In idiopathic follicular mucinosis single or multiple patches of hair loss resembling alopecia areata affect the scalp, trunk and limbs (Figure 23.10). The disease usually affects young individuals.

The pathology shows a lymphoid infiltrate around and within hair follicles and deposits of mucin within the hair follicle.

Differentiation between idiopathic mucinosis and mycosis fungoides associated follicular mucinosis is often impossible even with the PCR analysis of the infiltrate since a monoclonal T lymphocyte population is found in both types.

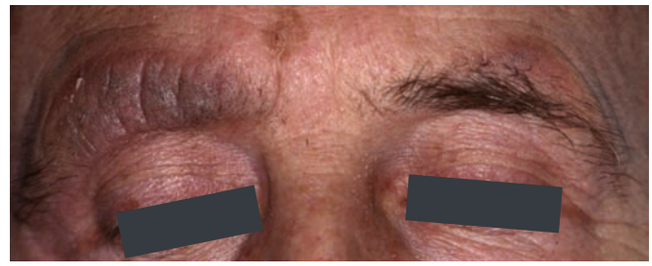


Figure 23.9
Eyebrows alopecia due to follicular mucinosis in a patient with mycosis fungoides.

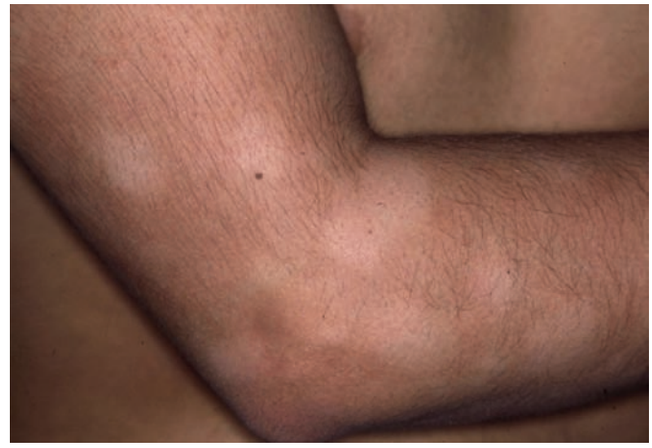


Figure 23.10
Multiple patches of hair loss in idiopathic follicular mucinosis.

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24 Body hair disorders

Body hair may be affected in alopecia areata and lichen planopilaris. This chapter reviews some conditions limited to body hair (Table 24.1).

Abdomen alopecia

Patchy alopecia of the abdomen can be a consequence of friction in patients who sleep on their abdomen and should be differentiated from alopecia areata.

Anterolateral leg alopecia

Patchy alopecia of the anterior and lateral legs is very common in men and possibly related to friction (see page 105). The skin is normal or dry without signs of inflammation (Figures 24.1–24.3).

Coiled hair

Coiling of body hair includes: circle hairs, rolled hairs and corkscrew hairs.

- Circle hairs are characterized by asymptomatic, perfectly circular (ring) hairs underneath the stratum corneum without follicular abnormalities or inflammation (Figure 24.4). The condition affects the back,

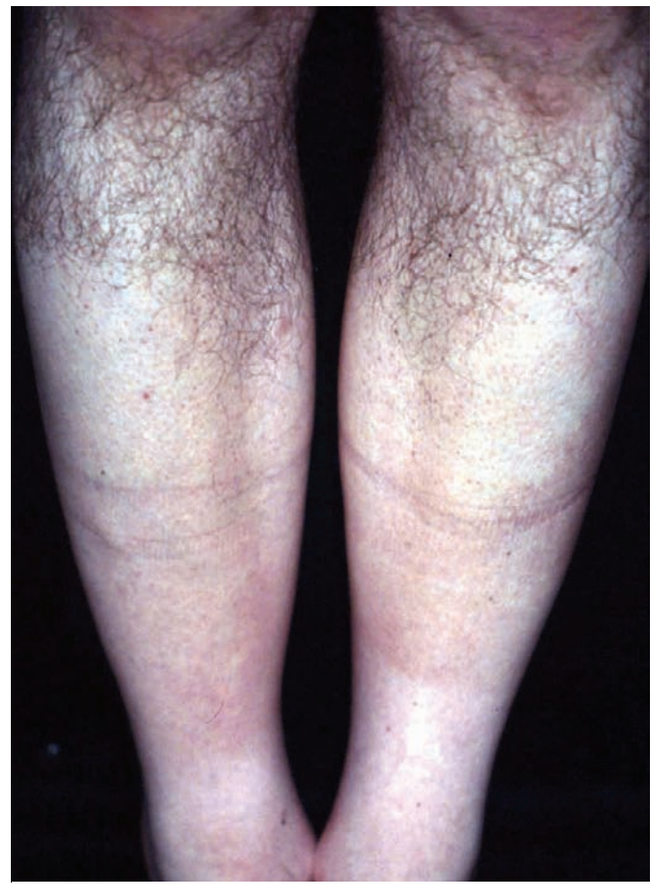
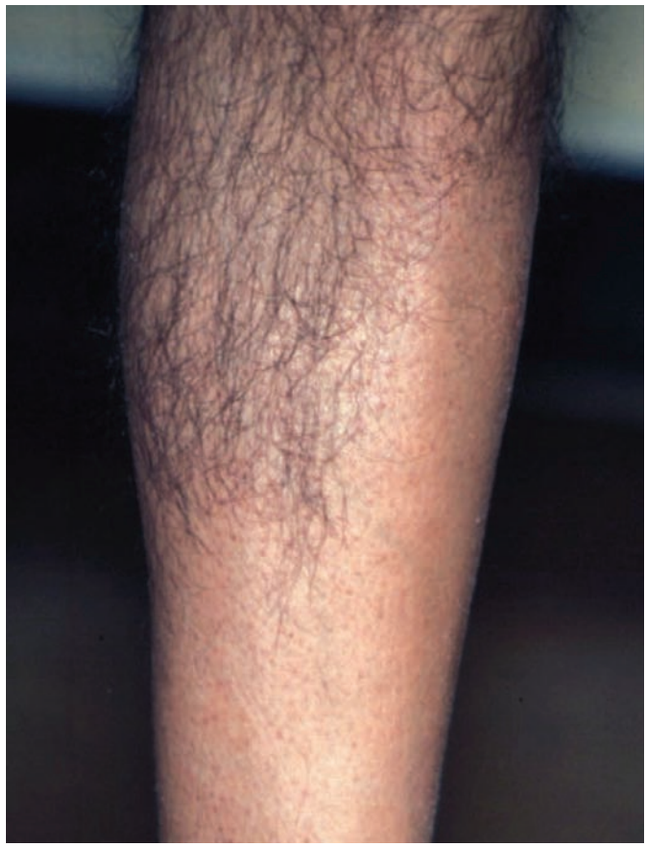


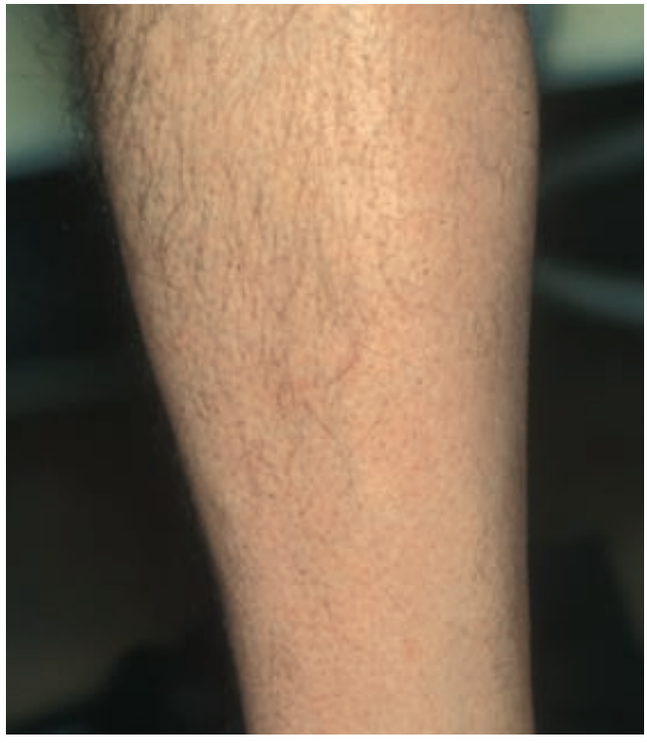
Figure 24.1
Friction alopecia of the anterior leg.

Table 24.1 Body hair disorders

	Causes	*Clinical characteristic*	*Site*
Circle hair	–	Thin, coiled hair	Back, abdomen of elderly obese
Rolled hair	Systemic steroids, dermatological conditions treated with topicals	Irregularly coiled hair within a hyperkeratotic papule, follicular keratosis	Legs
Matting of body hair	Repetitive friction	Knotting of body hair	Back
Trichostasis spinulosa	Retention of vellus hair	Bundles of vellus hairs retained within a follicle	Trunk, upper arms
Eruptive vellus hair cysts	–	Umbelicated papules	Sternal area, abdomen of young



24.2



24.3

Figures 24.2, 24.3
Friction alopecia of the anterior leg.

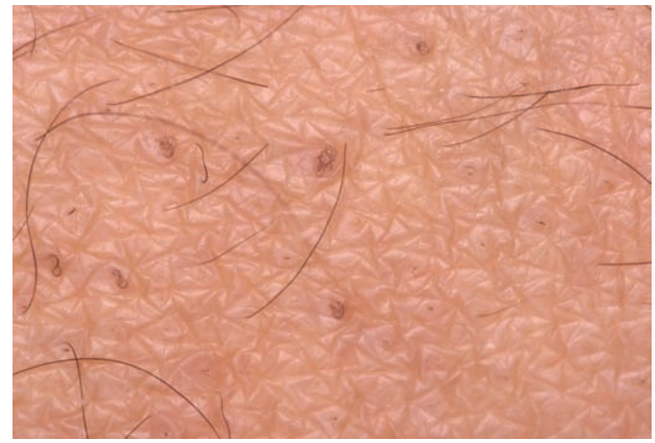
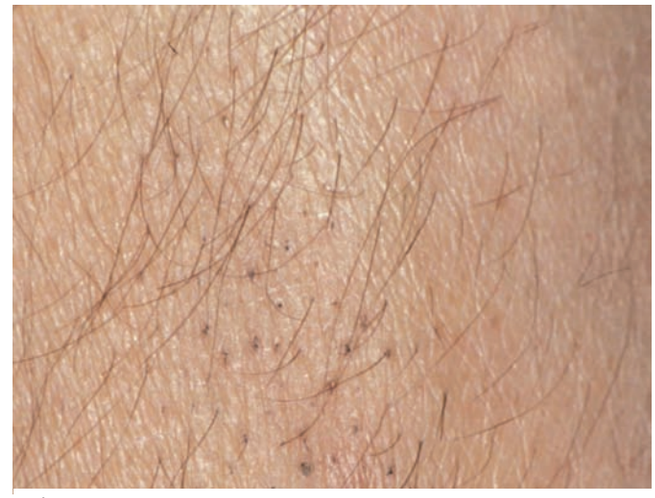
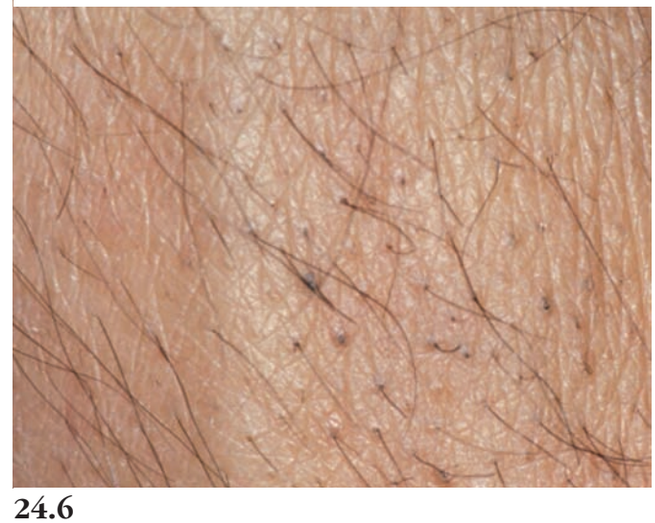


Figure 24.4
Circle hairs.



24.5



24.6

Figures 24.5, 24.6
Rolled hairs.

chest, abdomen, shoulders and thighs of overweight, very hairy males, usually after the age of 55.
- Rolled hairs differ from circle hair because of the presence of keratotic inflammatory follicular lesions and are usually seen in patients with dermatological conditions which require the application of topical drugs or in patients taking oral steroids with or without cyclosporin A (Figures 24.5, 24.6).
- Corkscrew hairs are typical of scurvy where they are associated with follicular hyperkeratotic papules, perifollicular hemorrhages and hair fracturing.

Matting of body hair

This disorder is characterized by knotting and rolling together of several hair shaft. The back is usually affected. The condition may be familial or acquired, usually in correlation with mechanical traumas, such as rubbing, massaging or regular application of topical drugs.

Trichostasis spinulosa (see page 101)

This results from retention of 5–50 telogen vellus hairs within the follicle. It may affect the face of the elderly or produce an itchy papular eruption of the trunk and upper arms of young adults.

Pseudofolliculitis of the legs

These are a common consequence of waxing, plucking or electrical plucking of leg and groin areas. The condition is rare in women who shave their legs.

In most cases a few follicular papules are present, but a more diffuse eruption with pustules may occasionally occur. The pathogenesis is similar to that of pseudofolliculitis of the beard.

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