Hair Loss

Reese L. Imhof, BA,* Dawn Marie R. Davis, MD,[†‡] Megha M. Tollefson, MD[†‡]

*Mayo Clinic School of Medicine, Rochester, MN
[†]Department of Dermatology and
[‡]Department of Pediatric and Adolescent Medicine, Mayo Clinic, Rochester, MN

PRACTICE GAPS

Hair loss is an often-reported and challenging complaint that may be caused by a wide range of conditions. The psychosocial impact of hair loss can be profound. Pediatricians should understand the causes of hair loss in children and adolescents, including the diverse range of congenital and systemic disorders associated with alopecia, and implement up-to-date management guidelines in clinical practice.

OBJECTIVES *After completing this article, readers should be able to:*

1. Know the common causes of hair loss in children and adolescents: from autoimmune (eg, alopecia areata, alopecia totalis) to psychiatric (eg, trichotillomania) disorders.
2. Gain appreciation of the genetic syndromes associated with hair loss in children.
3. Discuss treatment strategies for the various causes of hair loss.
4. Diagnose alopecia beyond tinea, giving consideration to hair products and cultural practices and their effect on hair loss.

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ABBREVIATIONS

AA	alopecia areata
ACC	aplasia cutis congenita
AE	anagen effluvium
AGA	androgenetic alopecia
AT	alopecia totalis
AU	alopecia universalis
CTA	congenital triangular alopecia
DLE	discoid lupus erythematosus
DSM-5	*Diagnostic and Statistical Manual of Mental Disorders, Fifth Edition*
FDA	Food and Drug Administration
FPHL	female pattern hair loss
KOH	potassium hydroxide
LAS	loose anagen syndrome
OCD	obsessive compulsive disorder
SAS	short anagen syndrome
SLE	systemic lupus erythematosus
TA	traction alopecia
TC	tinea capitis
TE	telogen effluvium
TNHL	transient neonatal hair loss

Hair loss in children and adolescents can result from various conditions, which may be acquired or congenital and range in severity. Hair loss can present in otherwise healthy patients, or it can be a sign of underlying malnutrition, toxicity, or systemic disease. Tinea capitis (TC), telogen effluvium (TE), traction alopecia (TA), trichotillomania, alopecia areata (AA), congenital alopecia, and hair cycle disorders are some of the most prevalent causes of hair loss in children (Table 1). (1)(2) Other causes include thyroid disorders, systemic lupus erythematosus (SLE), diabetes mellitus, structural abnormalities of the hair shaft, iron deficiency anemia, and malnutrition. (1)(2) Management of hair loss in pediatric populations is often difficult, but treatments are available for many conditions. Psychosocial support is an essential component when managing alopecia. Hair loss often causes emotional distress and may result in long-standing psychosocial complications, making a timely diagnosis essential. (1) An understanding of the various causes and management of hair loss is vital for all health-care providers.

BACKGROUND/PATHOPHYSIOLOGY

Understanding hair biology is helpful in appreciating the pathophysiology of various types of hair loss. The growth of a hair follicle is cyclical, consisting of the

Table 1. Summary of the Major Types of Alopecia in Children and Adolescents

ALOPECIA TYPE	ACUTE OR CHRONIC	CONGENITAL OR ACQUIRED	SCARRING OR NONSCARRING	PATHOGENESIS	TYPICAL EXAMINATION FINDINGS	TREATMENT
Tinea capitis	Usually acute, can be recurrent/ chronic	Acquired	Usually nonscarring, can be scarring in late stages	Dermatophyte infection	Patchy hair loss, erythema, scale, black dots (hair broken off at scalp surface), broken hair, cervical lymphadenopathy	Antifungal shampoos and oral antifungals until clear
Alopecia areata	Acute to chronic	Acquired	Nonscarring	Autoimmune mediated	Diffuse, patchy, or total hair loss; minimal or no erythema; no scale; "exclamation point hairs"; may see nail pitting	Off-label treatments include topical corticosteroids, intralesional corticosteroids, topical minoxidil, phototherapy, immunomodulators, topical immunotherapy
Telogen effluvium	Acute shedding 3–4 mo after triggering event	Acquired	Nonscarring	Many triggers, including illness, surgery, trauma, and medications	Diffuse hair thinning and shedding, no erythema or scale, positive hair pull test	Address underlying triggers, usually self-limiting
Anagen effluvium	Acute shedding days to weeks after triggering event	Acquired	Nonscarring	Triggers include chemotherapy or radiotherapy, malnutrition, infection, autoimmune disease, toxin exposure	Profound, diffuse hair loss, no erythema or scale	Address underlying triggers, usually self-limiting but may be permanent
Loose anagen syndrome	Chronic	Congenital	Nonscarring	Hereditary or sporadic mutations	Slow-growing hair or diffuse hair loss, hair texture abnormalities, no erythema or scale, positive hair pull test	Usually self-limiting and improves over several years, topical minoxidil (off-label) in severe cases
Short anagen syndrome	Chronic	Congenital	Nonscarring	Hereditary or sporadic mutations	Hair does not grow long, fine to normal density, no erythema or scale	Usually self-limiting and improves during puberty
Trichotillomania	Usually chronic	Acquired	Usually nonscarring, can be scarring in late stages	Impulse control disorder, grouped with obsessive compulsive, excoriation, body dysmorphic, and hoarding disorders	Patches of hair loss, broken hairs of different lengths, irregular borders, minimal to no erythema, and minimal to no scale	Mental health evaluation, cognitive behavioral therapy
Traction alopecia	Chronic	Acquired	Usually nonscarring, can be scarring in late stages	Repeated or prolonged pulling force on the hair, often from hairstyling or grooming practices	Patchy hair loss or hair thinning, typically involves the temporal scalp, perifollicular pustules and papules, broken hairs, erythema, and scale	Education and implementation of gentle hair care practices to avoid tension
Discoid lupus erythematosus	Acute to chronic	Acquired	Scarring	Autoimmune mediated	Patches of inflammatory lesions and diffuse hair loss, erythema, and scale	Topical, intralesional, or systemic corticosteroid, hydroxychloroquine

Continued

Table 1. (Continued)

ALOPECIA TYPE	ACUTE OR CHRONIC	CONGENITAL OR ACQUIRED	SCARRING OR NONSCARRING	PATHOGENESIS	TYPICAL EXAMINATION FINDINGS	TREATMENT
Aplasia cutis congenita	Chronic	Congenital	Scarring	Congenital deficiency of cutaneous tissue	Single circumscribed lesion with associated scarring hair loss, may be covered by a membrane or surrounded by a hair collar, no erythema or scale	Supportive care for most cases
Transient neonatal hair loss	Acute	Acquired	Nonscarring	Benign, physiological shedding of hair	Occipital alopecia or other focal patches of hair loss, no erythema or scale	Self-limiting and improves over time
Congenital triangular alopecia	Acute to chronic	Congenital	Nonscarring	Congenital deficiency	Triangular, oval, or lancet-shaped patches of alopecia, usually involves frontotemporal scalp, usually left-sided and unilateral, no erythema or scale	Self-limiting, usually remains stable and does not progress
Congenital atrichia	Chronic	Congenital	Nonscarring	Hereditary mutations involving human hairless gene	Total absence of hair at birth, papular lesions, no erythema or scale	Usually unresponsive to therapy
Congenital hypotrichosis	Chronic	Congenital	Nonscarring	Hereditary mutations involving human hairless gene	Decreased hair volume or diffuse hair thinning, no erythema or scale	Usually unresponsive to therapy
Androgenetic alopecia	Chronic	Acquired	Nonscarring	Androgen-mediated miniaturization of hair follicles	Patchy, progressive hair thinning in androgen-dependent regions, no erythema or scale	Topical minoxidil (off-label in patients aged <18 y)

anagen, catagen, and telogen phases. The anagen phase begins by 20 weeks' gestation. (2) In the anagen phase, which can last for up to 6 years, follicles are actively growing. (3) The catagen phase is a short, resting, transition phase lasting a couple of weeks. The telogen phase lasts 2 to 3 months, at which time hair is shed. Next, the anagen phase begins, and the hair cycle is repeated. In a normal hair cycle, approximately 90% of follicles are in the anagen phase, 1% are in the catagen phase, and 9% are in the telogen phase. (1)(4)(5) Of note, the first telogen phase usually occurs in infants between 4 and 8 months, but it can occur as late as 12 months old. This normal shedding is typically seen in infants during their health supervision visits. (2) Hair pigmentation and texture often change with this first cycle, (5) which may be a surprise to parents. The density of hair follicles also changes as an infant grows. (5) At birth there are approximately 5 million hair follicles, which have the ability to generate lanugo, vellus, or terminal hairs. Lanugo is soft, fine, unpigmented hair that is normally seen only in fetuses and neonates. In many cases lanugo will be shed in utero or if present at birth, lanugo hair will be replaced by vellus hair by the time infants reach 3 to 4 months old. (1)(5) Vellus hair is slightly pigmented, fine hair found on the face and extremities. (5) In infants and children, terminal hairs make up the scalp, eyebrows, and eyelashes, and vellus hairs are found in all other hair-bearing areas. During puberty, androgens stimulate vellus hairs to develop into terminal hairs that are thicker and have increased pigment. (1)(4)(5) Mature hair that develops in all areas of secondary sexual hair distribution, such as pubic and axillary hair, is terminal hair. Terminal hair is also found on the scalp, face, and extremities. (5) Although the phases of growth are the same for vellus and terminal hair, vellus hair generally has a shorter anagen phase. Similarly, scalp hair has a longer anagen phase and a greater anagen/telogen ratio than eyelash and extremity hair, which

is why scalp hair can grow longer than hair in other regions. (5)

In assessing pediatric patients with alopecia, understanding the history of a child's hair at birth and the development of their hair loss can aid in diagnosis. It is vital to determine whether the onset of hair loss is acute or more gradual. (2) Providers should obtain a detailed history, focusing on family history, medical history, nutrition, diet, and possible environmental or toxin exposures, and should also conduct a thorough physical examination of the head and scalp. The distribution and regions of hair loss should be determined on scalp examination. Other hair-bearing areas, such as the arms and legs, should also be assessed as part of a complete hair examination and evaluation. Assessing all hair-bearing areas will help determine the extent of hair loss and the types of hair affected (terminal versus vellus). When a diagnosis is uncertain, referral to a dermatologist for additional assessments can be helpful. Additional assessments include the hair pull test, the hair tug test, fungal screens, trichoscopy, trichography, and light microscopy. (1) The hair pull test involves grasping approximately 20 to 60 hairs and pulling away from the scalp, from the proximal to the distal end of the hairs. Hair should be pulled gently and not forcefully. It may not be possible to dislodge hairs, and this can be influenced by other factors, such as when the hair was washed. A positive hair pull test is when greater than 10% of the hairs are easily pulled out. Although not specific, a positive result indicates active hair loss with an increased number of hairs that are in the telogen phase. Hairs that are removed during the hair pull test can be evaluated by dermatopathologists with light microscopy to obtain more information on the features and stage of the hairs. (2)(5) In the hair tug test, a group of hairs is grasped using two hands, with one hand near the root and the other hand near the distal tip. The hairs are then pulled away using the hand at the distal end, looking to determine whether any of the hairs break in the middle. This test is performed to assess hair breakage and is useful in determining hair fragility. (5) Trichography is a semi-invasive procedure performed by dermatologists that involves using forceps to remove hair from different scalp regions followed by microscopic examination. Trichoscopy is a dermoscopic method of evaluating the hair and scalp using a dermatoscope to visualize at magnifications often ranging from 10-fold to 70-fold or higher. This is commonly performed by dermatologists in dermatology clinics. When examining the hair shaft, trichoscopy can often replace the need for hair mounts and microscopy. (5) Trichoscopy is a valuable noninvasive tool that can help visualize the hair and scalp to identify scarring versus nonscarring alopecias, as follicular ostia loss is typical of scarring alopecias, whereas follicular ostia are usually preserved in nonscarring alopecias. (1)(6)(7) Determining whether the hair loss is scarring or nonscarring is often the first step in formulating a differential diagnosis in a patient presenting with hair loss because diseases that affect the hair follicle unit are often one type or the other. Most hair loss in children results from nonscarring, acquired conditions, such as TC, TE, AA, TA, or trichotillomania. (1)(2) However, many of these conditions can lead to scarring and permanent hair loss if untreated because inflammation and scarring can occur over time. (1) Referral to a dermatologist would be considered in cases of diagnostic uncertainty or for patients who have failed treatment.

PEDIATRIC ALOPECIAS

Tinea Capitis

One of the most common causes of hair loss in childhood is TC, a dermatophyte infection that most frequently occurs in young children but that can affect all pediatric patients from infancy through late adolescence. (8) *Trichophyton tonsurans* and *Microsporum canis* are the organisms involved in most cases of TC in the United States. *T tonsurans* is the most common cause and is transmitted via human contact, and *M canis* is the second most common and is often spread by domestic animals. (2)(9)(10) Clinical presentations of TC may be heterogenous, with varying erythema, scaling, and pruritus accompanying hair loss (Fig 1). In acute infections, scale and erythematous patches are seen on clinical examination. In recurrent or chronic infections, diffuse scaling, broken hairs or black dots in lesions (from hairs being broken at the surface of the scalp), and cervical or occipital lymphadenopathy may be present. (2)(5) Although typically a nonscarring alopecia, chronic untreated TC can cause scarring. (1) Severe local inflammation may also present as kerions (Fig 2), which are fungal-associated abscesses most often seen in children aged 5 to 10 years and appear as boggy, purulent nodules. (1)(11)

In patients with suspected TC presenting with patchy lesions or diffuse scale, psoriasis, seborrheic dermatitis, and atopic dermatitis should all be included in the differential diagnosis. Trichotillomania and AA should be ruled out when broken hairs or black dots are present. Noninflammatory TC in particular may be difficult to differentiate from AA, but a notable distinguishing feature is that tinea involves both the scalp and hair. Although tinea almost always has scalp changes, AA rarely involves the scalp. (1)(2) When patients develop pustules and/or inflammatory papules,

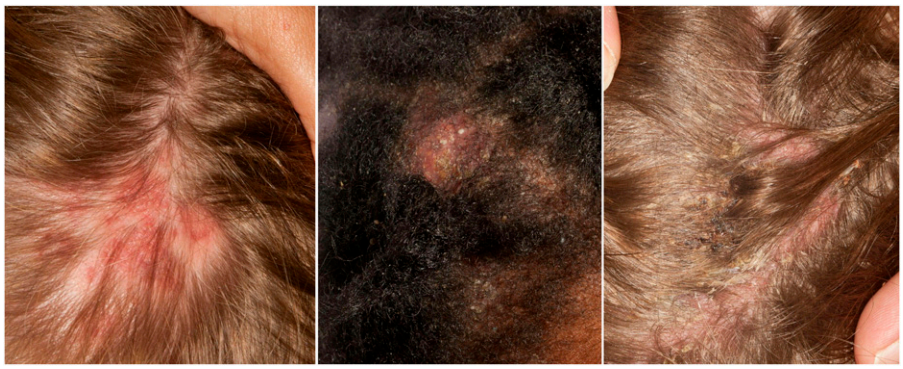

Figure 1. Heterogenous clinical presentations of tinea capitis with varying erythema, scaling, and pruritus accompanying hair loss.

folliculitis and cellulitis are included in the differential diagnosis. The differential diagnosis of TC with kerion formation includes cutaneous abscess and neoplastic processes. (2)

Methods for identifying TC include fungal potassium hydroxide (KOH) preparation and fungal culture from the affected area. (1)(2) However, cultures obtained from patients with kerions can produce false-negative results because kerions are primarily an inflammatory reaction. (12) Treatment of TC infections generally requires systemic medications such as oral griseofulvin and terbinafine until clear (2)(5) because topical azoles, topical allylamines, and ciclopriox olamine are typically ineffective. For TC infections caused by *Trichophyton* species, griseofulvin treatment for 8 weeks and terbinafine use for 4 weeks exhibit similar efficacy, whereas infections caused by *Microsporum* species may respond better to griseofulvin. (2) Of note, the US Food and Drug Administration (FDA) approves terbinafine for TC only in patients older than 4 years and griseofulvin in patients older than 2 years, although this treatment is used off-label for younger children. (2) Patients younger than 1 year often can be treated with topical azoles. (2) The use of ketoconazole shampoo may prevent transmission but is not a treatment. Management of kerions should include keratolytic emollients such as topical urea; glycolic, lactic, and salicylic acid; and soaks and may require short courses of oral corticosteroids, depending on the severity. (5) Treatment should be continued until repeated dermatophyte screening demonstrates cure. (2) Bacterial superinfection may also be present. Concern for secondary bacterial infection should prompt proper evaluation and treatment. Individuals with close contact to children with TC may require preventive treatment with ketoconazole shampoo. (1)(2)(13)

Alopecia Areata

AA is an autoimmune-mediated disease that causes hair loss. Although AA can present at any age, up to 2% of patients present before age 2 years and approximately 20% present before age 16 years. (1)(14)(15) It is suggested that AA in infants is underdiagnosed. (16) The lifetime prevalence of AA at any age is 2%, with an increased risk for patients with a history of other autoimmune disorders. (17)(18) Childhood AA is more prevalent in girls (boys may present with more severe disease, or this may be due to selection bias, with boys being less likely to present unless hair loss is severe). (19) Approximately one-quarter of all children with AA have a

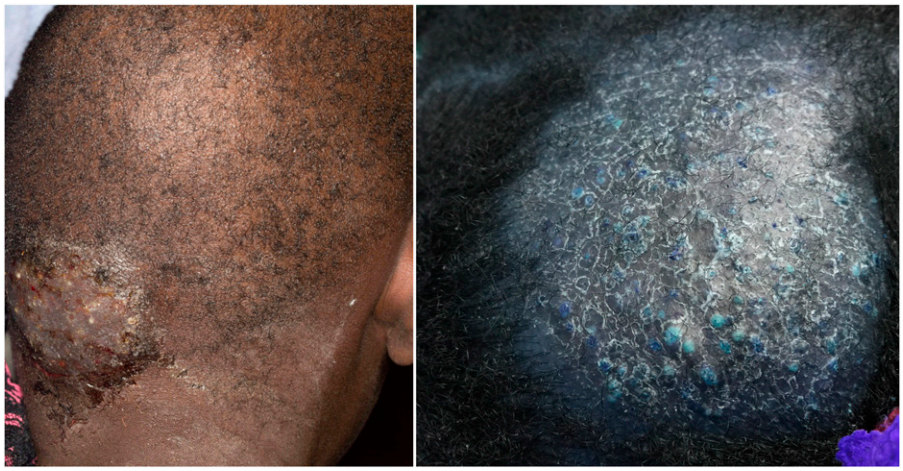

Figure 2. Kerion (fungal-associated abscesses) in patients with severe tinea capitis infections.

positive family history; current evidence suggests that AA may have a genetic component. (19)(20)

AA has a variety of different patterns but most often presents as well-circumscribed round patches of hair loss involving the scalp (Fig 3). These bald patches are typically asymptomatic, but some patients do experience pruritus or pain. (2)(21) In up to 5% of patients, AA can progress to involve total alopecia of the scalp, termed alopecia totalis (AT), or loss of all hair-bearing areas of the body, termed alopecia universalis (AU). (1)(21)

Some of the distinct patterns of hair loss associated with AA include a band of hair loss involving the scalp vertex with temporal and occipital regions spared (sisaipho type), or a band of hair loss involving the occipital scalp (ophiasis type) (Fig 4). Many patients with AA also experience nail changes, such as pitting or "rough nails" (trachyonychia). (1)(21)

Clinical observation of nonscarring, distinct regions of total hair loss, as well as "exclamation point hairs," can be helpful signs in making an early diagnosis of AA. Exclamation point hairs are short, broken hairs that are narrower or thinner close to the scalp surface and may also be described as tapered proximally at the scalp or "telescoping." The hair pull test will be positive in an active patch, and microscopy often confirms tapered hairs. Note that the hair pull test may be negative in a stable patch. Scalp biopsy is typically not required in making the diagnosis of AA, but in patients with atypical presentations, scalp biopsy can be helpful and will show a dense lymphocytic infiltrate involving the dermal papillae and anagen hair bulbs. (1)(2)

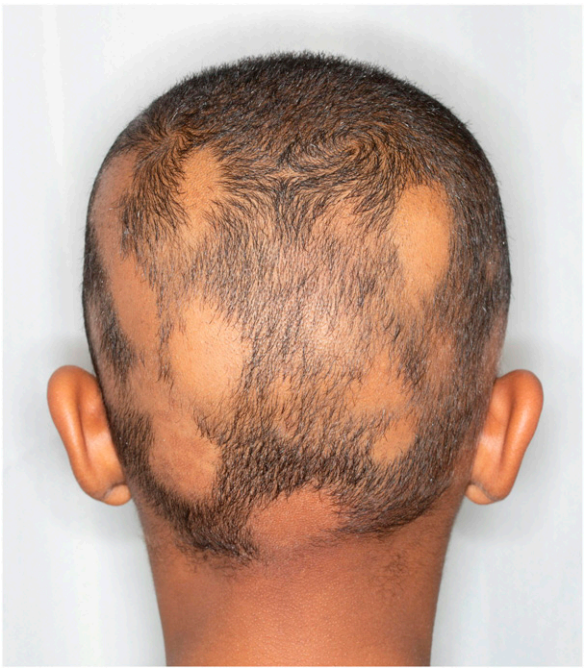

Figure 3. Round patches of hair loss seen in alopecia areata.

Figure 4. Ophiasis type of alopecia areata, characterized by a band of hair loss involving the occipital scalp.

In up to half of all patients with AA, regrowth may occur within 1 year even without treatment. (21) Spontaneous improvement is generally seen in patients with less severe AA who present with limited patches of hair loss. In more severe and chronic cases of AA, patients often present with nail changes, family history of AA, personal history of atopy, increased sites of involvement (AT or AU), ophiasis pattern, and hair loss that lasts for more than a year. Unfortunately, patients who present with AA in childhood are more likely to experience a chronic course. (21)(22)(23)(24)

AA may be associated with other autoimmune disorders, particularly autoimmune thyroid diseases and vitiligo. Although patients with AA may have a higher risk of developing other autoimmune conditions, routine screening for autoimmune disease is often not indicated in otherwise healthy patients with a negative review of systems and a negative family history due to insufficient clinical evidence of increased association in children at this time. (21)

Treatment of AA is often challenging, and there are currently no FDA-approved treatments specifically for AA. A variety of off-label treatment options exist for children with AA; however, patients with the more severe forms of AT or AU often do not respond well to treatment. (2)(21)(22)(23)(24)

Young patients may be responsive to topical corticosteroids, and this is often first-line treatment. (25) Most patients tolerate topical corticosteroids well, and systemic adverse effects are rare. Topical corticosteroids used to treat AA are typically class I to III and can be used alone or combined with topical retinoids or topical anthralin. (2) Intralesional corticosteroid injections may be beneficial in older patients who have fewer patches of hair loss. (1)(2) Dermal atrophy is a concern with continued topical or intralesional corticosteroid use, and patients should be monitored for this adverse effect. Systemic corticosteroids are generally not used in the pediatric population due to concern for systemic adverse effects and limited long-term efficacy. (2)(26) Other treatment options for AA include topical minoxidil (off-label), phototherapy, topical immunomodulators, and topical immunotherapy, including squaric acid dibutyl ester and diphencyprone. (1)(22)(23) Off-label use of topical minoxidil for AA may increase hair growth in the areas where applied but requires consistent long-term use. In addition, the efficacy and safety of topical minoxidil has not been well established in children. Children with particularly extensive disease may respond well to topical squaric acid dibutyl ester and diphenylcyclopropenone; however, there are adverse effects that may be difficult for some children to tolerate, including blistering, dermatitis, hyperpigmentation, and lymphadenopathy. (1)(22)(23)(27) Tofacitinib, a Janus kinase inhibitor, has recently been used successfully in children with AA in oral and topical forms; however, the efficacy rate is unknown. Studies are ongoing at the time of this publication. (1)(28) Unfortunately, despite these treatment options, AA continues to be difficult to treat, and relapse is often not preventable. Psychological counseling, support groups, and nonprofit organizations that provide hair prostheses are helpful in decreasing the burden of AA. (2)

NUTRITIONAL DEFICIENCIES ASSOCIATED WITH HAIR LOSS

Protein-Energy Malnutrition

Protein-energy malnutrition disorders include kwashiorkor, marasmus, and marasmic kwashiorkor. In the United States, cases have been reported related to "food fads," milk alternatives such as the substitution of rice "milk," food allergen avoidance, child abuse, and malabsorption disorders. (5) Hair symptoms seen in these disorders include thinning and alopecia, as well as hypopigmentation of hair and changes in texture. The "flag sign" of kwashiorkor is a notable hair finding that appears as alternating bands of dark and light hair color. (5) Approximately 80% of children with protein-energy malnutrition disorders have at least 1 hair symptom, with thinning and hypopigmentation being seen most often on clinical examination. (29) Resolution of the malnutrition corrects the hair loss in time. (5)

Vitamin C Deficiency (Scurvy)

Although perifollicular hemorrhage and follicular-based hyperkeratotic papules with corkscrew hairs are most often seen in patients with scurvy, hair loss has also been reported. (29) Diffuse nonscarring alopecia in infants has been suggested as an indicator of early scurvy because the typical cutaneous signs of scurvy are often not seen in infancy. (30)

Iron and Zinc Deficiency

Low iron and zinc levels have been associated with various types of alopecia, including AA and TE. Supplementing may aid in hair regrowth in patients with identified deficiencies. (29)

HEAVY METAL POISONING AND OTHER TOXIC AGENTS

Lead, copper, arsenic, mercury, selenium, and thallium poisoning have all been associated with hair thinning and alopecia. (29)(31)(32) Recent evidence supports that toxic levels of thallium, mercury, selenium, and colchicine are also often associated with hair loss. (32) Opiate use is important to consider as well because thallium and other heavy metals may be added to opiates by drug dealers to increase the weight of the product. (32)(33)

HAIR CYCLE DISORDERS

Telogen Effluvium

TE is a characteristically temporary, nonscarring diffuse hair loss condition that occurs when an increased number of hairs transition prematurely from the actively growing phase (anagen phase) to resting hair (telogen phase), resulting in hair density thinning and diffuse shedding. (1)(2)(34) TE can be triggered by stressors such as illness, surgery, trauma, medications, nutritional deficiencies, or metabolic disturbances, although in up to one-third of cases there is no known cause. (1)(2)(34)(35)(36)(37) TE often presents within 4 months of the triggering episode. In some cases, patients may not present until after the initial shedding episode, when regrowth has ensued. (1)(2) In these patients, on clinical examination, there will be regrowth seen in the frontal and temporal scalp, but patients will often describe a period of hair loss. (1)(2) In many cases TE may also coexist with other hair loss disorders. Diffuse AA, female pattern

hair loss (FPHL), and androgenetic alopecia (AGA) can present similarly to TE and should be ruled out. (1)(2)(38)(39) TE is often indicated by a positive hair pull test depending on when the patient last washed his or her hair. Patients suspected of having TE should be given a hair pull test, especially of the vertex scalp, to determine the amount of hairs in the telogen phase. (1)(2)(34)(36) Patients should also receive a complete blood cell count, metabolic panel, testing for lead exposure, and testing for nutritional deficits, including iron, zinc, biotin, and protein deficiency, if indicated by history and physical examination findings. (1)(2)

In most patients, TE resolves without treatment, reversing once the underlying triggers are addressed or resolved. Thus, observation and reassurance are most often recommended. Hair regrowth is commonly seen during the first year, although growth can occur at a slower rate. (1)(2) Rarely, some patients may develop chronic TE lasting 6 months or longer, which may not resolve on its own. Currently, treatment options are limited for patients with chronic TE. (1)(40)(41) In adults, treatment with topical minoxidil has shown varied response. (40)(41) It is unclear whether topical minoxidil is safe or effective in children with TE. (2)

Anagen Effluvium

Anagen effluvium (AE) is a hair cycle disorder in which shedding occurs during the normally active growing anagen phase, resulting in a diffuse nonscarring alopecia. (1) AE is rare in healthy pediatric patients and presents with profound, rapid hair loss involving most or all of the hair on the scalp, representing the large number of hairs in the anagen phase. (1)(34)(42) In patients with AE, anagen hair often has a tapered appearance, with breakage at the proximal hair shafts. (1)(34)(43) Similar to TE, AE is often preceded by a triggering event. Patients with a history of chemotherapy or radiotherapy, severe malnutrition, infection, autoimmune disease, or exposure to toxins are more likely to develop AE than are healthy patients. (1)(34)(42)(43)(44)(45)

Hair regrowth is observed in many patients within 3 months of resolution of the underlying cause or cessation of the offending medication. (1)(43) Unfortunately, some patients do experience permanent hair loss.

Treatment options remain limited in pediatric patients with AE. In adults, topical minoxidil, scalp compression, and scalp hypothermia have been beneficial in decreasing hair loss in patients receiving chemotherapeutic agents. It is unknown whether these treatments are safe or effective in children. (1)(43)(46)(47)

Loose Anagen Syndrome

Loose anagen syndrome (LAS) is an uncommon disorder in which there is early keratinization of the hair follicle's inner root sheath, resulting in weak attachment of anagen hair to the hair follicle unit. (1)(34)(48)(49)(50) LAS often manifests as painless loss of anagen hairs, with patients typically presenting from birth to early childhood with slow-growing hair or hair loss. LAS has been reported to be more common in females aged 2 to 6 years, although it may be underdiagnosed in males due to differences in hairstyling practices. (48)(49)(50)(51)(52) Parents often report that their child's hair has an odd texture and is unruly, frizzy, or "lusterless" in appearance (Fig 5). (49)(53)

LAS has a known genetic basis, with most cases reported as autosomal dominant with incomplete penetrance, although parents are often unable to recall whether they were affected as a child. LAS may also be caused by sporadic mutations. Mutations involving keratin have been identified in several familial cases of LAS. In addition, hereditary disorders such as Noonan syndrome, coloboma, and neurofibromatosis have been reported in association with LAS. (1)(54)(55)(56)(57)(58) LAS has also been reported in association with AIDS as a form of AIDS trichopathy. (52)

Patients are most often diagnosed by clinical examination. The hair pull test is a valuable, although not specific, diagnostic tool for LAS. Children with LAS have greater than 3, and often more than 10, loose anagen hairs on the hair pull test, whereas normal results would be only 1 to 2 loose anagen hairs. (53) Trichography and light microscopy are also helpful in diagnosing LAS, with trichography showing the presence of more than 50% loose anagen hairs and light microscopy showing a ruffled hair cuticle. (54)

In most patients with LAS the disorder has a self-limiting and benign course, with spontaneous hair regrowth even without treatment, although this may take several years.

Figure 5. Texture of hair in a child with loose anagen syndrome.

(1)(34)(49)(55)(56) Off-label use of topical minoxidil has been beneficial in decreasing hair loss in patients and can be particularly useful for patients with severe LAS; however, its safety is not well established in children. (49)(54)(57) Patients should also be counseled to limit the use of hair products and avoid styling that can aggravate hair loss. (1)

Short Anagen Syndrome

Short anagen syndrome (SAS) is a congenital hair cycle disorder in which hair does not grow long due to an underlying shortened anagen phase and a subsequent increase in telogen hairs. (1)(59)(60)(61) Children usually present between 2 and 4 years of age with short hair that is of decreased to normal density. Parents often notice that their child's hair has been short since birth without needing to be cut. (59) SAS may present similarly to the short hair seen in patients with LAS; however, patients with SAS will exhibit hairs of similar length that shed in the telogen phase, and patients with LAS have hairs of varying lengths that shed in the anagen phase. (1) Other areas of hair on the body may be normal. (61) Similar to LAS, SAS typically has a self-limiting and benign course, with most patients improving during puberty. (1)(59)(60)

TRAUMATIC HAIR LOSS

Trichotillomania

Trichotillomania is a relatively common condition that features chronic, repetitive, and compulsive urges to pull out one's own scalp, eyebrow, and/or body hair. (1)(2)(34) Trichotillomania can be debilitating and result in significant distress and functional impairment in addition to hair loss. (62)(63) The onset of trichotillomania is typically between 9 and 13 years of age. (64)(65)(66) Whereas in adults trichotillomania is more frequently seen in females, in childhood the female/male ratio has been shown to be fairly equal. (62)(63) Children usually present with nonscarring patches of hair loss that exhibit broken hairs of different lengths, irregular borders (often in unique, geometric shapes), and an absence of scale or excoriation (Fig 6). (1)(2)(65)(66) Trichotillomania can cause scarring and permanent hair loss in advanced, persistent cases. (1) Hair loss patches are more frequently seen on the side contralateral to the dominant hand. (1)(67) Hair pulling can occur at any site with hair, but most commonly involves the scalp (72.8%), eyebrows (56.4%), and pubic region (50.7%). (63) Of note, the occipital scalp is often not involved because pulling hair in this region is more painful. (1) Hair density is typically normal, and the hair pull test will be negative in patients with trichotillomania. (1)(68) Trichoscopy aids in the diagnosis of trichotillomania because there are specific findings that are highly suggestive of the condition, including irregularly broken hairs of varying lengths, 2 hairs that are broken at the same length and coming from the same follicular opening ("the V sign"), and coiled hairs. (5)(69)(70) Other causes of patchy, nonscarring hair loss, such as AA and noninflammatory TC, are important to include in the differential diagnosis of trichotillomania. (1)(2) Findings suggestive of trichotillomania versus AA include patches of broken hair of differing lengths, sparing of the occipital scalp, and the absence of patches of total hair loss. (1)(2) KOH and Wood lamp examination can help in ruling out noninflammatory TC. (70) Hair loss due to child abuse has been reportedly misdiagnosed as trichotillomania, and it is recommended that the possibility of abuse be considered during assessment. (71)

Some patients with trichotillomania may eat their pulled hairs and are at risk for developing bezoars, which can present with various gastrointestinal symptoms, including abdominal pain, nausea, vomiting, and bowel perforation. (72)

In the *Diagnostic and Statistical Manual of Mental Disorders, Fifth Edition (DSM-5)*, trichotillomania is now grouped with obsessive compulsive disorder (OCD), excoriation

Figure 6. Nonscarring patches, irregular borders, and an absence of scale or excoriation seen in trichotillomania.

disorder, body dysmorphic disorder, and hoarding disorder. (63) Patients with trichotillomania have been found to have higher rates of OCD, mood, anxiety and other mental health disorders in first-degree relatives. (63)(65)(66)(73)(74) Evidence suggests that there is a genetic component to trichotillomania. (2)(63)

The treatment and outcome of trichotillomania depend on the patient's age at presentation. In preschool children, trichotillomania is often self-limiting and may be characterized as a habit disorder that often resolves before the child enters grade school, with conservative management through behavioral modification. (1)(2) Adolescents who present with trichotillomania often have other psychiatric comorbidities. It is recommended that such patients undergo mental health evaluation and behavioral therapy with an experienced provider. (1)(2)(65)(66)

Although trichotillomania is classified with OCD in the *DSM-5*, treatment approaches differ. There is currently no universally accepted first-line pharmacological intervention for trichotillomania. (63) Psychotropic medications for trichotillomania show variable response in adults, with a paucity of data in pediatric populations. (1)(65) There is currently no strong evidence of a treatment response with selective serotonin reuptake inhibitors. (63) N-acetylcysteine, a glutamatergic agent, and the antipsychotic agent olanzapine have demonstrated some benefit in treating trichotillomania in adults. (63)(75)(76) Cognitive behavioral therapy approaches have been shown to be effective in treating trichotillomania in pediatric patients, particularly habit reversal therapy with a trained therapist. (63) There is currently not enough data to recommend the use of off-label topical minoxidil in patients with trichotillomania.

Traction Alopecia

TA develops as a result of repeated or prolonged pulling forces on the hair, usually due to hairstyling or grooming practices. (1)(2) Although TA has been reported in children in the first year of life, the prevalence increases with age and is most common in adult women. (77)(78) Women of African descent have higher rates of TA. (2)(78) Children with TA typically present with hair loss involving the temporal scalp; however, the frontal and occipital scalp can also be involved in some patients. (79) Some patients may present with acute-onset patchy hair loss, and others may present with thinning hair in areas of traction. Signs typical of TA include perifollicular pustules and papules, broken hairs, scaling, and perifollicular erythema in the early stages. (1)(2)(78) Late-stage TA is more likely to present with scarring and permanent hair loss. (78) The "fringe sign"—the presence of a margin of thin retained hairs along the frontal or temporal hairline—is a sensitive and specific clinical finding in both early and late TA. (77)(78) Another useful clinical finding in diagnosing TA is the presence of "tenting"—elevation of the scalp skin into a tent shape when the hair is pulled tightly. (77)(78)(80) Most patients with TA are unaware that their hairstyling is applying excessive tension and causing the hair loss. (78)(79) When specifically asked about hairstyling practices, most patients will recall symptoms related to hairstyling, such as tenderness or the development of pimples and crusting. (78) Specific hairstyles associated with TA include ponytails/pigtails, braids, chignons, twists, tight buns, cornrows, dreadlocks, sisterlocks, weaves, extensions, and curlers. (1)(2)(77)(78)(79)(80) In addition, chemical relaxing and straightening products can also increase the risk of developing TA. (78)

Treatment primarily consists of implementing new hair care practices to avoid tension. Loosening the hairstyle is recommended, as well as avoiding heat and chemicals. (1)(2) Oral or topical antibiotics are recommended in patients with concurrent folliculitis, (34)(78) and topical or intralesional corticosteroids can be useful in patients with inflammation. (78)(81) Topical minoxidil has shown promise in promoting hair growth in patients with TA. (78) Many patients can experience regrowth of hair within months of treatment, although some patients with severe or chronic TA may have irreversible hair loss. (1)(2)

Discoid Lupus Erythematosus

Discoid lupus erythematosus (DLE) is an autoimmune disorder that rarely presents in pediatric patients but can present as an acquired scarring alopecia. (1)(82) Patients with DLE develop scaly, erythematous, inflammatory lesions that most frequently involve the head and neck. The scalp skin may be thin and shiny, with prominent telangiectasias. These patches of inflammation can scar and cause irreversible hair loss. (1) DLE lesions may be misdiagnosed as TC due to similarities in initial presentation and are usually diagnosed based on typical scalp biopsy findings. (1) Unlike in adults, children with DLE often do not test positive for antinuclear antibodies. (82)(83) However, children with DLE are more likely to progress to develop SLE, with studies reporting 26% to 40% of patients progressing to SLE. (82)(83) In contrast to adults, DLE seems to affect male and female children at similar rates. (1) Patients with DLE may be more likely to have a family history of autoimmune or rheumatologic disease. Treatment options focus on slowing disease activity to decrease scarring and include hydroxychloroquine and topical, intralesional, or systemic corticosteroids. (1) Monitoring for the development of systemic disease is recommended in pediatric patients with DLE. (82)(83)

Other Scarring Alopecias

Additional types of scarring alopecia include central centrifugal cicatricial alopecia, lichen planopilaris, and frontal fibrosing alopecia. These conditions are progressive, scarring hair disorders that primarily affect adult women. Although rare, these scarring alopecias have been reported in pediatric and adolescent patients and may be underreported. (84)(85)(86)(87)(88)(89)(90)(91) Management can be challenging because there is currently no gold standard pharmacologic treatment. (86)(87)(88)(89)(90)(91)

APLASIA CUTIS CONGENITA

Aplasia cutis congenita (ACC) is a rare congenital deficiency of cutaneous tissue, most often involving the scalp. Patients typically present at birth with a single circumscribed scalp lesion and associated scarring alopecia, but in rare cases lesions may occur on the trunk and limbs. (1)(92)(93) Lesions may be covered by a membrane or ulcerated, with complications including hemorrhage, thrombosis, and infection. (1)(92)(94) If lesions are covered by a membrane or surrounded by a hair collar, patients are more likely to have underlying defects, and further imaging is recommended. In most patients, supportive care is recommended because over time most lesions will epithelialize spontaneously. (1) Patients are usually left with a permanent scar and absence of hair growth of the affected area. ACC may be difficult to differentiate from nevus sebaceous, a benign hamartoma that can have a similar presentation. ACC most often presents in isolation with a focal absence of skin and hair, or it can occur as a heterogenous group of syndromes with varying severity. (92)(94)(95)(96) Trisomy 13, Johanson-Blizzard syndrome, Adams-Oliver syndrome, and Wolf-Hirschhorn syndrome have all been associated with ACC. (1)(92)(96) Fetal exposure to methimazole, misoprostol, methotrexate, angiotensin-converting enzyme inhibitors, benzodiazepines, valproic acid, and cocaine have also been linked to ACC. (92)(95) ACC may be excised in older adolescence or adulthood if the area is cosmetically concerning to the patient.

TRANSIENT NEONATAL HAIR LOSS

Transient neonatal hair loss (TNHL), also known as neonatal occipital alopecia, is a common phenomenon in which neonates develop occipital alopecia and often other patches of hair loss. (1)(2) This is a benign condition that most frequently occurs in healthy babies younger than 3 months and is felt to be a physiological shedding of hair. (97)(98) Although TNHL was originally thought to be related to friction and sleep position, recent evidence suggests that there is no relationship between sleep position and development of TNHL. (97)(98) No treatment is necessary for TNHL because hair will regrow spontaneously over time.

CONGENITAL TRIANGULAR ALOPECIA

Congenital triangular alopecia (CTA), previously known as a Brauer nevus or temporal triangular alopecia, is a benign, nonscarring, and nonprogressive alopecia. CTA is characterized by triangular, oval, or lancet-shaped patches of alopecia, usually involving the frontotemporal scalp. (1)(99) The patches of hair loss seen in CTA tend to be left-sided and unilateral in approximately 80% of patients. (99)(100)(101) Most patients present with CTA at 2 to 9 years of age, although it can present at birth. (1)(102)(103) In rare cases, CTA can be associated with other disorders, including phakomatosis pigmentovascularis and trisomy 21. (99) Although CTA does not progress, it often remains stable, and hair regrowth is not always possible.

CONGENITAL ATRICHIA

Congenital atrichia is a rare condition that results in the complete absence of hair, usually due to mutations in the human hairless gene (HR) inherited in an autosomal recessive manner. (1)(34)(104)(105) Patients typically present with a total absence of hair at birth, but in some patients hair loss may occur later in life due to programmed follicular destruction. (1)(34)(106) Congenital atrichia is often also associated with papular lesions involving the scalp, face, neck, trunk, and limbs. (1)(104)(105)(106) Other syndromes that may be associated with congenital atrichia include progeria, Moynahan syndrome, and hidrotic ectodermal dysplasia. (106)

CONGENITAL HYPOTRICHOSIS

Congenital hypotrichosis is an inherited hair loss disorder that typically presents in children 2 years and older with decreased hair volume or extremely thin hair. Hair thinning is usually diffuse, but the amount of hair loss and the quality of hair can vary. (1)(34) Similar to congenital atrichia, mutations involving the hairless gene are often responsible for congenital hypotrichosis, although it is less severe than congenital atrichia. (1)(34)(107) Marie Unna hereditary hypotrichosis is a form of congenital hypotrichosis that is inherited in an autosomal dominant manner, with progressive hair loss that starts around puberty. (108)(109) Congenital hypotrichosis can also be associated with inborn

errors of metabolism, epilepsy, Ehlers-Danlos syndrome, juvenile macular degeneration, skeletal and dental abnormalities, and other chromosomal disorders. (107)(109) Genetic counseling is often beneficial in these cases. Unfortunately, in many cases patients may not respond to treatment. (1)(109)

ANDROGENETIC ALOPECIA

AGA is considered the most prevalent cause of hair loss in adults, presenting as nonscarring, progressive alopecia that typically involves androgen-dependent areas of the scalp (the vertex, temporal, midfrontal, or parietal scalp). (1)(34)(110)(111)(112)(113)(114)(115) FPHL is often considered a form of AGA that occurs in women and presents with a pattern that differs from what is seen in male pattern AGA. Patients with FPHL typically experience thinning of the central scalp, and, in contrast to male patients with AGA, rarely experience thinning of the frontal, temporal, or vertex scalp. (110)(115)(116)(117) AGA and FPHL may be seen as early as adolescence. AGA is believed to be mediated by dihydrotestosterone receptors of the hair follicle unit, causing miniaturization of hair follicles in genetically susceptible individuals. (110) Evidence suggests that the androgen receptor gene may be involved in the pathogenesis. (113) AGA is rare in pediatric patients with normal androgen levels before puberty, although cases have been reported in pediatric patients who had a strong family history of AGA. (1)(34)(111)(112)(113)(114)(115)(118) Trichoscopy and family history are valuable tools in making the diagnosis. A biopsy is usually not necessary but often will show signs of inflammation. (110)(115) It is suggested that boys with early-onset AGA and adolescent girls with AGA receive a full hormonal evaluation. (115) Patients with congenital adrenal hyperplasia, polycystic ovary syndrome, and other conditions associated with hyperandrogenism may have an increased risk of AGA or FPHL. (115)(119)(120) Topical minoxidil is the most common treatment recommended in adolescent patients with AGA. (114)(115)

ACQUIRED HAIR SHAFT DISORDERS

Hair shaft disorders encompass a variety of congenital and acquired disorders that result in structural hair defects, typically presenting with increased hair breakage, difficulty with growing the hair, or changes in hair appearance and texture. (1)(121) History, trichoscopy, and hair pull and tug tests can all aid in diagnosing hair shaft disorders. (1)(34)(122)(123)(124) Hair shaft disorders with fragility are more common and usually acquired. (121) Trichorrhexis nodosa is a common acquired hair shaft disorder that is usually caused by damage from hair products or styling. In trichorrhexis nodosa, there are characteristic points where the hair is weak and breaks easily. (122) Another acquired hair shaft disorder is known as *bubble hair abnormality;* exposing hair to high temperatures can create gas bubbles in the hair shaft. (122)(124) Patients with acquired hair shaft disorders should be counseled to avoid hair products and styling practices that can damage the hair shaft. Improvement is expected after discontinuing use of the underlying product or practice, but this can take years. (122)

CONGENITAL HAIR SHAFT DISORDERS

Congenital hair shaft disorders are rare and include trichothiodystrophy, trichorrhexis invaginata, monilethrix, pili torti, pili trianguli et canaliculi, and trichorrhexis nodosa. (34)(121)(122)(125) Various genetic mutations and genetic associations have been identified in each of the congenital hair shaft disorders. Genetic counseling is recommended for patients with a suspected congenital hair shaft disorder. (121) Gentle hair care, such as avoiding chemical products, braids, heat, brushing, and teasing, is recommended for all hair shaft disorders associated with fragility. (121) Unfortunately, there are limited treatment options. Some disorders, including pili torti, trichorrhexis invaginata, and pili trianguli et canaliculi, may improve as the child ages. (121) Monilethrix and trichorrhexis nodosa can improve with topical minoxidil therapy, and oral retinoids have been used to treat both monilethrix and trichorrhexis invaginata. (121)

Summary

- Hair loss in children can be divided into congenital/hereditary or acquired causes. Congenital or hereditary causes of hair loss in children can be further divided into localized/focal or diffuse causes. Acquired causes of hair loss in children can be divided into scarring or nonscarring. Nonscarring acquired causes can be inflammatory, patchy, diffuse, or localized/focal. (1)
- Most hair loss in children results from nonscarring, acquired conditions, such as tinea capitis (TC), telogen effluvium, alopecia areata (AA), traction alopecia, or trichotillomania. (1)(2)
- TC is the most common cause of hair loss in children. Based on strong research evidence as well as consensus, TC should be ruled out in children

presenting with hair loss because chronic untreated TC can progress to severe inflammation and permanent scarring. (1)(2)(8)

- AA is an autoimmune-mediated type of alopecia that is relatively common. Based on some research evidence as well as consensus, patients with AA may be at an increased risk for other autoimmune disorders, particularly autoimmune thyroid diseases and vitiligo. (19)(20)(21)(22)(23)(24)
- Based primarily on consensus due to lack of relevant clinical studies, routine screening for autoimmune disease is often not indicated in otherwise healthy patients with AA and a negative review of systems. (21)
- Based on some research evidence as well as consensus, hair products can contribute to hair loss in patients with traction alopecia, trichorrhexis nodosa, and loose anagen syndrome. Patients with acquired hair shaft disorders should be counseled to avoid hair products and styling practices that can damage the hair shaft. (1)(2)(77)(78)(79)(80)(122)
- Based on some research evidence as well as consensus, alopecia is often distressing, and it is important to provide psychosocial support while counseling patients and their families about prognosis and treatment. (1)

SUGGESTED QUALITY IMPROVEMENT PROJECT:

- Decrease use of hair fungal cultures when considering pretest probability likelihoods.

References for this article can be found at http://pedsinreview.aappublications.org/content/41/No. 11/570.

PIR QUIZ

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1. A 4-year-old African American boy is seen in the clinic with multiple patches of hair loss on the scalp. On close examination of these patches, the examining clinician noted broken hairs and black dots. Which one of the following is the most likely diagnosis in this patient?

   A. Atopic dermatitis.
   B. Psoriasis.
   C. Seborrheic dermatitis.
   D. Telogen effluvium.
   E. Tinea capitis.

2. A 5-year-old girl has been steadily losing hair, and now hair loss involves her entire scalp. You suspect alopecia totalis. The mother is very worried and would like to start treatment right away. Which one of the following is the best next step in management?

   A. Intradermal corticosteroid injections.
   B. Oral griseofulvin.
   C. Oral corticosteroids.
   D. Scalp biopsy.
   E. Topical corticosteroids.

3. A 2-year-old boy recently adopted from Afghanistan is seen in the office. The child is noted to be below the 3rd percentiles for height and weight. On physical examination he appears malnourished. He is noted to have alternating dark and light hair color. He is diagnosed as having failure to thrive. Which one of the following is the most likely cause of the hair finding in this patient?

   A. Iron deficiency.
   B. Kwashiorkor.
   C. Lead poisoning.
   D. Scurvy.
   E. Zinc deficiency.

4. You are seeing a 15-year-old girl who recently immigrated from Guatemala. She was diagnosed as having pulmonary tuberculosis and is currently receiving treatment. In addition to some initial weight loss, the parents report that the patient has significant diffuse hair loss. Test results for nutritional deficiencies and complete blood cell count, metabolic panel, and lead levels are all normal. You attribute the hair loss to telogen effluvium based on a hair pull test. Which one of the following is the best next step in management?

   A. Reassurance.
   B. Scalp compression.
   C. Scalp hypothermia.
   D. Trial of topical minoxidil.
   E. Trial of topical corticosteroids.

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5. A 10-year-old girl is diagnosed as having trichotillomania based on a history of repetitive pulling of her hair and physical examination demonstrating broken hairs of different lengths. Based on the *Diagnostic and Statistical Manual of Mental Disorders, Fifth Edition (DSM-5),* this patient's diagnosis is part of the manifestations of which one of the following psychiatric conditions?

   A. Anxiety.
   B. Mood disorder.
   C. Obsessive compulsive disorder.
   D. Oppositional defiant disorder.
   E. Schizophrenia.

Hair Loss

Reese L. Imhof, Dawn Marie R. Davis and Megha M. Tollefson

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