
Hair Loss in Children

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Abstract

Hair diseases represent frequent complaints in dermatology clinics, and they can be caused by a number of conditions reflected by specific diagnoses. Hair loss is not uncommon in the pediatric group, but its patterns in this group are different from those seen in adults. Additionally, in children, these disorders can have psychological effects that can interfere with growth and development. Hair is easily accessible for examination, and dermatologists are in the enviable situation of being able to study many disorders using simple diagnostic techniques. To fully understand hair loss during childhood, a basic comprehension of normal hair growth is necessary. Knowledge of the normal range and variation observed in the hair of children further enhances its assessment. This chapter has been written in an attempt to facilitate the diagnostic process during daily practice by helping to distinguish between acquired and congenital hair diseases. It can sometimes be difficult to differentiate between abnormality and normality in neonatal hair aspects. Management of hair disorders can be quite a daunting task for the attending physician and mandates a holistic approach to the patient. Some hair disturbances have no effective treatment, and for others, no single treatment is 100% successful. If no effective treatment for a hair loss disease exists, a cosmetic approach is important.

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Introduction

Hair loss in children occurs in a wide range of conditions that may be congenital or acquired. A congenital abnormality may be an isolated finding in a healthy patient or may occur as a feature of a multisystem syndrome. Recognition of a hair disorder may enable the diagnosis of a particular syndrome. The clinical presentations of pediatric hair disorders range from subtle to disfiguring. Alopecia is not uncommon in the pediatric population but has patterns that are different from those seen in adults. The occurrence of these problems during childhood can cause psychological and emotional stress to both children and their parents. A good knowledge of the normal hair cycle, embryology and clinical features is necessary.

Embryology and Normal Hair Development

Hair follicles are derived from an interaction between the embryological ectoderm and mesoderm, which begins at 9 weeks of gestation. Primary follicles first develop on the eyebrows, upper lip and chin. Then, hair follicles develop over

the scalp in a frontal to occipital direction and progress over the body in a cephalocaudal direction [1]. By 18–20 weeks of gestation, the entire initial population of follicles has formed, including those on the scalp [2]. Each follicle is capable of producing three different types of hair as follows: lanugo, vellus and terminal. Lanugo hair is nonmedullated, fine, soft, and usually nonpigmented, and it can be found on the bodies of newborns. This type of hair is shed by 3–4 months after birth. Vellus hair is short, fine, light-colored, barely noticeable, and covers almost the whole body. During puberty, the androgen hormone causes most of vellus hair to turn into terminal hair. Terminal hair is larger, thicker and strongly pigmented, and it is found on the scalp, eyebrows, axillary and pubic areas, chest and face [1].

Human hair grows in a continuous cyclic pattern known as the hair cycle. The hair cycle presents with different phases as follows: the anagen or growth phase (85–90% of hairs), the catagen or regression phase (<1% of hairs), the telogen or resting phase (4–15% of hairs) and, finally, the shedding exogen phase. At birth, about 5 million hair follicles cover the human body, and approximately 100,000 are scalp hairs [2]. Newborn hairs are all anagen, and during childhood, there is a gradual transition of scalp hairs from vellus to terminal hairs [3].

Diagnosis

A newborn can have the following three presentations of hair (normal variants): a full head of hair, or little or no hair. The beginning of abnormal hair growth sometimes occurs during infancy; thus, it is difficult to predict which newborn will have a hair pathology. Knowledge of a patient's personal and family history, a thorough clinical examination, as well as general and specific diagnostic procedures are important for a correct diagnosis and early treatment [4]. A de-

tailed history is essential for an accurate diagnosis. The key points in a patient's history are the following: age of onset (congenital or acquired); onset of hair loss (sudden or insidious); extent of alopecia (localized or diffuse); physical and mental development (underlying syndrome?); past medical history (surgery, autoimmune disease, or medication); psychiatric disorders; and family history of alopecia [3, 5]. Similar to the history evaluation, an exhaustive physical examination should be performed to assess the following components: type of alopecia (localized or generalized/scarring or nonscarring); hypotrichosis or alopecia; hair shaft anomalies; hair quality; and hair color. A thorough scalp examination is also important to evaluate the existence of erythema, edema, pustules, scaling, atrophy or scarring.

The presence of short stature, abnormal bone development, defective hearing, dysmorphic features, impaired vision or other physical findings could indicate an underlying metabolic or autoimmune disease.

Usually, the diagnosis of the most common forms of hair loss can be made only by clinical and physical examinations. A hair pull test distinguishes between loss from follicles and loss due to hair shaft fragility. To perform this test, around 50–60 hairs are grasped between the index finger and thumb and then lifted with gentle traction. The pull test is considered positive if more than 10% of hairs are released [5]. False positives can occur if the test is performed on a day in which hair has been washed.

To confirm breakage of the hair shaft, a tug test should be performed. The tug test consists of the grasping of a hair between the finger and thumb near its exit from the scalp and the pulling of the distal part [6]. If the hair is fragile, a fracture will occur in the shaft. Trichoscopy, a noninvasive method, has emerged as a valuable tool in the differential diagnosis of most hair and scalp diseases. It is also important to evaluate the therapeutic response of hair loss [7].

Alopecia Areata

Alopecia areata (AA) is a nonscarring, autoimmune and inflammatory pattern of alopecia that occurs in both children and adults. It is a common disorder, affecting different ethnicities with equal incidences between genders. The prevalence of AA is approximately 0.2% in the general population, and its lifetime risk is estimated to be approximately 1.7% [8]. Although AA has been considered rare in young infants, recent studies have suggested that this disorder could occur in 1–2% of patients less than 2 years of age [9]. In children younger than 16 years of age, AA has been reported to occur in 21–24% of patients [9, 10].

The predisposition to AA is genetically determined, with 5–25% of patients possessing a strong family history [10, 11]. There is also an increased frequency of AA in individuals with Down syndrome [12–15]. AA may be seen in association with autoimmune disorders, such as atopic dermatitis, allergic rhinitis, asthma, vitiligo and autoimmune thyroid disease [13].

There are several clinical presentations of AA, which are usually classified according to the hair loss pattern or extent. The classic presentations are as follows: patchy AA (the most common pattern), alopecia totalis (the complete absence of terminal scalp hair), and alopecia universalis (the total loss of terminal scalp and body hair) [13]. Less frequent patterns of AA include the reticulated pattern, ophiasis type, sisaipho type and diffuse thinning over a part of or the total scalp [16, 17]. Another variant, acute diffuse and total alopecia, has been described and is characterized by rapid progression and extensive involvement along with a good prognosis.

The typical appearance of AA is of a well-demarcated localized patch that is asymptomatic, round or oval with a smooth surface. There may be single or multiple lesions with no associated epidermal changes, such as scaling. In some cases, slightly reddened skin can be present. AA can af-

fect the scalp or any hair-bearing area on the body. This disorder is associated with nail involvement, including nail pitting, trachyonychia, brittle nails, onycholysis and koilonychia [16–18].

Trichoscopy examination reveals the presence of short ‘exclamation mark’ hairs at the periphery of the lesion (pathognomonic of AA) and ‘yellow dots’ in a follicular distribution. The hair pull test is typically positive in active AA with the presence of telogen club hairs and dystrophic anagen hairs [5, 19]. A scalp biopsy is usually unnecessary to establish the diagnosis of AA, except in the case of diffuse shedding. The hallmark histological finding is a dense lymphocyte infiltrate comprising mainly T cells around the anagen hair bulb matrix and the dermal papillae. The main differential diagnosis of AA in children includes tinea capitis, trichotillomania (TTM), transient neonatal hair loss (TNHL) and congenital triangular alopecia.

Tinea capitis is a common cause of patchy hair loss in infants, although individuals of all ages are occasionally affected. This condition involves the invasion of scalp hairs by dermatophyte fungi, including *Trichophyton* and *Microsporum* species. The clinical picture of this disease varies according to host immunity, the degree of inflammatory response and the type of hair invasion by the pathogen. The key feature is patchy hair loss with various degrees of inflammation and scaling and the easy removal of hairs from the affected area [19]. A trichoscopic hallmark of this disease is the presence of comma hairs. Fungal potassium hydroxide preparations and cultures and Wood’s lamp are also helpful in establishing the diagnosis of tinea capitis. Oral antifungal treatment is required to treat this disease. The aim of treatment is a clinical and mycological cure [20].

TTM is classified as an impulse control disorder and is a self-inflicted compulsion or habit-tic to pull or pluck at the hair. It is characterized by single or multiple patches with the presence of

broken hairs of different lengths. TTM can be more difficult to differentiate from AA [21] (see Traumatic Alopecia for more information).

TNHL, which is also known as 'neonatal occipital alopecia', appears as a bald patch on the occipital region. Initially, it was thought to be secondary to friction due to babies sleeping in the supine position. According to a prospective study, 20% of neonates are born with an observable deficiency of the occipital scalp hair. This entity can be present at birth and is related to the physiology of hair shaft shedding. TNHL appears in healthy babies from birth until approximately the second month of life without accompanying symptoms and with spontaneous resolution [2]. It is important to inform parents of the benign course of TNHL and its absence of a relationship with the sleeping positions of babies.

Congenital triangular alopecia is also known as temporal triangular alopecia or Brauer nevus [22, 23]. It is a localized, congenital, nonscarring form of alopecia that might be present at birth or acquired during the first 10 years of life. Typical lesions are triangular or lancet shaped, can be unilateral or bilateral, are several centimeters in width and are confined to the front temporal region [23]. Apparently, the lesions are hairless, although very fine vellus hairs may be seen. Trichoscopy can aid in the differential diagnosis of this disease by the observation of normal follicular openings containing vellus hairs and the absence of specific features associated with AA [7].

There is no consistently reliable treatment for AA. Its natural history is uncertain, and spontaneous remission occurs in some patients.

Depending on the extent of involvement and the patient's age, watchful waiting is often a sensible approach. In the majority of children with patchy AA, hair will regrow entirely within 1 year without treatment. The aim of therapy is cosmetically acceptable hair regrowth.

Therapeutic options available for AA in children are more limited than for adults. Topical corticosteroids are commonly used for the treat-

ment of this disease and are the first-line choice of many dermatologists.

Topical corticosteroids may be applied painlessly and have benign side-effect profiles [1]. Topical potent fluorinated corticosteroids remain an acceptable form of treatment for children with AA but have been reported to have some effects [19, 24, 25]. Children younger than 10 years of age with AA of recent onset tend to be the most responsive [19]. If hair growth occurs, this treatment should be continued with regular monitoring to prevent cutaneous atrophy [26]. Although rare, the development of systemic effects must also be monitored in children. Relapse after discontinuation of treatment could happen [27]. Injectable triamcinolone acetonide can be useful in children with patchy hair loss. Children younger than 10 years of age are not usually treated with intralesional corticosteroids because of the pain caused by its injection [16]. Older children (>10 years) may be prepared to consider this treatment for limited areas of hair loss if they are able to tolerate the injections [19].

Systemic corticosteroids have been considered in the treatment of widespread AA or AA that is refractory to other local treatments, but careful reviews of the protocols (doses, lengths of treatments and side effects) are mandatory.

Topical minoxidil is widely used in children and adults for the treatment of AA. Systemic absorption of the drug is typically minimal with topical therapy. Although side effects are rare, they have to be taken in consideration, especially in young children. According to Herskovitz et al. [28], a 2-year-old male patient developed generalized hypertrichosis after 2 months of treatment with 5% minoxidil foam for AA. This report highlights the risk of serious cutaneous or systemic side effects due to the possibility of the systemic absorption of topical minoxidil.

Other treatments for AA include topical immunotherapy (topical agents that induce hair growth by provoking allergic contact dermatitis,

such as dinitrochlorobenzene, squaric acid dibutylester, and diphencyprone), anthralin (induces inflammatory irritant dermatitis), phototherapy, immunosuppressants and immunomodulators [1]. Widespread forms of AA are generally more difficult to cure, and no treatment appears to prevent relapse.

An earlier onset of AA may be indicative of a more widespread disease, although it is impossible to predict its severity. In patients with few patches of alopecia, prognosis is generally good, and regrowth might occur within 6–12 months. Indicators of a poor prognosis are the presence of immune disease, atopy, a family history of AA, young age at onset, nail involvement, extensive hair loss and an ophiasis pattern [17]. The course of AA is unpredictable, and response to treatment can be changeable.

Androgenetic Alopecia in Children

Androgenetic alopecia (AGA) is a nonscarring, patterned alopecia with a typical pattern of distribution and with genetic involvement [29]. Family history predisposes individuals to the early development of this disease and rapid progression of the alopecia. The onset of AGA is gradual, slowly developing over a period of years. For its development, the presence of androgens is necessary in combination with genetically susceptible hair follicles. The onset of AGA is not expected to occur in children without abnormal androgen levels [30]. If a healthy prepubertal child presents with AGA, an endocrine evaluation is strongly recommended [30]. AGA is not uncommon in adolescents and should be considered as a cause of hair loss. Minoxidil topical solution appears to be effective and well tolerated in adolescent boys and girls with AGA [31]. Finasteride has not been studied in the treatment of males younger than 18 years of age; thus, its safety and efficacy in adolescents with AGA has not been determined.

Disturbances of Hair Cycle

Anagen Hair Loss: Anagen Effluvium and Loose Anagen Syndrome

The hair follicle undergoes periods of cyclic growth in the following phases: the anagen or growth phase, the catagen phase, the telogen phase and the shedding exogen phase. At any given time, 85–90% of follicles are in the anagen phase. During this phase, mitotically active matrix cells in the hair bulb differentiate and divide, resulting in hair growth [32]. The duration of the anagen growth phase of scalp hair varies from 2 to 6 years [1]. About 1% of scalp hairs are in the catagen phase, which lasts about 3 weeks. Approximately 10% of follicles are in the telogen resting phase, which lasts about 3 months, after which these hairs are shed. The final step of the hair cycle, exogen, is when the hair is released from the follicle. Anagen loss results from the shedding of a large amount of hairs during the anagen phase. The daily loss of telogen hairs is considered normal, but if a loss of anagen hair occurs, potential causes should be promptly analyzed. The causes that affect children are the same as those affecting adults, and a similar evaluation should be performed.

Anagen Effluvium

In anagen effluvium, hair loss is profound because up to 90% of scalp hair is normally in anagen. Hair loss characteristically occurs within 1–2 weeks of the insult.

The most frequent and easily recognizable cause of anagen effluvium is radiotherapy and systemic chemotherapy (e.g. doxorubicin, cyclophosphamide, vincristine, or bleomycin) [33]. Other causes that must be considered are exposure to toxic agents, such as mercury and colchicine, boric acid intoxication (e.g. exposure or ingestion of household pesticides), and ingestion of certain plants (*Lecythis ollaria* and *Leucaena glauca*) [34]. Severe protein malnutrition can also lead to anagen effluvium.

Diagnosis is easily made according to the patient's clinical history and the presence of dystrophic anagen hairs. If the insult is removed, normal hair regrowth is expected because the hair cycle was only temporarily interrupted.

Loose Anagen Syndrome

In loose anagen syndrome, the anchoring of growing anagen hairs to follicles is impaired, with a lack of adhesion of the hair shaft to the hair follicle [19]. Because the growing anagen hairs are not anchored normally, they can be easily and painlessly plucked from the follicle.

Children between 2 and 7 years of age are the most frequently affected with this syndrome. At birth, the hair is sparse but appears normal, with no fragility or breakage [35]. Around 2–3 years of age, the hair becomes unruly and remains so until it spontaneously becomes normal at 5–7 years.

The hair of affected children does not grow long, and parents commonly state that there is no need for a haircut. Using light microscopy, hairs with distorted anagen roots without inner root sheaths and ruffled cuticles can be observed [35]. Trichogram analysis reveals a preponderance of anagen hairs (>97%) [36].

During childhood, gentle handling decreases hair shedding. No treatment is necessary because the hair spontaneously reverts to normal around puberty, although some physicians have reported the use of a 5% minoxidil solution causing clinical improvement [37, 38].

Telogen Effluvium

Telogen effluvium is an abnormality of hair cycling that can occur at any age. It is a reaction pattern in which a percentage of hairs move prematurely from anagen to telogen, resulting in a diffuse increase in hair shedding. This shedding occurs in response to a pathologic or physiologic alteration in the health condition.

The main causes of telogen effluvium are high fever, surgery, drugs (allopurinol, colchicine, beta-blockers, or antihypertensives), systemic ill-

nesses, endocrine disorders, nutritional disorders (protein-calorie malnutrition, zinc deficiency, or starvation), severe emotional stress, immunization (following bivalent human papillomavirus) [39] and pregnancy. Acute telogen effluvium occurs approximately 3 months after a triggering event. Chronic telogen effluvium is considered when shedding continues longer than 6 months. A detailed history should be sought, including a thorough drug history, and full clinical examination should be performed [40]. A hair pull test is positive when a high percentage of telogen hairs is obtained (>20%) [5]. This test should be performed at the vertex, parietal and occipital areas [33].

Telogen effluvium is less frequent in children than in adults, and in children, it is more likely to be related to sudden disease or trauma. This condition is managed by the treatment of the underlying disorder with appropriate replacement drugs or medical therapy [19]. The prognosis of telogen effluvium is very good if the precipitating event is eliminated.

Traumatic Alopecia

Trichotillomania

TTM is characterized by repetitive and self-induced compulsive hair pulling [41]. The most frequent site is the scalp, but it can also involve the eyebrows, eyelashes and pubic hair. During infancy and early childhood, TTM is more frequent in boys, while during puberty and in adults, there is a strong female predominance [42].

Clinically, patients present with areas of alopecia of different shapes, irregular borders and the presence of hairs of different lengths [21]. This condition is usually a nonpermanent form of alopecia, but if the same area is persistently plucked, scarring may result with persistent hair loss [1]. Typically, there is no scaling on the scalp, although some excoriations can be seen. Hair density is normal, and the pull test is negative [5].

Trichoscopy reveals the presence of flame hairs (specific for TTM) with the v-sign and tulip hairs, which are highly characteristic features of TTM [43].

Three subsets of individuals affected by this condition appear to exist as follows: preschool age children, preadolescents to young adults, and adults. In young children, TTM is usually a habit comparable to thumb sucking, and it normally has a self-limited and benign course of hair pulling with regrowth of the hair if the condition is managed conservatively. Several reviews of childhood TTM have related its onset in young children to stressful situations. This type of traumatic alopecia is more frequent in preadolescents to young adults with an average age of onset of between 9 and 13 years [21]. These patients tend to have more chronic and relapsing courses of hair pulling.

TTM in adults may be secondary to underlying psychiatric disturbances and has a more prolonged course.

Management of this disorder is difficult and is usually based on a person's age. No specific treatment has been established that is completely effective. Preadolescents to young adults without associated psychiatric conditions may benefit from nonpharmacological interventions, such as behavior modification programs. If TTM is associated with psychological/psychiatric disorders, referral to a psychiatrist for evaluation or treatment is recommended. Pharmacological interventions include serotonin-specific re-uptake inhibitors, tricyclic antidepressants and more recently, N-acetylcysteine, a glutamate modulator [44].

Traction Alopecia

Traction alopecia, a pattern of alopecia caused by physical trauma, is induced as a consequence of constant traction while hairstyling (e.g., tight ponytails, cornrows or rollers). It is more frequent in black women and children. Afro-textured hair is particularly easily damaged by such procedures.

Clinical manifestations include hair thinning, focal decreases in hair density and perifollicular erythema [45]. Short, broken hairs, folliculitis and follicular papules may be seen [1]. With prolonged traction, perifollicular scarring and cicatricial alopecia develop, which are more evident along the hairline. During its early stages, alopecia is initially reversible with a change in hairstyling methods. If patients maintain continuous traction, permanent hair loss occurs due to hair follicle loss.

Hair Shaft Disorders

Hair shaft disorders can be inherited or acquired and sometimes can aid in diagnosing an underlying disease. A reliable diagnosis of a hair shaft abnormality can only be made via an evaluation of the wide structural variations found in normal hair.

Hair shaft disorders can be divided in two subcategories as follows: (1) hair shaft abnormalities WITH increased fragility and breakage; and (2) hair shaft abnormalities WITHOUT increased fragility. Table 1 has a description of the principal hair shaft abnormalities.

Special Disorders with Hypotrichosis in Children

There is an extensive list of genetic disorders associated with hypotrichosis. The detailed analysis of these syndromes falls outside of the ambit of this chapter.

Congenital Atrichia and Hypotrichosis

Atrichia congenita is a rare form of irreversible alopecia with an autosomal recessive mode of inheritance that is usually associated with a mutation in the human hairless gene located on chromosome 8 [55, 56].

This condition is characterized by follicular agenesis or programmed follicular destruction.

Table 1. Hair shaft disorders

Disease	Genetic/clinical features	Diagnosis	Associated diseases	Management
<i>Hair shaft abnormalities WITH increased fragility and breakage</i>				
Trichorrhexis nodosa [46–48]	<p>Congenital or Acquired (++)</p> <p>Acquired TN is frequently due to weathering due to external causes, such as shampooing, styling, UV radiation, wetting and natural friction</p> <p>Hairs appear dry and brittle with a tendency to break at different lengths</p> <p>Three variants of acquired TN:</p> <p>Proximal ATN: seen in patients after years of hair straightening</p> <p>Distal ATN: acquired, cumulative cuticular damage</p> <p>Circumscribed ATN: scalp, moustache or beard</p>	<p>Light microscopy:</p> <ul style="list-style-type: none"> – nodes along the hair shaft that cause hair to break easily; distal nodes generally indicate hair weathering; and the absence of cuticle cells <p>Trichoscopy:</p> <ul style="list-style-type: none"> – light magnification: nodular thickenings along hair shafts that appear lighter in dark hair shafts – high magnification: small fibers with an appearance suggestive of the ends of two brushes aligned in opposition 	<p>Mental retardation</p> <p>Argininosuccinic aciduria</p> <p>Muenke syndrome</p> <p>Defective teeth and nails</p> <p>Its presence in infants or young children should trigger a search for an underlying metabolic problem</p>	<p>Change in hairstyling procedures</p> <p>Sun hair protection</p>
Monilethrix (beaded hair) [1, 49, 50]	<p>Congenital (AD): mutation in the hair cortex-specific keratin genes <i>KRT86(++)</i>, <i>KRT81</i>, <i>KRT83</i>, and <i>DSG4</i></p> <p>Clinically, there is normal hair at birth, but within the first months of life, a characteristic moniliform aspect of the hair shaft develops.</p> <p>Perifollicular erythema and follicular hyperkeratosis are common features</p> <p>The hair is short, fragile, brittle and breaks spontaneously or as a result of friction</p> <p>In its mildest forms, only the scalp is involved. In extensive cases, there may be eyebrow, eyelash and nail involvement</p>	<p>Light microscopy: hairs possess a beaded appearance</p> <p>Trichoscopy:</p> <ul style="list-style-type: none"> – uniform elliptical nodes and intermittent constrictions of the hair shaft, causing variation in hair shaft thickness – hairs are bended regularly at multiple locations and have a tendency to fracture at constriction sites – periodic constriction of the hair shaft 	<p>Keratosis pilaris</p>	<p>No specific treatment</p> <p>Retinoids, glycolic acids and minoxidil may be helpful in some cases</p> <p>There is a wide spectrum of severity, and it tends to remain constant throughout life</p>
Pili torti [50, 51]	<p>Congenital (AD (++)/AR) or acquired: isolated or associated with other conditions</p> <p>At birth, the hair is normal. Changes gradually appear between the 3rd month and 3rd year. More common in girls with blond hair</p> <p>Clinically, hairs are fragile, with a twisted and spangled appearance</p>	<p>Light microscopy: twists at irregular intervals along the shaft</p> <p>Trichoscopy: hairs are twisted and rotated at 180°. In a field of view, only part of the hair usually appears abnormal. Twists are better seen with dry trichoscopy using high magnification</p>	<p>Björnstad syndromes (pili torti associated with sensorineural hearing loss)</p> <p>Muenke disease</p> <p>Bazex-Dupré-Christol's syndrome</p> <p>Crandall syndrome</p> <p>The presence of pili torti, requires further evaluation for neurological and ectodermal disorders</p>	<p>No treatment</p> <p>May improve in puberty</p>
Trichorrhexis invaginata (bamboo hair) [50–52]	<p>Congenital (AD): mutation in the <i>SPINK5</i> gene</p> <p>Clinically, the hair is short, brittle, sparse and very fragile. May affect the scalp, eyebrows and body hair</p> <p>Defective cornification of the cortex</p>	<p>Light microscopy: intussusceptions of the distal shaft into the proximal shaft, with a bamboo appearance</p> <p>Trichoscopy: golf tee hair (scalp and eyebrow)</p>	<p>Marker for Netherton's syndrome (AR; triad of ichthyosis, atopic diathesis and trichorrhexis invaginata)</p>	<p>No specific treatment</p> <p>Avoidance of trauma</p>

Table 1. Continued

Disease	Genetic/clinical features	Diagnosis	Associated diseases	Management
Trichothiodystrophy	Congenital (AR), characterized by sulfur-deficient hair Clinically, only the hair may be affected (fragile) or it may be associated with ichthyosis, photosensitivity (50%), decreased growth, and mental handicap	Light microscopy: irregular, undulating contours and transverse fractures throughout the hair shaft ("tiger tail") Trichoscopy: non-specific Diagnosis is performed by sulfur and/or amino acid analysis of the hair	BIDS or Tay's syndromes (brittle hair, intellectual impairment, decreased fertility, and short stature) IBIDS (ichthyosis and BIDS) PIBDS syndrome (photosensitivity and IBIDS)	No treatment currently available Sun protection
<i>Hair shaft abnormalities WITHOUT increased fragility</i>				
Pili annulati (ringed hair) [46, 53]	Rare Congenital (AD) or sporadic: abnormalities appear at birth or during infancy Clinically, the hair looks shiny but otherwise normal, and 20–80% of the hair is affected No fragility, and the hair can grow long	Light microscopy: hair shafts with alternating bright (normal hair) and dark bands (abnormal hair; the medulla is expanded by air cavities within the hair) Trichoscopy: hair shafts with regular light bands corresponding with dark cavities, visible by light microscopy	No hair or systemic abnormalities	No treatment currently available Aesthetic defect
Woolly hair syndrome [46, 50]	Three types of woolly hair: Hereditary woolly hair (AD): mutations in plakoglobin genes and desmoplakin genes. Hair color is variable Familial woolly hair (AR): mutations in the <i>LPAR6</i> and <i>LIPH</i> genes. Hair is sparse, thin, and short Woolly hair nevus: localized Clinically, hair is fine and tightly curled. Occurs in Caucasians	Light microscopy: ovoid cross sections, 180-degree longitudinal twisting, trichorrhexis nodosa and pili annulati Trichoscopy: Crawling snake with short wave cycles and broken hairs	Hereditary woolly hair: palmoplantar keratoderma and cardiac abnormalities Familial woolly hair: associated with hypotrichosis Woolly hair nevus: associated with linear epidermal or pigmented nevi (>50%)	No specific treatment Can improve with age
Uncombable hair (pili trianguli and canaliculi) [46, 54]	Congenital (AD): hair abnormalities appear around the age of 3 years Clinically, the hair is unruly, dry and impossible to control with a brush or comb	Light microscopy (gold standard): in more than 50% of hairs, there is a triangular or reniform shaft with a longitudinal groove or flattening Trichoscopy: triangular or reniform hair shaft	Absence of systemic abnormalities	Improves spontaneously with aging

TN = Trichorrhexis nodosa; AD = autosomal dominant; AR = autosomal recessive; DSG4 = desmoglein 4; LIPH = lipase H.

Hair at birth can be sparse (or inexistent) and progress to a total and permanent absence of scalp hair over the first 5 years. In another variant of the disease, neonates are born with lanugo hair, which is shed during the first few months of life and is never replaced [19, 50]. Congenital atrichia may occur, either as an isolated phenomenon or

in association with a number of rare syndromes. To be distinguished from AA totalis, a scalp biopsy may be required.

Congenital hypotrichosis is a less severe form of atrichia congenita, in which hair is not absent but is diffusely thinned [19]. There is a profound reduction in the number of hair follicles on the

scalp, which usually but not always is present from birth. Hypotrichosis may not be noticed until the age of 2 years because of variation in the quality and quantity of the hair normally present at birth, and it usually occurs as an isolated defect.

Marie-Unna Hereditary Hypotrichosis

Marie-Unna hypotrichosis is an autosomal dominant condition, which was first described in 1925 by Dr. Marie Unna in 27 affected members of a family over seven generations [50]. This rare hypotrichosis is characterized by isolated progressive alopecia without abnormalities of the nails or teeth or intellectual or gross motor development. Typically, this diffuse hair defect occurs as an isolated phenomenon, although some reports have described of its associations with Ehlers-Danlos syndrome and juvenile macular degeneration. At birth, hair may be normal or sparse. Around the third year of life, hair becomes coarse and twisted, resembling an 'ill-fitting wig' [19, 57]. During puberty, the hair is gradually lost from the crown, ultimately resulting in complete baldness due to the destruction of the follicles with scarring [19]. Lashes, eyebrows and body hair are also sparse or absent.

Ectodermal Dysplasia

Ectodermal dysplasia represents a heterogeneous group of inherited disorders affecting more than one ectoderm-derived tissue, leading to abnormalities of the hair, nails, epidermis, teeth and eccrine glands. Scalp hair is usually fine and short but silky in texture [19]. Eyebrows and/or eye-

lashes are also affected. These patients should be examined thoroughly, with particular attention paid to other ectodermal structures.

Conclusion

Hair problems in children occur relatively frequently in a wide range of conditions that may be congenital or acquired. Not all congenital or hereditary hair disorders are present at birth, and those that are often go unrecognized. Usually, these hair disorders are isolated abnormalities, and the child is otherwise well. A number of disorders occur as part of a multisystem syndrome; thus, recognition of the clinical presentation may enable the diagnosis of a particular syndrome. If a disease is hereditary, genetic counseling should be offered. A detailed clinical history accompanied by examination are essential for the accurate diagnosis of the pathology.

Hair is very important, both cosmetically and functionally. Alterations in the normal appearance of hair can predispose individuals to low self-esteem and a negative body image. Pediatric hair disorders causes psychological and emotional stresses in both children and parents. It is important that parents are provided a clear understanding of the etiology and natural history of the disease.

The management of a hair loss disorder should be adapted according to the age of the patient. An early identification of the pathology leads to better treatment results, including psychological and cosmetic support. Unfortunately, for many hair disorders, there is no effective, reliable therapy.

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