# 9 Management of noncicatricial circumscribed alopecia

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## COGNITION LIES IN THE EYE OF THE BEHOLDER

The skin and hair are gratifying for diagnosis. One has but to look, and recognize, since everything to be named is in full view. Looking would seem to be the simplest of diagnostic skills, and yet its simplicity lures one into neglect. To reach the level of artistry, looking must be a skillful, active undertaking. The skill comes in making sense out of what is seen, and it comes in the quest for the underlying cause, once the disorder has been named. In many instances, a specific diagnosis is made in a fraction of a second if it is a simple matter of pattern recognition. The informed look is the one most practiced by dermatologists; it comes from knowledge, experience, and visual memory. Where the diagnosis does not come from a glance, the diagnostic tests come in (i.e., the dermatological techniques of examination, and the laboratory evaluation).

By definition, alopecia is the acquired condition of recognizable hair loss. The term derives from Greek with reference to the loss of hair in patches in foxes afflicted with sarcoptic mange. Traditionally, the alopecias have been classified into generalized or circumscribed alopecia, congenital or acquired alopecia, and alopecia with or without evidence of scarring.

When examining the scalp, the distribution of hair loss, the presence and characteristics of associated anomalies, and the presence of scarring should be noted.

In the case of congenital absence or lack of hair, the correct terms are *atrichia* and *hypotrichosis*, respectively.

Aquired hair loss in patches signifies either alopecia areata, traumatic alopecia, trichotillomania, or scarring alopecia.

Finally, increased fragility of the hair shaft may lead to pseudoalopecia from breakage of hair, particularly in regions with higher friction, such as the occiput.

*Congenital triangular alopecia* (of Sabouraud), also known as temporal alopecia or temporal triangular alopecia, is a congenital disorder that usually appears in childhood as a focal patch of fine vellus hairs with a usually triangular configuration in the temporal region of the scalp (Figure 9.1). The condition basically represents the most common of a variety of congenital circumscribed hypotrichoses of the scalp, and may appear either uni- or bilaterally. The suggested frequency for this condition in the general population has been estimated around 0.11%. The majority of cases are detected in childhood between the ages of 2 and 9 years.[1]

The condition presents usually as an isolated finding; rare cases have been associated with *Setleis syndrome* that is characterized by scar-like, circular depressions on the temples (Figure 9.2), which may resemble forceps marks and (much as with aplasia cutis congenita) give rise to litigations with respect to alleged obstetrical injury.[2]

A patchy type of hair loss, particularly over the cranial sutures, and associated with cutaneous atrophy of the scalp, is a typical finding of the *Hallermann–Streiff syndrome* (oculomandibular dyscephaly). Patients have a typical bird-like face, due to a sharply curved, thick, pointed nose, and mandibular hypoplasia. Associated anomalies, believed to derive from a developmental defect of the branchial arch, are brachycephaly, dental, and ocular abnormalities.[3]

Hair restoration surgery using follicular unit transplantation is usually successful for treatment of congenital triangular alopecia.[4]

*Alopecia areata* is a common hair loss condition characterized by an acute onset of nonscarring hair loss in usually sharply defined areas. Any hair-bearing area can be affected, but the most noticeable are the scalp, the beard area, and the eyebrows. The characteristic patch of alopecia areata is usually round or oval and is completely bald and smooth (Figure 9.3a). Some patients lose hair in only a small patch, while others may have more extensive or less frequently diffuse involvement. Some patients show a reticular pattern of alopecia resulting from confluence of multiple small patches of alopecia (Figure 9.3b). Ophiasis is a form of alopecia areata characterized by the loss of hair in the shape of a wave at the circumference of the head (Figure 9.3c). The name derives, again, from the Greek word for snake, because of the apparent similarity to a snake shape and the pattern of hair loss. Occasionally, alopecia areata may progress to complete baldness, which is referred to as alopecia totalis, and when the entire body suffers from complete hair loss, it is referred to as alopecia universalis.

Alopecia areata is considered to be of autoimmune origin with an organ-specific, T cell-mediated assault on the hair follicle at the level of the bulb. The hair bulb is an immune protected site with deficient major histocompatibility complex (MHC) expression. There is circumstantial evidence suggesting that alopecia areata results from

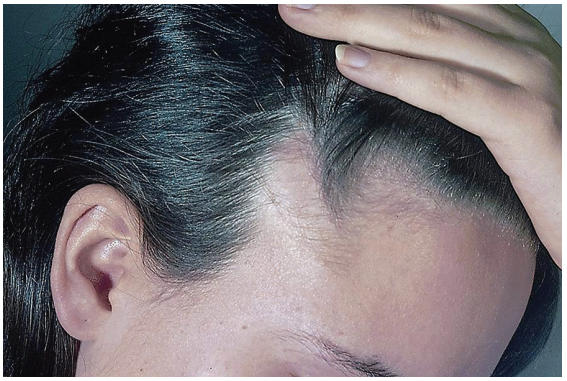

*Figure 9.1* **Congenital triangular alopecia. (Reproduced from Trüeb RM. *Haare Praxis der Trichologie*. Steinkopff, Darmstadt 2003. With permission.)**

loss of immune privilege with presentation of autoantigens. A peribulbar lymphocytic infiltrate induces hair follicle keratinocytes to undergo apoptosis resulting in inhibition of cell division in the hair matrix.

The progress of alopecia areata in an individual patient is unpredictable, though a large surface area, a long disease duration, and associated nail abnormalities (nail pitting, and trachyonychia, rarely onychomadesis) have been associated with a poor prognosis.

With respect to treatment of alopecia areata, a recent meta-analysis of published trials states that only a few treatments have been well evaluated in randomized trials.[5] The authors found no randomized controlled trials on the use of diphenylcyclopropenone (DCP), intralesional corticosteroids, or dithranol, although commonly used in clinical practice. Although topical steroids and minoxidil are widely prescribed and appear to be safe, there is no convincing evidence that they are beneficial in the long term. Most trials have been reported poorly and are so small that any important clinical benefi s are inconclusive. Of 17 trials including 6–85 participants with a total of 540 participants assessing a range of interventions that included topical and oral corticosteroids, topical ciclosporin, photodynamic therapy, and topical minoxidil, none showed significant treatment benefit in terms of hair growth when compared with placebo. The authors came to the conclusion that considering the possibility of spontaneous remission, especially for those in the early stages of the disease, the options of not being treated therapeutically or, depending on individual preference, of wearing a wig may be alternative ways of dealing with this condition.

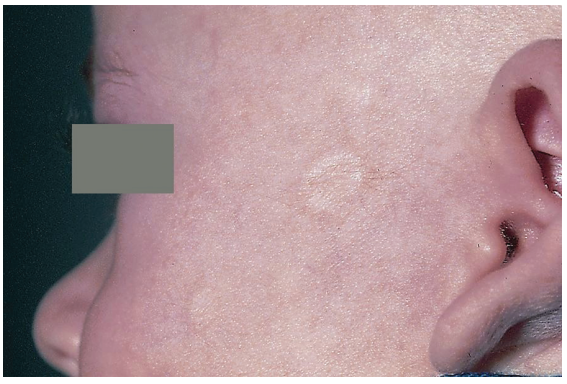

*Figure 9.2* **Setleis syndrome. (Reproduced from Trüeb RM. *Haare Praxis der Trichologie*. Steinkopff, Darmstadt 2003. With permission.)**

The spontaneous remission rates for patchy alopecia areata are one-third within 6 months, half within one year, and two-thirds within 5 years; thereafter, complete remissions are rare. Recurrence rates within 5 years are 80%, and within 20 years are 100%. Total remission rates for alopecia totalis or universalis with a disease duration of 5 years or more are 1% in children and less than 10% in adults.

Nonetheless, depending on patient age, surface area, and disease duration, a treatment algorithm can be designed: Single patches of alopecia areata are best treated with intralesional triamcinolone acetonide 10 mg/mL by jet injector or insulin syringe on a monthly basis, for an average of three to a maximum of six consecutive treatments (in children 5 mg/mL, for eyebrows 2.5–5 mg/mL, and for beard 5 mg/mL by insulin syringe).

With pulse corticosteroid therapy (500 mg IV methylprednisolone on 3 consecutive days, in three cycles 4 weeks apart) within 6 months of disease onset remission rates are 88% for multilocular alopecia areata with a surface area <50%, 59.4% with a surface area >50%, and 21.4% in alopecia totalis. After 6 months of disease onset the remission rate is 15.8%.[6]

With 0.05% topical clobetasol ointment under occlusion (Saran wrap) on 6 consecutive nights per week over 6 months, regrowth of hair is achieved in alopecia totalis or universalis in 17.8%. Despite positive clinical results obtained using clobetasol ointment with occlusive dressing, this approach has a low patient compliance, especially in patients with residual hair.[7]

More recently, 0.05% clobetasol foam applied twice daily on 5 consecutive days per week proved to also be an effective, safe, and well-tolerated treatment with good cosmetic acceptance and patient compliance.[8]

In a retrospective study of 68 patients with severe alopecia areata (>40% scalp hair loss) treated for at least 5 months with topical diphenylcyclopropenone (DCP), Pericin and Trüeb[9] found an overall response rate of 70.6%, with 30.9% complete remission and 39.7% partial remission. Among the investigated prognostic factors for the outcome of DCP therapy, only the extent of alopecia areata at the time of initiation of treatment was found to be of significance. Total remission rates were as follows:

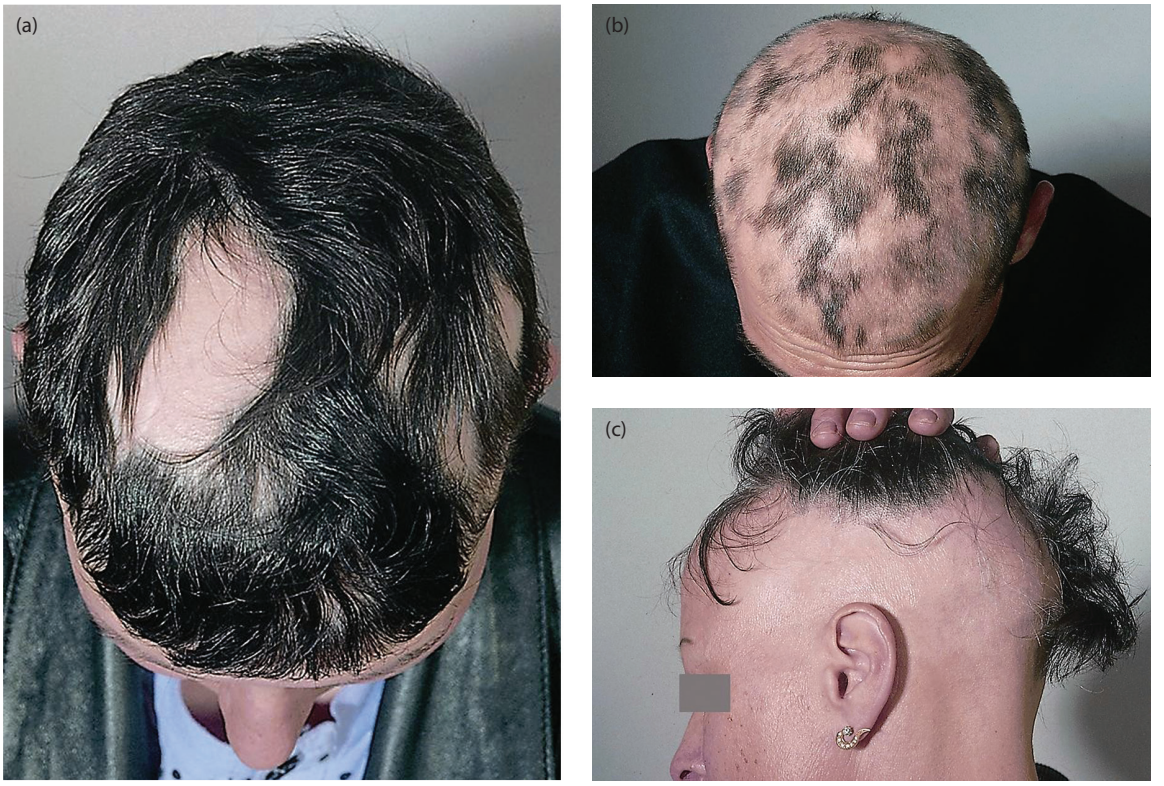


***Figure 9.3*** **Alopecia areata: (a) typical bald patch of alopecia areata, (b) reticular pattern, and (c) ophiasis pattern. (Reproduced from Trüeb RM. *Haare Praxis der Trichologie*. Steinkopff, Darmstadt 2003. With permission.)**

for multilocular alopecia areata were 43.8%, for subtotal alopecia areata and ophiasis 33.3%, and for alopecia totalis or universalis 21.4%, irrespective of disease duration.

Joly[10] proposed the use of methotrexate (MTX) alone or in combination with low doses of oral corticosteroids in the treatment of alopecia areata totalis and universalis, with an overall success rate of 64%. Best results are achieved with subcutaneous MTX in the maximal dosage of 30 mg weekly in combination with 20 mg oral prednisone daily, with regrowth of hair beginning within 2 to 4 months of therapy. Drug toxicities are to be carefully weighed against treatment benefi .

Ultimately, the options available for adapting to the disease rather than treating in an effort to cure are to be taken into consideration in selected long-standing widespread cases or recurrent small spot disease.

New drug treatment opportunities, based on the results of genome-wide association studies that implicate T cell and natural killer cell activation pathways, are paving the way to new approaches in future clinical trials for alopecia areata. Currently, there are ongoing studies with the CTLA4-Ig fusion protein abatacept (blocks costimulation of T cells), anti-IL15Rβ monoclonal antibodies (blocks activation of $CD8^+$ T cells), and the Janus Kinase (JAK) 3 inhibitor tofacitinib and JAK 1/3 inhibitor ruxolitinib (block signal transduction at the IL-15 receptor).[11]

Platelet-rich plasma (PRP) is blood plasma that has been enriched with platelets. As a concentrated source of autologous platelets, PRP contains and releases through degranulation several growth factors and cytokines. These include PDGF, TGFb, FGF, IGF-1 and IGF-2, VEGF, EGF, IL-8, and KGF. PRP has gained popularity among a limited number of dermatologists with a primary commercial interest, though the use and clinical validation of the method for diverse dermatologic conditions is still in the early stages. Results of basic science and preclinical trials have yet to be confirmed in large-scale controlled clinical trials. A Cochrane review on PRP has been performed in chronic wounds.[12] Even if the conclusion of the respective review was positive, this would have not been surprising, because blood platelets have a physiologic role in wound healing. In contrast, it is more difficult to comprehend the role of platelet-derived growth factors and cytokines for hair growth and treatment of hair growth disorders. For this reason, a stringent evaluation of existing data in favor of PRP for treatment of hair-related disorders is indispensable. There exists a single, double-blind, placebo- and active-controlled, half-head study to

evaluate the effects of PRP on alopecia areata published in 2013.[13] However, in an era in which a more comprehensive understanding of the immunologic basis of alopecia areata has opened the venue to more targeted treatments of alopecia areata, in the opinion of the more stringent scientific community, the proposal of PRP, with its poorly defined mode of action, rather represents an intellectual step backward.

## TRAUMATIC ALOPECIA

*Traction alopecia* is defined as hair loss resulting from either prolonged or repetitive pulling force to the hair. Clinically, traction alopecia most often affects the frontal and temporal scalp areas. However, it has been extensively reported in the literature to occur on many different regions of the scalp, depending on an individual's hair grooming practices, which may or may not be related to the ethnic or cultural background. The condition was originally described in female subjects in Greenland who developed hair loss along the hairline from wearing tight ponytails. In 1958, Slepyan[14] reported alopecia in American girls wearing ponytails, and stated that the patches of baldness need not be limited to the margins of the scalp, since alopecia may occur along any line of traction. Traction alopecia is also seen occasionally in long-haired people who use barrettes to keep the hair out of their faces. The more recent literature has focused on traction alopecia from African hair-braiding styles. It has been pointed out that in African females the likelihood of developing traction alopecia increases when traction is applied to chemically processed hair. The risk of developing traction alopecia also increases in the presence of androgenetic alopecia, and with age. With androgenetic alopecia, the hair seems to be less resistant to traction, while with age, traction alopecia is likely to result from a longer history of the aforementioned hair practices.[15] Finally, the risk of developing traction alopecia is substantial in hair weaves, that are worn to conceal hair loss, usually resulting from androgenetic alopecia. Hair weaving involves creating a braid around the head below the existing hairline, to which a hairpiece is attached. Since the hair of the braid is still growing, it requires frequent maintenance, which involves the hairpiece being removed, the natural hair braided again, and the piece tightly reattached. The tight braiding and snug hairpiece cause tension in the hair that is already at risk.

Traction alopecia generally does not present any diagnostic difficulties, provided the possibility is considered. Diagnostic challenges may be encountered if the clinical suspicion is not high or if no history of traction is obtained. The earliest clinical manifestation of continuous traction on hair is perifollicular erythema. Occasionally, keratin cylinders may surround the hairs just above the scalp surface. Not infrequently patients complain of localized dandruff with itching. Eventually the erythema around the follicles will evolve into folliculitis and minute folliculopustules may become evident. In general, patients who develop any symptoms, including pain, pimples, stinging, or crusts with hairdressing are at increased risk of developing traction alopecia. The process gradually leads to loss of hair, which becomes irreversible after sustained traction. By the time the alopecia is evident, the scalp usually no longer shows inflammatory changes. Although the hair loss has become permanent, the skin does not have the quality common to the usual types of cicatricial alopecia, remaining soft and pliable. On close inspection there is a decrease in the density of follicular orifices. In traction alopecia of the marginal hair line, Mirmirani et al. made the observation that the presence of retained hairs along the frontal and/or temporal rim, which they termed the fringe sign (Figure 9.4), may be a useful clinical marker seen in 85% of women with the condition.[16]

Treatment of traction alopecia depends on whether or not long-standing traction has resulted in permanent loss of hair. Accordingly, management of traction alopecia is divided into prevention, treatment of early disease, and treatment of long-standing disease. Prevention is key in girls, and involves educating parents on the importance of loosening the hairstyle. Brushing the affected area with the misbelief of stimulating hair growth should be avoided as well. In adults with early traction alopecia, the hairstyle should also be loosened. Moreover, chemicals or heat are to be avoided. Intralesional triamcinolone and oral tetracycline antibiotics may be beneficial in suppressing perifollicular inflammation, while added topical minoxidil may promote hair growth in some patients. Ultimately, with long-standing disease, surgical procedures, such as hair transplants in the form of micrografting, minigrafting, and follicular unit transplantation, may be considered.

*Postoperative pressure alopecia* represents yet another peculiar form of traumatic alopecia frequently mistaken

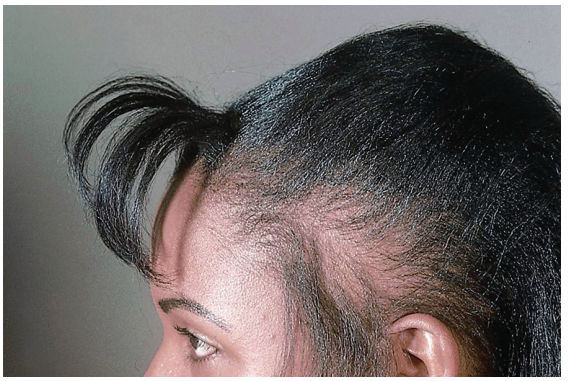

***Figure 9.4*** **Fringe sign in traction alopecia. (With kind permission from Springer Science+Business Media: Female Alopecia. *Guide to Successful Management*, 2013, Berlin, Germany, Springer, Trüeb RM.)**

for alopecia areata, localized to the occipital scalp (Figure 9.5). Patients typically complain of occipital pain and tenderness within 24 hours of surgery. Signs observed within the first week after surgery may be focal swelling, edema, crusting, and ulceration. The hair loss sets in between 2 and 3 weeks after surgery and is complete within 28 days. The condition was originally reported after open cardiac surgery, and is now increasingly observed following lengthy plastic surgical procedures. It occurs after prolonged pressure on the scalp during general anesthesia with intubation, with the head fixed in one position, and is understood to be due to pressure-induced ischemia. Risk factors for hair loss and scarring include length of the anesthesia, prolonged endotracheal intubation, prolonged head immobilization, intraoperative use of the Trendelenburg position, and additional factors potentially aggravating ischemia of the scalp, such as severe hypotension, massive blood loss, and use of vasoconstrictors.[17]

The most important aspect of prevention of this complication of surgery is knowledge of its existence and pathophysiology. A prospective study that incorporated head repositioning every 30 minutes in cardiac surgical patients significantly reduced the incidence of alopecia from a prospectively determined value of 14% to 1%.[18] Fortunately, the condition is self-limiting in most cases with spontaneous regrowth of hair occurring within 3 months. In cases of necrosis, ulceration, and scarring, the alopecia is permanent.

*Temporary radiation-induced epilation following neuroradiologically guided embolization procedures* represents yet another condition frequently mistaken for alopecia areata. Since the anagen hair follicle is highly susceptible to x-ray exposure, loss of hairs results from acute damage to the actively dividing matrix cells of anagen follicles (dystrophic anagen effluvium). Hair loss occurs within the area exposed to x-rays, and most patients notice hair loss 2 to 3 weeks following radiation exposure (Figure 9.6). Hair loss occurs on hair-bearing skin with doses above 300 to 400 cGy and is permanent only with a single dose above 1200 cGy, and if the dose is fractionated, not until the dose exceeds 4500 cGy. Krasovec and Trüeb[19] originally reported temporary radiation-induced epilation following a neuroradiologically guided embolization procedure in the dermatologic literature. Endovascular procedures have become a widely used treatment of cerebral vascular malformation. Transient radiation-induced alopecia following neuroradiologically guided therapeutic embolization of cerebral vascular malformation is probably underreported, since it seems not to be uncommon. It has been misinterpreted to be due to arterial occlusion; moreover, excluding the differential diagnosis of alopecia areata may be difficult, since the bald patch is devoid of inflammatory signs, and hair loss is characterized by dystrophic hair. The patient history, chronology of events, and localization and geometry of the bald patch usually allow correct diagnosis.

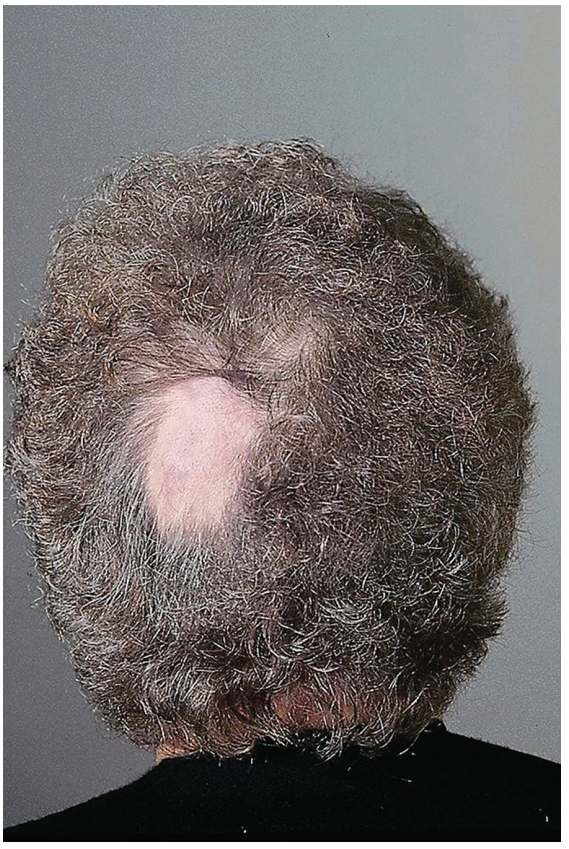

***Figure 9.5*** **Postoperative pressure alopecia. (Reproduced from Trüeb RM. *Haare Praxis der Trichologie*. Steinkopff, Darmstadt 2003. With permission.)**

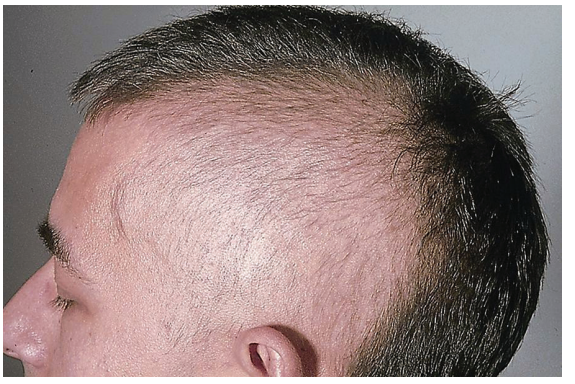

***Figure 9.6*** **Temporary radiation-induced alopecia. (Reproduced from Trüeb RM. *Haare Praxis der Trichologie*. Steinkopff, Darmstadt 2003. With permission.)**

Because the hair loss is temporary, complete hair regrowth occurs spontaneously 2 to 4 months after x-ray exposure, and patient education is paramount to prevent needless anxiety.

## TRICHOTILLOMANIA AND RELATED DISORDERS

*Trichotillomania* involves the repetitive, uncontrollable pulling of one's hair, resulting in noticeable hair loss. It represents a disorder of impulse control. The disorder usually begins between early childhood and adolescence. It occurs six to seven times more frequently in children than in adults, before the age of 6 males predominate, thereafter females. Most commonly, scalp hair is pulled, resulting in ill-defined areas of incomplete hair loss. In the affected areas, there are different lengths of hair, short, longer, and normal. When the hair is pulled in the centroparietal area of the scalp, sparing the lateral margins and the nape of the neck, a tonsural pattern may result, that has been termed tonsure trichotillomania (Figure 9.7).[20]

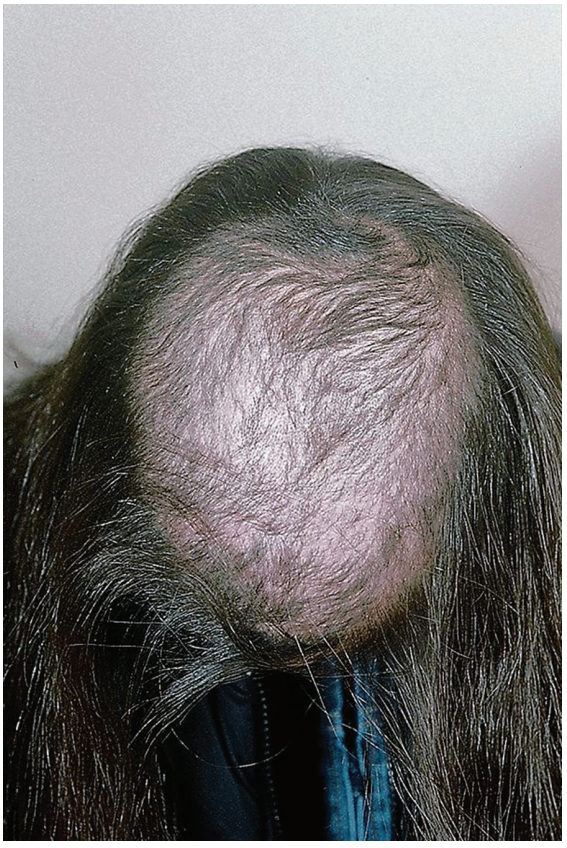

***Figure 9.7*** **Tonsure trichotillomania. (Reproduced from Trüeb RM. *Haare Praxis der Trichologie*. Steinkopff, Darmstadt 2003. With permission.)**

In younger children, trichotillomania results from a mild form of frustration in a climate of psychosocial stress, and soon becomes a habitual practice. From puberty onward, trichotillomania is related to more severe pathologic psychodynamics, and prognosis is more guarded, particularly tonsure trichotillomania in the female adolescent.

The younger the patient, the smaller the percentage of cases referred to a psychiatrist; the rest are treated by the dermatologist who applies his or her own psychiatric knowledge (liaison psychiatry). A proper follow-up is required to establish whether improvement has actually occurred. When the symptom is present in adolescents or adults, competent help from a psychiatrist should be sought. The primary treatment approach for trichotillomania is habit reversal combined with stress management and behavioral contracting. Parents can help by recognizing the problem in its early stages and getting involved in its treatment. Treatment may involve self-monitoring of hair-pulling episodes as well as the feelings and situations that are most likely to lead to hair pulling. Youngsters are then systematically introduced to new behaviors; for example, squeezing a ball or tightening their fist whenever they feel the urge to pull at their hair. Relaxation training and other stress-reduction techniques may also be used, including reward charts that help track and monitor a child's progress with the added incentive of earning small rewards for continued progress. In addition, cognitive therapy is found to be effective.

Children with trichophagy should be screened for iron deficiency as part of their evaluation, since the association of pica—an unusual craving for nonfood items—and iron deficiency has been reported. The compulsive oral behavior characteristically resolved with the oral administration of therapeutic doses of iron. It must be kept in mind though, that iron deficiency may either be a cause of trichophagy or result from gastrointestinal bleeding in the case of trichobezoar.

In a dermatologic setting, a pharmacologic approach may be most feasible for patients who refuse to be referred elsewhere. Basically, the same pharmacologic agents are used for the treatment of trichotillomania as for obsessive–compulsive disorder: the older tricyclic antidepressants imipramine and clomipramine, and the newer selective serotonin reuptake inhibitors (SSRIs) fluoxetine, fluvoxamine, sertraline, and paroxetine. Physicians using SSRIs for treatment of patients with obsessive–compulsive disorders or trichotillomania are cautioned that the duration of treatment is critical in determining adequate treatment. Improvement continues to occur when the drugs are taken beyond 8- or 12-week trials. A patient showing a partial response after 4–6 weeks would be expected to continue to improve during the following weeks. Cessation of pharmacotherapy results in a relapse in the majority of patients. Despite success with SSRIs, patients with obsessive–compulsive disorders tend to

respond to medication with only partial symptom reduction, suggesting that obsessive–compulsive disorders may be a neurobiological heterogeneous disorder that may require alternative treatment options in the individual patient. For example, successful treatment of five adult trichotillomania patients with a combination of the SSRI escitalopram with the anticonvulsant topiramate was originally reported. Subsequently, Lochner et al.[21] performed an open-label pilot study to investigate the efficacy and safety of topiramate in 14 adults with trichotillomania. They found that topiramate may be useful in the treatment of trichotillomania, and suggested that future studies should investigate the efficacy of topiramate in an appropriately powered randomized placebo-controlled trial.

An interesting new therapy is based on the glutamate modulator *N*-acetylcysteine.[22] It is hypothesized that *N*-acetylcysteine, an amino acid, restores the extracellular glutamate concentration in the nucleus accumbens and, therefore, offers promise in the reduction of compulsive behavior. In a 12-week, double-blind, placebo-controlled study performed in 50 individuals with trichotillomania (45 women and 5 men with a mean age [SD] of 34.3 [±12.1]), Grant et al. orignially found that *N*-acetylcysteine (dosing range, 1200–2400 mg/d) demonstrated statistically significant reductions in trichotillomania symptoms. No adverse events occurred in the *N*-acetylcysteine group, and *N*-acetylcysteine was well tolerated.

To examine the efficacy of *N*-acetylcysteine for the treatment of trichotillomania in children, Bloch et al.[23] again performed a double-blind, placebo-controlled (add-on) study with a total of 39 children and adolescents aged 8 to 17 years with trichotillomania randomly assigned to receive the active agent or matching placebo for 12 weeks. No significant difference between *N*-acetylcysteine and placebo was found on outcome measures. It is noteworthy that on several measures of hair-pulling, subjects significantly improved with time regardless of treatment assignment: in the *N*-acetylcysteine group, 25% of subjects were judged as treatment responders, compared to 21% in the placebo group.

*Trichotillomania in connection with alopecia areata* may pose a special diagnostic challenge. It may result from scratching at the site of alopecia areata that is symptomatic with pruritus, initiating a habit-forming behavior. Alternatively, patients with a mental predisposition may artificially prolong the disfigurement as the hair on the bald patches of alopecia areata regrows, with the aim to maintain gratification of dependency needs, which were being met during alopecia areata.[24]

*Traumatic alopecia due to child abuse (battered child)* is, though uncommon, yet another important differential diagnosis to take into consideration in a child with unexplained hair loss and other signs of physical trauma.[25]

*Trichoteiromania* is the term originally coined by Freyschmidt-Paul et al.[26] for breakage of hair by forcefully rubbing an area of the scalp. The typical clinical presentation is that of a bald patch with broken hairs (Figure 9.8). In contrast to trichotillomania, traumatic changes to the hair shaft are more conspicuous, with splitting at the ends of the hairs, giving the impression of white tips. The underlying mental disorder varies among the patients, though an underlying cutaneous sensory disorder, not explained through any specific dermatological disorder, is a common denominator in all cases. Although trichotillomania is considered to be an obsessive–compulsive disorder, the underlying mental disorder in trichoteiromania represents a more heterogeneous group, including anxiety, depression, or somatoform disorder.[27]

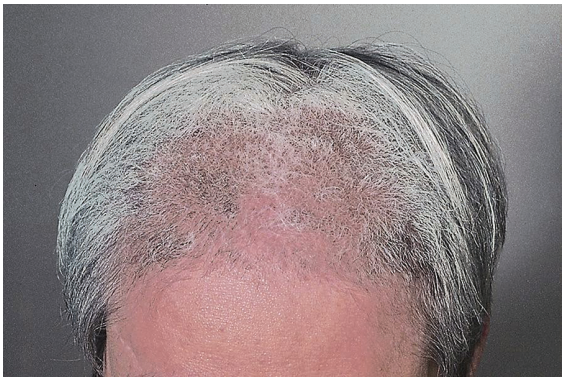

***Figure 9.8*** **Trichoteiromania. (Reproduced from Trüeb RM. *Haare Praxis der Trichologie*. Steinkopff, Darmstadt 2003. With permission.)**

Cooperation with the psychiatrist is indicated, as much as the management and prognosis of trichoteiromania will depend on recognition of the underlying mental disorder and its specific psychotherapeutic and pharmacological treatment.

*Trichotemnomania* is an area of cut-off air as just one of varied manifestations of factitious disorder. Factitious disorder with physical symptoms is a condition in which the patient creates lesions on the skin to satisfy a psychological need of which he or she is not consciously aware, usually a need to be taken care of by assuming the sick role. Patients with factitious disorder create the lesions for psychological reasons, and not for monetary or other discrete objectives as in the case of malingering. Patients knowingly fake symptoms but will deny any part in the process. They desire the sick role and may move from physician to physician in order to receive care. They are usually loners with an early childhood background of trauma and deprivation. The majority of patients suffer from borderline personality disorder. The condition for which dermatologists are consulted often has already occasioned many visits to other physicians. The patient typically presents a bundle of normal investigative findings and a shopping bag filled with oral and topical

medications. Though the possibilities are limitless, consistent is a "hollow" history—a term that refers to the patient's vagueness and inability to give details of how the lesions evolved.

With respect to treatment, the essential and probably most difficult step is to secure an enduring and stable patient–physician relationship. For achieving this goal most clinicians advocate a nonconfrontational strategy, reframing the factitious manifestation as a "cry for help." An interesting approach is that of "contract conference." In this approach the psychiatrist emphasizes the need for the patient to express himself or herself in the common language of difficult relationships, feelings, and problems in living instead of the (factitious) language of illness. After that the patient and clinician can focus their efforts on resolving those real problems. Once a stable relationship is installed, the management of the disorder must be oriented to avoid unnecessary hospitalizations and medical procedures. Another important issue in the management of this condition is recognition and adequate treatment of frequently associated disorders, such as personality disorders, depression, drug and/or alcohol abuse and dependency, etc. Occlusive dressings are used as a diagnostic tool rather than an effective therapeutic intervention, because success is only of temporary nature. Some case reports focus on the use of pharmacological agents. A good response has been reported to the antipsychotic drug pimozide; other clinicians, because of the resemblance to the obsessive–compulsive disorder, advocate the use of clomipramine, or the SSRIs fluoxetine and fluvoxamine maleate. In the vast majority of patients, the condition remains chronic.

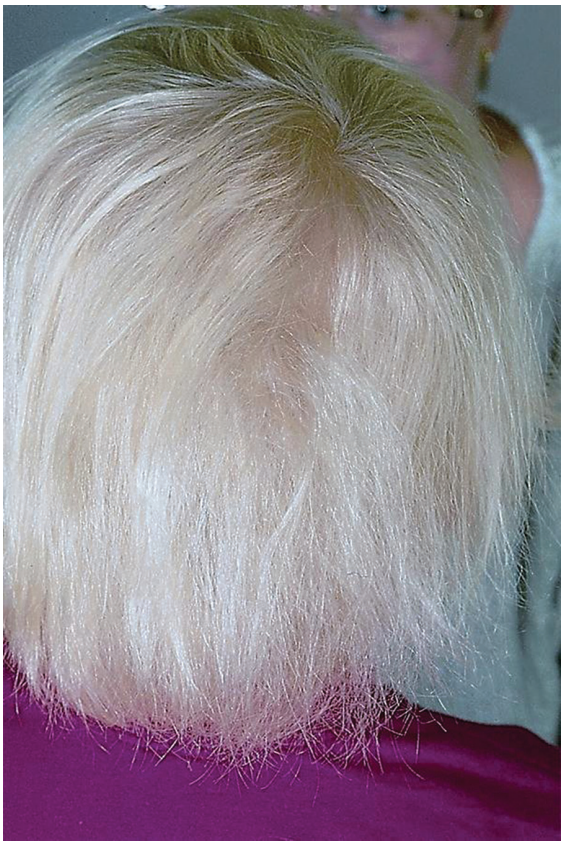

***Figure 9.9*** **Loose anagen hair. (Reproduced from Trüeb RM. *Haare Praxis der Trichologie*. Steinkopff, Darmstadt 2003. With permission.)**

## LOOSE ANAGEN HAIR

*Loose anagen hair*, characterized by easily pluckable anagen hairs, is a disorder predominantly observed in children. The diagnosis is based on the following criteria: on pull-test, painless extraction of >10 anagen hairs (devoid of hair root sheaths), and in the trichogram >80% of plucked hairs are anagen hairs devoid of sheaths.[28] Clinically, the hair may show uneven ends. Additionally, there may be variations in hair texture, and the hair is often dry and lustreless (Figure 9.9). Loose anagen hair has originally been reported in girls but may be under diagnosed in males simply because of hairstyle differences between boys and girls. The condition often recedes with age but can be seen in adulthood, either as a continuation of the disorder that has lingered since childhood, or as late-onset loose anagen hair.

There is no specific treatment for loose anagen hair, except for careful grooming of the hair to avoid extracting the loose anagen hairs. Oral biotin may be beneficial for the strength and texture of the hair.

## ALOPECIA PARVIMACULATA

*Alopecia parvimaculata* (of Dreuw) represents yet another peculiar form of circumscribed alopecia in the pediatric population, in which hair is lost in very small, irregularly shaped patches (Figure 9.10). The etiology has remained obscure, but in its original description may have been related to small epidemic outbreaks of parasitic infection.[29] In 90% regrowth of hair occurs spontaneously, in 10% alopecia is permanent and is understood to represent a pediatric variant of the pseudopeladic state of Degos (i.e., the nonspecific end stage of a variety of scarring alopecias).

## PSEUDOALOPECIA FROM HAIR BREAKAGE

Finally, circumscribed patches of hair loss can result from hair breakage (pseudoalopecia) that is either due to an inherent weakness of the hair shaft, or results from excessive trauma to the hair shaft. The patches of hair loss are usually in areas of higher friction. Already 15 minutes of hair scratching will cause hair to break.

A simple hair-feathering test is used to determine abnormal hair fragility with hair shaft breakage. The distal 2–3 cm of the hairs are grasped between the thumb and index finger. After rubbing the distal hair ends between the thumb and index finger, a brisk pull is made

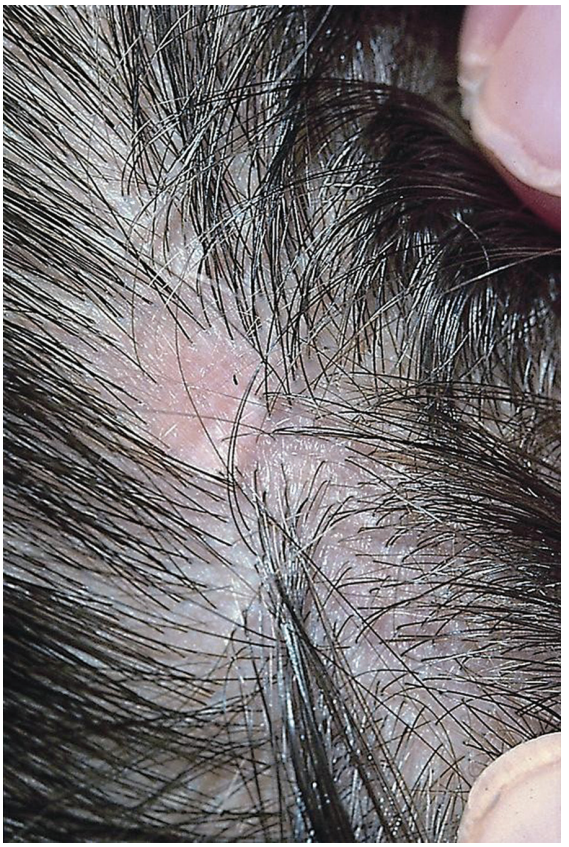

*Figure 9.10* **Alopecia parvimaculata. (Reproduced from Trüeb RM. *Haare Praxis der Trichologie*. Steinkopff, Darmstadt 2003. With permission.)**

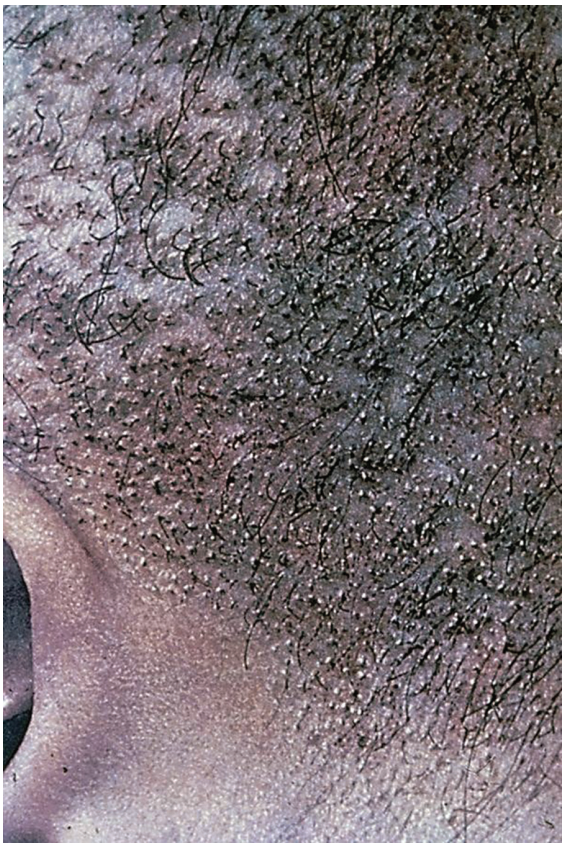

*Figure 9.11* **Monilethrix. (Reproduced from Trüeb RM. *Haare Praxis der Trichologie*. Steinkopff, Darmstadt 2003. With permission.)**

on the ends of the hairs. The thumb and index finger are then checked for short broken hair fragments. In case of increased hair fragility in the hair-feathering test, a light microscopic study of the hair shaft usually reveals the hair shaft efect underlying abnormal breakage.

Hair shaft abnormalities result from genetic conditions and/or exogenous factors affecting the integrity of the hair shaft.[30] Definitive diagnosis is based on microscopic examination of the hair shaft, though dermoscopy represents an effective tool for quick diagnosis of hair shaft disorders on the patient. Hair shaft abnormalities with increased fragility and hair breakage that can be visualized by dermoscopy include the following:[31]

*Monilethrix* is characterized by a beaded appearance of the hair due to periodic thinning of the shaft. The uncommon hereditary condition results in hair fragility and patchy dystrophic alopecia. The alopecia is more severe in the occipital region that is more exposed to friction. Typically, the occipital scalp also presents follicular keratosis (Figure 9.11).

*Pili torti* is characterized by hair that is flattened and twisted through 180° on its axis at varying intervals; the hair can additionally be grooved and has then also been termed *pili torti et canaliculi* or twisting dystrophy. The hair shaft anomaly can be seen as an isolated hereditary trait, in a number of genetic syndromes, including Menkes kinky hair syndrome, Björnstad syndrome (with sensorineural deafness), and the ectodermal dysplasias. Twisting dystrophy in a progressive patch of alopecia in the vertex region is a typical finding in Marie Unna's hereditary hypotrichosis (Figure 9.12).[32]

*Trichorrhexis invaginata* or bamboo hair is the marker for Netherton syndrome, a rare autosomal recessive condition with bamboo hair, ichthyosis, and atopic dermatitis. The hair shaft shows multiple nodes along its length. The nodes consist of a proximal cup-shaped portion and a distal ball-shaped portion, resembling the joint of bamboo. Hair breakage corresponds to the nodes. Eyebrows are affected as well as scalp hair. Simple examination of eyebrow hairs may increase the likelihood of early

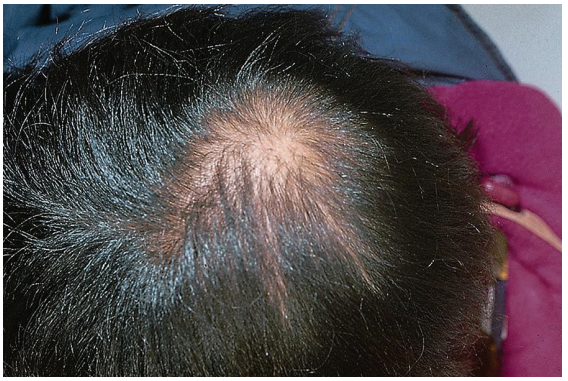

***Figure 9.12*** **Marie Unna's hereditary hypotrichosis. (Reproduced from Trüeb RM. *Haare Praxis der Trichologie*. Steinkopff, Darmstadt 2003. With permission.)**

diagnosis of Netherton syndrome in the newborn with congenital erythrodermic ichthyosis.

*Trichorrhexis nodosa* refers to white knots with transverse fractures along the hair shaft. Dermoscopy reveals brush-like hair fracturing. In general, trichorrhexis nodosa is an unspecific finding related to excess stress of hair in relation to its fragility. It can be observed in hair shaft abnormalities with increased fragility, or more frequently as a consequence of hair weathering.

*Pili annulati* are defined by characteristic alternating light and dark banding in the hair shaft, due to air-filled spaces between the macrofibrillar units of the hair cortex. A priori, not a hair shaft anomaly with increased fragility, the significance of pili annulati lies in that affected hair is more susceptible to weathering, particularly in combination with androgenetic alopecia. With onset of hair thinning due to androgenetic alopecia, progressive reduction of hair shaft diameter may cause increased fragility and trichorrhexis nodosa-like hair shaft racturing.[33]

For treatment, trauma must be minimized, and hair care products and conditioning agents that improve the structural integrity of damaged hair fibers and increase tensile strength are available.

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