ELSEVIER

Disorders of Hair in Infants and Children Other Than Alopecia

MARTIN S. WADE, MB, BS
RODNEY D. SINCLAIR, MB, BS

Hair disorders in children encompass a wide range of conditions with varied pathology that can be either congenital or acquired. Of the congenital conditions, the hair abnormality may be either an isolated occurrence in an otherwise healthy child or one of the signs composing a clinical syndrome. There is a vast array of such syndromes, of which most are rare. Discussion here is limited to the more common and those syndromes that have a hair abnormality as a prominent feature.

When describing any pathological condition, it is essential to have an accurate concept of what constitutes the norm to allow any deviation from this to be easily recognized. This is often difficult when dealing with children's hair, as there is a great degree of variation in what is accepted as normal. Social and cultural influences also play a part. The amount of scalp hair seen in normal, healthy newborns is one example, and some conditions such as hypotrichosis may not be recognized until after early infancy for this very reason.

Hair disorders range from subtle to particularly disfiguring and may have far-reaching consequences. Abnormal hair may be more upsetting to the parents than the younger child, who may not yet have the self-awareness to realize that a problem exists. One parent may blame the other, especially when an inherited or infective cause is found. Siblings may develop unsupported fears that they, too, may be involved. If the condition is disfiguring, the affected child will almost certainly, at some stage, become the victim of ridicule at school, which may result in lifelong psychological problems. In such cases, it is important to ensure that parents take a proactive approach by meeting with teachers at school entry.

Management of hair conditions in children requires a holistic approach. Children with head lice, for example, need to be treated personally as well as other close contacts, the school informed, and the child kept at home until treatment has commenced. When there is no known effective medical treatment for a particular condition, such as hereditary hypotrichosis, cosmetic camouflage methods can be used while parents are told of the natural history and offered genetic counseling.

From the Department of Medicine and Dermatology, St. Vincent's Hospital, Fitzroy, Victoria, Australia.

Address correspondence to Martin S. Wade, MB, BS, St. Vincent's Hospital, Department of Medicine & Dermatology, Fitzroy Victoria 3065, Australia.

E-mail address: sinclair@svhm.org.au

Embryology and Normal Hair Development

Human hair growth is cyclic, with each follicle producing many distinct hairs during a lifetime. There are approximately 5 million hair follicles on the body, of which, on average, only 100,000 are on the scalp. There is some variation in the number of scalp follicles, ranging from about 70,000 to 150,000 for brunettes, with blondes having 10% more and redheads 10% less.[1] Most of the follicles on the body produce vellus hairs that are cosmetically insignificant, and many do not even protrude from the skin's surface.

Hair follicles develop as epidermal down growths that invaginate the dermis and subcutaneous fat and enclose at their base a small stud of highly specialized dermis known as the dermal papilla. Follicles exist as pilosebaceous units that also give rise to the sebaceous glands, arrector pili muscles, and, in certain areas, apocrine glands. The hair growth rate is relatively constant at about 1 cm/mo, and hairs appear on the skin's surface in groups of three, known as Meijeres trio group. The hair follicles first form on the eyebrow, upper lip, and chin at between 9 and 12 weeks gestation, and at birth the full complement of hair follicles is present. Hair follicle formation occurs in a frontal to occipital wave on the scalp and a cephalocaudal wave on the body, reaching the feet at about 22 weeks. The first hair grows from the follicle between weeks 16 and 22. The hair grows for about 10–12 weeks to a length of 2–3 cm and covers the entire body from head to toe. These fine and nonpigmented hairs are known as lanugo hairs. The growth then terminates in similar frontooccipital and cephalocaudal waves, with the follicles entering telogen via the involutional catagen phase. Telogen terminates with the development of the second hair bud forming at the base of the bulb. New anagen hair growth results in the commencement of shedding of the first coat of hair between weeks 32 and 36. At full term, there are two consecutive waves of hair on the scalp, each of which is running from the forehead to the

655 Avenue of the Americas, New York, NY 10010

0738-081X/02/$–see front matter
PII S0738-081X(01)00226-7

Table 1. Most Common of Tinea Capitis by Continent

Europe	*Australia*	*N. America*	*S. America*	*Africa*	*Asia*
M. canis	*M. canis* *T. tonsurans* (Aborigines)	*T. tonsurans* *M. canis* *M. audouinii*	*T. violaceum* *M. canis*	*T. violaceum* *T. soudanense* *M. audouinii* *M. canis* *T. yaoundi*	*T. violaceum*

occiput. Over the occiput, the primary hairs do not enter telogen until 8–12 weeks after birth and then fall, producing a well-defined area of alopecia. This has been described as occipital alopecia of the newborn.

The second hair produced by the follicle is different from the first. Site variation is introduced, with the growth phase of the scalp hairs progressively elongating and the body hairs shortening to the point where most of the hairs are shorter than the first. These grow for about 4–8 weeks, reaching a length of about 1 cm uniformly over the body. The second coat is shed during the first 3–4 months of life, as the second set of hairs pass through telogen and are replaced by the third set.

On the body, the third hair is smaller than the second hair and many no longer protrude from their pore or do so only as fine, light, vellus hairs. In contrast, the scalp hair follicles enlarge and produce thicker and more pigmented hairs known as terminal hairs.

This site specificity also mirrors how follicles react to pubertal androgens. Scalp hairs miniaturize in response to androgens while body hairs enlarge. The degree of enlargement is highly variable, with pubic and axillary hair being most sensitive and most capable of enlarging in response to physiological levels of adrenal androgens. Facial, chest, thigh, abdominal, and buttock hair require at least physiological levels of gonadal androgens but are more likely to appear with abnormally high levels of either adrenal or ovarian androgens. There is enormous person-to-person variation in the response of follicles to circulating androgens, and this is influenced by a variety of genetic and metabolic factors.

The fourth and subsequent hairs from scalp follicles continue to enlarge and elongate until a steady state is achieved. The average duration of anagen is about 3 years, producing hairs that would grow to a final length of approximately 36 cm; however, the range is large and may extend to 6 or 7 years. By late adolescence, the anagen duration tends to be fixed and remains constant in the absence of an acquired disorder such as androgenetic alopecia.

The synchrony of neonatal hair growth is progressively lost, and the wave of hair growth and shedding from the frontal to occipital scalp is disturbed and takes on a random, mosaic pattern. This contrasts to our mammalian counterparts which have a seasonal molt, while humans tend to shed only a few hairs each day.

In summary, there are four main types of hair produced in the fetus and young child. These are as follows:

1. Lanugo hairs, which are formed and shed during the seventh and eighth months in utero. These are fine, soft, unpigmented hairs that have no central medulla.
2. Vellus hairs, which are also fine, unmedullated, and unpigmented hairs shorter than 2 cm.
3. Terminal hairs, which are coarse, pigmented, long hair found on the scalp, eyebrows, and eyelashes before puberty and additionally pubic, axillary, chest, and beard areas after puberty.
4. Indeterminate hairs also exist on the scalp of infants at 3 months and last until the age of 2 years. These are coarser than lanugo hairs, are sparsely pigmented, and have a poorly defined medulla. Hair leaves the scalp at an angle that is precisely determined, so that streams and pattern are formed. Whorls and streams appear as early as 12 weeks of gestation and can usually been seen on the parietal scalp. A single parietal whorl can been seen in 95% of babies and it is usually clockwise. The remainder have two or rarely three whorls. A frontal cowlick due to counter stream of hair from the forehead is present in 7% of babies.

Abnormal scalp hair patterns with absent or aberrant whorls may be seen on the heads of children with abnormal brain development, such as microcephaly. Displacement of the scalp line occurs in a number of syndromes. A congenitally low anterior hairline occurs in Cornelia de Lange syndrome, lipoatrophic diabetes, fetal hydantoin syndrome, and Rubinstein-Taybi syndrome, while a low posterior line occurs in Noonan's and Turner's syndromes. A congenitally high anterior hairline is seen in myotonic dystrophy.

At puberty there is a transition from vellus to terminal hair in certain regions, contributing to the secondary sexual characteristics. Initially this is long and downy but then develops into coarser and more curly hair. The onset of pubic hair growth is earlier in girls, with the mean age of onset being 11.7 years compared with 13.4 years for boys.[2] Axillary hair follows some 2 years later. Facial hair in boys appears at approximately the same time, commencing at the corners of the lips. Terminal body hair then develops in a fairly predictable manner, commencing with the lower legs, thighs, forearms, ab-

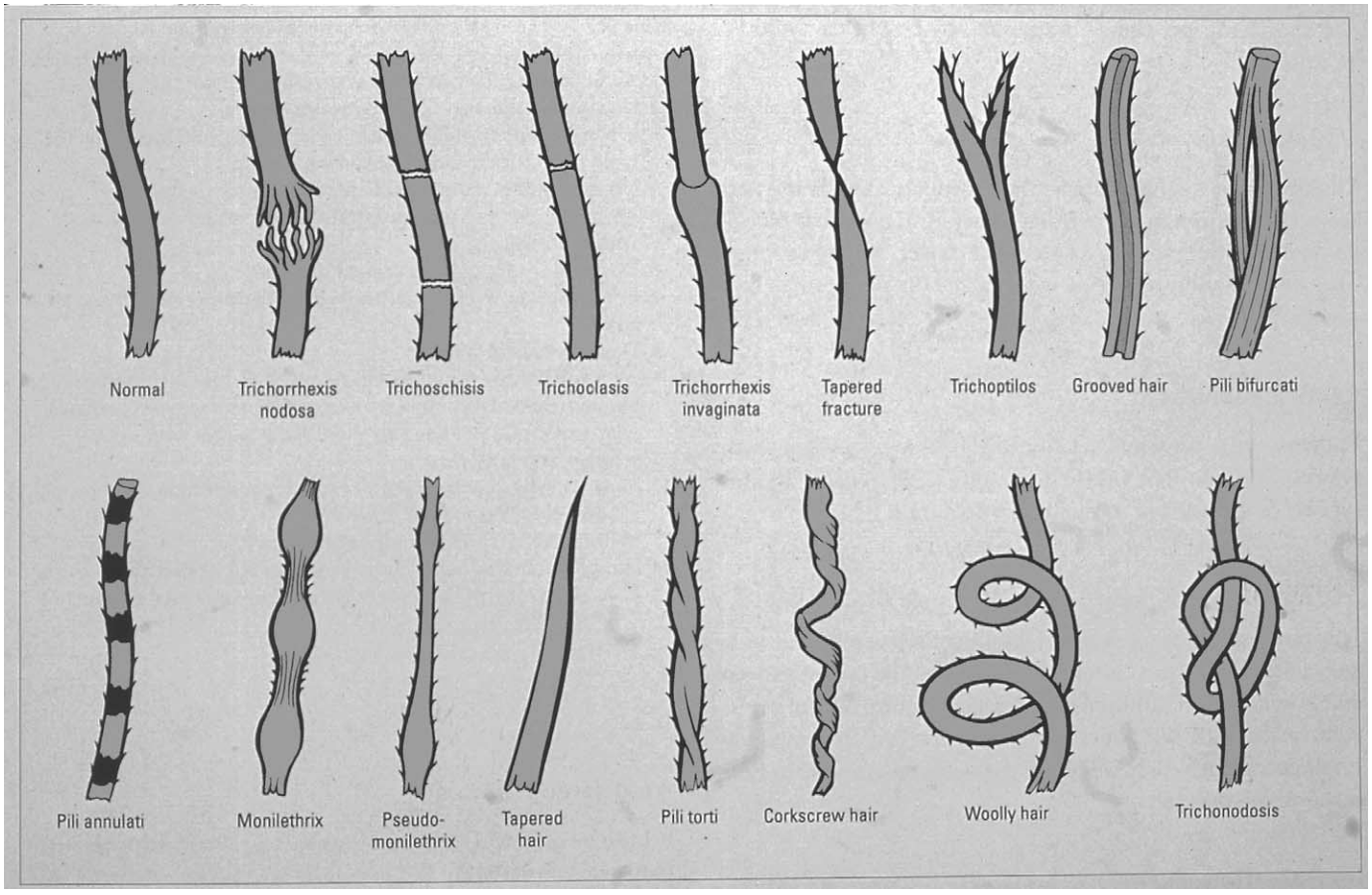

Figure 1. *Graphic representations of various hair disorders.*

domen, buttocks, chest, back, upper arms, and then the shoulders.[3]

Hypertrichosis

Hypertrichosis is defined as an excessive growth of hair on skin not normally hairy. The word "growth" is emphasized, as the number of hair follicles present is not altered. Hypertrichosis indicates a difference in the quality and length of the hair that is produced from the follicle. Although the word "excessive" is subject to cultural and racial influences and personal preferences, most cases are self-evident. The pathogenesis of hypertrichosis predominantly involves elongation of the anagen phase. Although some degree of hair follicle and hair fiber enlargement may be seen, it is commonly not pronounced.

Unlike hirsuties, the hair growth in hypertrichosis is not androgen driven and the pattern does not mimic hair growth in men. The desire to distinguish hypertrichosis from hirsuties is based on practical considerations: hypertrichosis is not androgen dependent and does not respond to antiandrogen therapy. Thus, the natural history, prognosis, and response to treatment of these two conditions are different. Unfortunately, the distinction between hypertrichosis and hirsuties is not clear clinically in all cases, and the two conditions are also not mutually exclusive.

Hypertrichosis can be classified as generalized or localized and can occur as an isolated phenomenon or as part of a syndrome. There are many causes, including hereditary, congenital, or drug induced, or as a result of a neuroectodermal abnormality. Of these, an acquired, generalized form, usually referred to as prepubertal hypertrichosis, is the most common in children.

Localized Hypertrichosis

Hairy Ears, Eyelashes, Eyebrows, and Antecubital Fossae

Congenital hairy ears have been reported in otherwise-normal infants, in babies with XYY syndrome, and in babies born to diabetic mothers whose sugar control appears to be unrelated.[4] Extremely long eyebrows or eyelash hairs are usually familial and may be first noticed in infancy. This may also be the case with eyebrows that join in the center, known as synophrys, which, when present, usually occurs in an otherwise-normal child but may be associated with a syndrome, such as Cornelia de Lange, Rubinstein-Taybi, or con-

genital trichomegaly with dwarfism.[5] Long eyebrows are commonly the most noticeable aspect of drug-induced hypertrichosis due to minoxidil, cyclosporin, or diazoxide.

In familial hypertrichosis cubiti, excessive hair develops in infancy over the lower third of the upper arm and the upper third of the forearms. The hypertrichosis is progressive for a few years before partially regressing around puberty. The mode of inheritance has not been defined, and an association with short stature is seen in 10% of cases.[6]

Faun Tail

Faun tail refers to a congenital, midline, well-demarcated dorsal patch of hypertrichosis. The relevance of this form of localized hypertrichosis is that it may indicate an otherwise-occult spinal abnormality, such as spina bifida occulta.[7] The hair is usually pigmented, with long, silky hair overlying normal skin. Within the tuft of hair, however, there may be a sinus or dimple representing incomplete formation of the spinal tract and resulting in a possible portal of entry for infection. Less commonly, a lipoma or capillary nevus may be associated.

Spinal dysraphism (failure of spinal fusion) underlying the faun tail is four times more common in girls than boys and usually results in the spinal cord's becoming transfixed by bone. There may be no neurological deficit at birth, but with the differential growth of the vertebrae and spinal cord with time, traction is placed on the cord and neurological abnormalities begin to manifest in the first 5 years of life. Permanent neurological deficits can be prevented in most cases with early prophylactic surgery. The faun tail may occur at any level of the vertebral column but is most common in the sacrum. The growth of hair is presumed to be due to an as-yet-unidentified growth factor secreted by neural tissue.

Hair Collar Sign

Of presumably similar etiology is the hair collar sign, which refers to the collar of fast-growing, long, terminal hairs that surround a bald nodule of skin on an infant's scalp. The nodule may contain heterotopic brain tissue and usually overlies an embryonic fusion line. This ring of hypertrichosis has also been reported with other neuroectodermal defects, including dermoid cysts, dermal sinus tumors, encephaloceles, and meningoceles. Communication with the brain must be considered and excluded with imaging and is considered more likely if the cyst is seen to fill when the baby cries.

Inclusion Dermoid Cyst of the Scalp

Inclusion dermoid cysts develop from sequestrated epithelial cells along the lines of embryonic fusion. As the cranial bones grow together, islands of epidermal cells are cut off from the surface epithelium. Dermoid cysts are often lined by an epidermis that contains cutaneous appendages. Fully mature follicles, sebaceous glands, and eccrine glands are often present, whereas apocrine glands and smooth muscle fibers are found in approximately 20% of cases.[8]

The natural history of inclusion dermoid cysts of the scalp is varied; however, the lesions characteristically begin as a small, solitary nodule in the midline or overlying an embryonic fusion line. There is overlying loss of hair, with or without the hair collar sign, and with time the lesion may flatten and eventually scar over.[9] The history of a cyst present at birth allows a flattened lesion to be differentiated from aplasia cutis.

These cysts have a potential for intracranial extension, so before excision, the patient should have a computed tomography scan with bone windows, and those associated with partial-thickness bone defects or intracranial extension should be referred to a neurosurgeon for management.[10] Although some authors advocate that all dermoid cysts should be removed as a routine to prevent potential intracranial infection,[11] the most common reasons for their removal are because they may be enlarging, become repeatedly infected superficially, may discharge fluid, or for cosmetic reasons. Malignant degeneration is exceedingly rare, and prophylactic excision is not indicated.[12]

Congenital Or Acquired Melanocytic Nevi

Localized hypertrichosis may occur secondary to either congenital or acquired melanocytic nevi. The congenital pigmented hairy nevus is probably the most common cause of localized hypertrichosis in infancy. The presence of hair in a nevus bears no relation to its malignant potential. The differential diagnosis with a hairy melanocytic nevus is the hypertrichosis and hyperpigmentation occasionally seen overlying congenital smooth muscle hamartomas. The latter usually shows some induration and may become transiently raised on rubbing. Biopsy is sometimes required to differentiate the two. The lesions of congenital smooth muscle hamartomas tend to decrease in size with time.

Becker's Nevus

This is also characterized by hyperpigmentation and hypertrichosis; however, the pigmentation is not melanocytic in nature. As the lesion progresses the pigmentation may fade, leaving only a patch of hypertrichosis. The lesion usually presents in adolescence and is thought to be due to increased androgen cytosol receptors in the involved skin; however, it has also been reported in infancy. The lesions are usually singular and on the trunk, especially around the shoulder region. Acne may occasionally be present within the Becker's nevus. The incidence is approximately 0.5% and is 10 times more common in men.[13] Multiple lesions are rare, as are associated structural abnormalities.

Epidermal Nevus Syndromes/Depigmented Hypertrichosis
All the well-defined epidermal nevus syndromes are lethal gene syndromes, apart from the nevus comedonicus syndrome. There has been a recent case report of depigmented hypertrichosis following Blaschko's lines, with associated cerebellar and ocular malformations. The patches of hypertrichosis were first noted at 17 months of age on slightly pigmented nevoid skin of the trunk and arms. Hair in some of the patches grew as long as 8 cm. Patches of white hair developed on the scalp at puberty. This may be an example of an autosomal lethal gene surviving in its mosaic form, which would account for its sporadic presentation, asymmetrical distribution of lesions, and the lack of complete organ involvement.[14,15]

Acquired Localized Hypertrichosis
Constant inflammation or irritation may result in localized hypertrichosis, with the most common example being the increased hair growth seen under a plaster cast, which then regresses once the cast has been removed. Chronic dermatoses, however, do not usually result in hypertrichosis.

Generalized Hypertrichosis

Congenital Hypertrichosis Lanuginosa
This is a rare, autosomal dominant condition characterized by the retention and continued synchronized growth of the fetal lanugo hair. One third of cases have been reported to be sporadic.[16] The thick coat of lanugo hair may be present at birth or develop within the first few months of life. The hair is silvery-gray and may grow to as long as 5 cm. The glabrous areas of the body are spared: palms, soles, lips, glans penis, and terminal phalanges. The amount of hair may regress but the norm is for the condition to progress. At puberty, longer lanugo hairs grow in the secondary sexual sites rather than the usual terminal hair. There are usually no associated features.

Congenital Syndromes With Hypertrichosis
Hypertrichosis with gingival fibromatosis, hypertrichosis with osteochondrodysplasia, and X-linked hypertrichosis are rare forms of congenital hypertrichosis and can be distinguished from congenital hypertrichosis lanuginosa by the nonsynchronous growth of hair, the presence of the associated abnormalities, and the fact that the hair becomes pigmented at puberty.

Cornelia de Lange syndrome is rare and consists of generalized hypertrichosis and abundant scalp hair with low frontal and nuchal hair lines. The eyebrows are bushy and meet in the center, and the eyelashes are long and curled. The syndrome may be associated with physical and/or mental retardation, characteristic facies, short digits, a high arched palate, and a distinctive low-pitched cry. Rubinstein-Taybi syndrome is the combination of generalized hypertrichosis, physical and mental retardation, cryptorchism, broad thumbs, and, once again, a characteristic facial appearance. Coffin-Siris syndrome displays generalized, congenital hypertrichosis, prominent eyebrows and long eyelashes, and sparse scalp hair. There is associated severe mental and growth retardation and the congenital absence of the distal phalanges and nails of the fifth fingers and toes.

POEMS, an acronym for *P*eripheral neuropathy, *O*rganomegaly, *E*ndocrine dysfunction, *M*onoclonal gammopathy, and *S*kin changes, is associated with a generalized yet patterned hypertrichosis on the extensor surfaces of the limbs, the malar region, and the forehead. Hyperpigmentation and edema may also be seen in the affected regions.

Prepubertal Hypertrichosis
This is the most common form of hypertrichosis found in children. It is usually present at birth and increases in severity during early childhood. The extent is variable and subject to ethnic and racial interpretation. On the face, the eyebrows are bushy and there is terminal hair on the temples spreading across to the forehead. There is increased growth of terminal hair on the back and proximal limbs. The history of the timing of onset and the distribution usually allow prepubertal hypertrichosis to be differentiated from hirsuties in the postpubertal girl.

Drug-Related Hypertrichosis
Generalized hypertrichosis is quite common with the use of some systemic drugs. The main drugs implicated are minoxidil, cyclosporin, phenytoin, and diazoxide. With minoxidil it occurs in 70% of children,[17] and the stimulation of vellus hair growth is seen over the face, shoulders, and extremities. It appears weeks to months after the commencement of treatment and regresses within months of ceasing the medication. Cyclosporin-induced hypertrichosis is seen in 60% of treated children,[18] is more common in children and adolescents compared with adults, and is more common again in children being treated for graft-versus-host disease. The hair growth is diffuse and resolves several months after stopping treatment.

Prolonged use of corticosteroids can induce hypertrichosis that is most marked on the forehead, temples and sides of the cheeks, back, and extensor surfaces of the arms. Steroid-induced asthma may also be associated with this hypertrichosis.

Treatment

When a precipitating cause is apparent, removal of this stimulus will result in resolution of the unwanted hair growth. Physical methods of hair removal can be used for congenital or other causes of acquired hypertrichosis. The easiest is bleaching with hydrogen peroxide to camouflage unwanted pigmented hairs; however, this

may be impractical when the area involved is large or the hairs particularly long. Plucking and waxing stimulate reactivation of the resting telogen hairs into anagen and so only provide a short delay before the new hair emerges. Both of these methods are painful and may cause folliculitis, especially where the hair is curly or kinky. Shaving and chemical depilatory creams remove only the portion of the hair fiber that protrudes above the surface of the skin, and reemergence of hair, though less noticeable, is almost instant. Depilatory creams act by dissolving keratin and often irritate the skin, which also contains keratin. Home epilators are, in reality, no more effective than plucking. Electrolysis offers a degree of permanent hair removal by delivering a brief pulse of electricity through a needle inserted into the ostium to cauterize the dermal papilla. It is expensive, time-consuming even for small areas, can be painful, and may result in scarring. Multiple treatments are required, and up to 80% regrowth can be seen in an area treated only once.

Psychological counseling may be required for the more severe, generalized cases or those that pose a difficult cosmetic problem. This counseling should be aimed at patient and peer acceptance of the condition and attempt to allow the child to socialize and interact normally.

Hirsuties

Hirsuties is nearly always a condition of postpubertal females, and thus, is only seen in older children and is usually encompassed by adult medicine. Hirsuties is described as the androgen-dependent growth of terminal hair on the body of a female in the same pattern and sequence as that of the postpubertal male. This description, however, is a little tenuous, as most girls develop hair in an identical pattern to boys at puberty, the only difference being that girls produce less body hair. Therefore, hirsuties relates more to the degree and quality of body hair growth. Recognizing the male androgen-dependent pattern is important because it allows hirsuties to be differentiated from generalized hypertrichosis.

Cultural norms and fashion define acceptable amounts of hair that girls are happy to accumulate on their face and body, so the first consideration in diagnosis is whether a pathological amount of hair exits. If a girl is not considered hirsute but simply has unwanted hair, advice can be given concerning cosmetic camouflage or removal of hair. When to investigate is the next consideration, and this is determined by the age and rate of onset of the hirsutism, the degree of hirsutism, and the presence of any other features of virilization such as androgenetic alopecia, male habitus, deepening of the voice, and clitoromegaly. Menstrual irregularity and acne are also signs of hyperandrogenism, but their high natural incidence in adolescence must be taken into consideration. Onset of hirsuties before puberty, rapid progression of hirsuties, and clitoromegaly are suggestive of an androgen-secreting tumor.

A minority of girls will require pharmacological treatment for hirsuties. This may be in the form of an oral contraceptive pill containing 2 mg cyproterone acetate with 25 μg ethinyl estradiol for 21 of the 28 days of the menstrual cycle or cyproterone acetate alone in higher doses; spironolactone can also be used. Many parents do not like the thought of their daughter's being placed on an oral contraceptive, so a careful explanation of the treatment at the outset may prevent problems later.

Hereditary and Congenital Hypotrichosis

Hypotrichosis is best divided into localized and generalized types. Excision of the lesion or hair transplantation may be a management option for localized hypotrichosis in an older child. The value of an accurate diagnosis in generalized cases is to provide some idea of the natural history of the condition, detect known associated anomalies, reassure the parents, and provide genetic counseling.

Localized Hypotrichosis

Aplasia Cutis Congenita

Aplasia cutis congenita is a congenital anomaly seen in 0.03% of live births and can be sporadic, of autosomal dominant inheritance, or the hallmark of a syndrome.[19] Eighty-five percent of cases are found on the scalp and may be shallow, involving only epidermis, or may extend down to bone. The most common site is over the parietal whorl. Healing in utero may occur, leaving only a scar devoid of hair at birth, or the lesion may be ulcerated. Rapid healing usually occurs in the postnatal period when the lesions are small, but if this is unobtainable, a skin graft may be required. Most lesions are singular and 1–2 cm in diameter. In approximately 20% of cases, associated developmental defects are found.

Dermal Hypoplasia

Focal dermal hypoplasia, also known as Goltz syndrome, is an X-linked dominant condition that is lethal in males. Females demonstrate lionization and present with streaks of dermal hypoplasia, hypopigmentation, telangectasia, and herniations of fat along Blaschko's lines. Leonine facies, absent eyebrows, puckered periorbital skin, and symmetrical scarlike defects on each temple (following Blaschko's lines) may be associated. The epidermis is intact and uninvolved over the affected regions.

Temporal Triangular Alopecia

This condition was first described by Sabouraud in 1905 and given the name *alopecia triangulare congenitale de la temp,* which has been translated into English as congenital triangular alopecia. Although this rare condition is most commonly seen in young children, case reports of onset in adult life encourage the use of the name *temporal triangular alopecia.*[20] The permanent hair loss or hypotrichosis is localized and usually occurs in the temporal region in infancy, compared with congenitally. There is equal sex incidence. Histology shows a normal epidermal and dermal architecture with a normal number of hair follicles, which are capable of producing only vellus hairs. The lesion is noninflammatory and scarring is absent.

Generalized Hypotrichosis

Generalized hypotrichosis may occur either as a single anomaly or may be just one characteristic of a vast number of syndromes that may have been described only once or twice. Discussion here is limited to generalized hypotrichosis, in which the absence of hair is the main clinical finding.

Reduction of scalp hair follicles is seen in these conditions to a varying degree and could be considered a continuum of the one disease. Hereditary hypotrichosis does not need to be present at birth, and follicular drop-out may ensue over the first 5 years of life. This is thought to be due to programed destruction rather than follicular agenesis. Conversely, hypotrichosis present at birth may not be noticed until 2 years of age owing to the wide variation in quantity and quality of scalp hair seen in normal babies at birth. Scarring may or may not be present.

Atrichia Congenita

Atrichia congenita is most commonly seen as an autosomal recessive condition, but it also exists in an autosomal dominant inheritance pattern and is the most severe form of generalized hypotrichosis. It is usually an isolated phenomenon, but facial papular cysts, immunodeficiency, epilepsy, mental retardation, deafness, ocular abnormalities, ichthyosis, skeletal abnormalities, inborn errors of metabolism, and premature aging syndromes have been associated. The baby may be born with a complete absence of hair, or the infant may progress to this over the first 5 years of life. A scalp biopsy can be performed to exclude alopecia areata totalis when there is no family history, as well as give information on the follicle count, state of the follicles, and the presence of other cutaneous abnormalities.

There are several variants of atrichia congenita. In one variety the neonate is born with lanugo hair on the scalp, which is shed in the first few months of life and never replaced. Madarosis is the congenital absence of eyebrows, which may occur with or without the absence of scalp hair.

Marie Unna Hereditary Hypotrichosis (MUHH)

First described by Dr. Marie Unna in 1925 in a German family over seven generations, this disorder is characterized by progressive alopecia in an autosomal dominant fashion.[21] The condition is exceedingly rare, and there have been fewer than 12 families of Caucasian origin reported. From about 1–2 years of age, hair becomes increasingly wiry and difficult to manage. This had been described as being similar to horse hair or that of a doll's wig. At or after puberty, hairs become increasingly lost from the scalp, proceeding most often to severe baldness. The pattern mimics that of severe androgenetic alopecia; however, eyebrows and eyelashes may also be affected. Histology reveals a nonscarring process leading to follicular drop-out with prominent sebaceous glands. Other abnormalities of the nails, teeth, and intellectual and gross motor development have not been described in association with this condition.[22]

MUHH is most likely a single-gene disorder due to a strong inheritance pattern in the families that have been reported. MUHH has been linked to a locus near the hairless gene on chromosome 8.[23] A mutation in the hairless gene has recently been discovered to cause congenital atrichia; however, no mutation has been found to date in the nearby loci to account for MUHH.[24]

Structural Abnormalities of the Hair Shaft

Numerous abnormalities of the hair shaft have been demonstrated on light microscopy, some due to an inherited disorder but others due to the normal weathering process. This in turn can produce varied appearances of the hair, including areas of alopecia, or frizzy, coarse, or limp hair. Hair may be described as unruly because it is twisted, curled, or irregularly shaped, all of which prevent the hair fibers from sitting together neatly. Spangled hair refers to hair that is either twisted or has alternating light and dark bands that reflect light in different directions. There are a number of recognized hair shaft disorders, each with distinctive features, the more common of which are described below.

Weathering

Weathering refers to the structural damage caused by external forces. It is caused by environmental factors such as wetting, ultraviolet radiation, and natural friction as well as damage done by cosmetic procedures such as combing and brushing. Weathering is more common in girls because they are more likely to grow their hair longer and style it.

The hair shaft consists of a hydrophobic lipid envelope that surrounds the proteinaceous cuticle, which in turn surrounds the cortex and central medulla. It is this

lipid exocuticle that gives hair its luster and silky texture and is the first to be destroyed by weathering, followed by the cuticle itself. This gives rise to limp, lusterless hair with a coarse texture. Later changes include cortical damage, terminal fraying, and longitudinal fissures between exposed cortical cells, followed by transverse fissures and nodes of the type seen in trichorrhexis nodosa as the weathering becomes more severe.[25] When trichorrhexis nodosa is seen in young children, the possibility of an underlying metabolic cause should be considered. Bleaching or perming hair in teenagers may distort the shaft, which in turn increases the likelihood and extent of frictional damage. Certain hair shaft disorders will also cause hair to weather to a greater degree than normal, such as the twists in pili torti or the internodes in monilethrix.

Treatment for weathering is prevention. This includes the avoidance of all unnecessary trauma and limitation of the use of hair styling products. Washing of the hair with conditioner is advised.

Uncombable Hair

This is an autosomal dominant genodermatosis characterized by triangular hair. The hair is first noted to be abnormal at about 3 years of age and is seen to be unruly and resistant to all forms of grooming with a brush or a comb. The pathognomonic feature on light and electron microscopy is that >50% of the hairs have a triangular or kidney-shaped cross section with a longitudinal groove along almost the entire length of the hair. The condition rarely improves with age.

Pili Torti

The hair shaft in pili torti is flattened and twisted through 180° at irregular intervals along the shaft. The hairs are fragile and snap off if subjected to moderate trauma. Not all hairs are affected, and the proportion of affected hairs varies from person to person. The twisting gives the hair a spangled appearance, and the fragility may lead to circumscribed areas of baldness at sites of friction and trauma. In severe cases, there may be only a short, coarse stubble over the entire scalp. In areas not subjected to trauma where the hair is allowed to grow long, the twisting renders it unruly.

Pili torti has a variable clinical presentation. Classically the hair is normal at birth and is gradually replaced by spangled, blond, abnormal hair between the third month and third year of life. The hair remains abnormal until puberty, when it darkens, becomes less fragile, and grows to an acceptable length.

An inherited mutation in the keratin 17 (*K17*) gene has been identified in the pili torti phenotype of the type II form of pachyonychia congenita.[26] K17 is an epithelial, hard keratin intermediate filament found in the medulla compartment of hair and the presumptive medulla precursor cells located in the hair follicle matrix. K17 is limited to facial and scalp hair, and this may explain the anatomic distribution of pili torti in the condition.

Menkes' kinky hair syndrome is an X-linked recessive condition resulting in an abnormality of copper metabolism. Affected boys have pili torti, growth retardation, and progressive psychomotor retardation. Females are carriers and display patchy areas of hair loss due to random X chromosome inactivation.

Twisting Hair Dystrophy

There are a number of heterogeneous disorders in which half and three-quarter twists of the hair are seen at irregular intervals rather than the full 180° twists seen in pili torti. Acquired twisting is also almost invariably seen at the edge of a scarring alopecia and is due to follicular distortion as a result of the scarring process.

Woolly Hair

This refers to tightly coiled hair over all or part of the scalp that resembles Negroid hair in an individual of another race. Generalized woolly hair can be inherited in either an autosomal dominant or recessive fashion. The hairs are fine and of irregular caliber, with occasional loose twists and of varying strength. The hair may relax from curly to wavy with age, and associations with enamel hypoplasia, ocular defects, deafness, ichthyosis vulgaris, keratosis pilaris atrophicans, and Noonan's syndrome have been reported.

Localized, nevoid woolly hair may be present at birth but appears not to be genetically determined. Acquired woolly hair also exists, but this appears to be a distinct condition that is usually a precursor of androgenetic alopecia.

Pili Annulati

Children rarely present with this autosomal dominant condition, and usually it is discovered incidentally. Sporadic cases have been reported. The child's scalp hair has a banded or slightly spangled appearance and is more noticeable in less pigmented hair.[27] As with other genetic disorders, severity of the phenotype is extremely variable, with some individuals displaying gross banding clinically, to cases in which light microscopy is necessary for the diagnosis. The light band is normal hair, whereas the dark band represents air cavities within and around cortical cells in the cortex of the hair shaft.[28] These bands are seen on transmitted-light microscopy, more easily seen on fixed hair specimens rather than air-mounted ones. Use of cross polarization aids detection of banding in subtle cases.

On scanning electron microscopy, the cuticle is intact but appears cobblestoned and fluted. This feature can be explained by the cuticular cells' being thrown into folds at the points where the underlying cortical cells

have collapsed because of multiple cavities within and around them. Transmission electron microscopy reveals a large number of abnormal holes, varying in shape and size, ranging from <1 μm to >10 μm in the cortical cells. The medulla is normal.

Monilethrix

This rare autosomal dominant condition has recently been shown to be due to mutations in the helix termination motif of two type II human hair keratin genes, *hHb1* and *hHb6*.[29] The mutations found predict possible structural abnormalities, including misalignment of keratin filaments and the fiber interaction that may result; however, no definite link between clinical features or severity has been shown between the different mutations. In particular, the mutations provide no explanation for the periodic nodes. There is a suggestion that *hHb1* may produce a milder phenotype and that nail dystrophy may be commoner with *hHb1* mutations.

Clinically the hair is characterized by keratosis pilaris, beaded hairs, and fragile hairs. The beading may produce a slightly spangled appearance. The dominant clinical feature is hair that fails to grow long owing to fragility, and both beaded and nonbeaded hairs are fragile. This condition has a high but variable penetrance, giving rise to a wide spectrum of presentations from hair that is nearly normal to hair that is not able to be grown >2 cm.

Trichorrhexis Invaginata

This condition is colloquially known as bamboo hair because of its appearance under the light microscope. It may occur alone, but when combined with ichthyosis and atopic diathesis, it known as Netherton's syndrome. Like monilethrix, the primary abnormality is one of keratinization of the hair fiber, which allows intussusception of the harder, keratinized distal segment of the hair fiber into the softer, proximal segment. This gives the appearance of bamboo hair, and when the distal segment breaks off the end of the hair fiber takes on a tulip appearance.

Loose Anagen Syndrome

Loose anagen syndrome is a newly described condition seen exclusively in children, and it predominantly affects girls.[30] Here, anchorage of the growing anagen hair to the follicle is impaired, and these hairs can be easily and painlessly plucked from the follicle. As a result, most hairs do not remain in situ until completion of the anagen phase. This results in affected children's having hair that does not grow long and that is of uneven length. The hair is unruly and has an unusual sticky feel to it. Bald patches are easily produced if a handful of hair is pulled in a childhood dispute and is often the trigger for presentation.

Children between the ages of 2 and 7 years are most commonly affected. Their hair appears normal at birth but becomes unruly, sticky, and uneven at 2–3 years and remains so until it spontaneously reverts to normal at 5–7 years. It is quite common for these children to have never had a haircut. It is inherited in a polygenic fashion, and it is common to find that apparently normal parents or older siblings have easily plucked hair. The characteristic feature seen on light microscopy is an anagen hair with a misshapen bulb, a crumpled proximal hair cuticle without an inner root sheath that has been extracted by gentle traction.

Trichotillomania

Trichotillomania refers to a patient's compulsion or tic to twist and pluck hairs. The scalp and eyebrows are the most common sites, but any area may be involved. It is twice as common in girls compared with boys. The clinical findings are unusual patterns of hair loss and a stubble of short hairs of irregular length, owing to the fact that it is difficult to manually pluck out hairs shorter than a few millimeters. Alopecia areata may be a differential diagnosis on clinical grounds, as these short hairs may be mistaken for "exclamation mark" hairs. One can differentiate between the two by plucking the short hairs: in alopecia areata, they will come out easily, whereas the anagen hairs of trichotillomania will require a tug.

Fractured hair fibers and evidence of perifollicular trauma may be seen on biopsy, but follicles devoid of their hair fiber surrounded by normal follicles may be all that is seen. Trichotillomania is mostly a nonpermanent alopecia, but if the same area of hair is continually plucked, scarring may result in permanent hair loss.

Infantile trichotillomania occurs in children who continually play with their hair and is a benign, self-limiting form. These children are often unaware that they are playing with their hair or are too young to understand and eventually grow out of this habit. If the child continually plays with the same area (usually an area easily accessible with the dominant hand), the hair fibers may fracture and produce a clinical picture similar to trichotillomania.

Adolescent trichotillomanics, on the other hand, often describe scalp dysesthesia or a tingling sensation in the hair they are targeting next. The children are aware of their actions but may often be secretive about their plucking, limiting it to times when they are alone or in bed at night. This can be difficult for the treating physician, as the child will deny such behavior and the parents refuse to believe the diagnosis. This can create great anxiety and discord in the family, with the parents seeking many medical opinions before they accept the diagnosis. An identifying cause for the behavior should be sought and counseling arranged. Psychiatric referral is recommended in recalcitrant cases.

Infections and Infestations of Hair

Tinea Capitis

Tinea capitis is the invasion of scalp hair by dermatophyte fungi, which can occur in one of three ways. Anthrophilic infections are acquired from close person-to-person contact as well as being passed on by the shared use of hairbrushes, combs, and hats. Zoophilic infections are acquired from animal contact and geophilic infections arise in children playing in infected soil. Some infections can be both zoophilic and geophilic, and *Microsporum canis*, which is classified as zoophilic, can also have limited passage from person to person.

M. audouinii, Trichophyton violaceum, and *T. schoenleinii* have a predilection to hair; however, nearly all dermatophytes are able to infect hair. The predominant organism causing tinea capitis varies from country to country: in Europe it is *M. canis*, whereas in the United States it is *T. tonsurans*.

Classification of scalp ringworm is based on the pattern of hair invasion. Endothrix (from within) infections are only due to *Trichophyton* species. Ectothrix (from outside) infections may be divided into small-spored (2–3 μm) types, which are only caused by *Microsporum* species, or large-spored (5–10 μm) types, which may be caused by either *Microsporum* or *Trichophyton* species.

Generally, anthrophilic fungi produce noninflammatory lesions, whereas zoophilic and geophilic fungi produce lesions with marked inflammation, such as kerion and favus. Inflammatory tinea capitis tends to be self-limiting over 6–10 weeks, but it generally heals with scarring. Infection is far more common in children than in adults.

Small-spore ectothrix infections classically produce annular lesions or gray-patch ringworm. Brittle hairs break off close to the scalp surface to create circular patches of partial alopecia. The broken hairs have a dull gray appearance owing to their coating of arthrospores. Inflammation of the scalp is minimal, but fine scaling is characteristic. There is usually a fairly sharp margin and there may be several such patches throughout the scalp. Each fluoresces green with Wood's lamp examination. With some infections due to *M. audouinii*, there may be minimal hair loss and the mild scaling may mimic seborrhoic dermatitis.

Large-spored ectothrix infections may produce scalp ringworm that resembles small-spored ectothrix. Alternatively, there may be more marked inflammation of the scalp. Occasionally a folliculitis or kerion occurs, especially with zoophilic and geophilic fungi. Agminate folliculitis is a moderately severe inflammatory response and consists of well-defined, dull red plaques studded with follicular pustules. A kerion is a painful, boggy, elevated, purulent inflammatory mass. Hairs fall out rather than break off, and any remaining hairs can be easily and painlessly pulled out. Thick crusting with matting of adjacent hairs is common. The usual organisms responsible are *T. verrucosum* and *T. mentagrophytes*. Occasionally, an insidious anthrophilic infection will suddenly develop into a kerion.

In endothrix infections, the arthrospores remain confined within the cuticle, and affected hairs are severely damaged and break off at the scalp surface. Children present with black dot ringworm, where broken hairs within an angular patch of alopecia appear as black dots. With the anthrophilic fungi, there is usually minimal scaling or inflammation of the scalp. Sometimes hair loss is minima, and all that can be seen is a mild folliculitis or seborrhoic dermatitis–like scaling.

Majocchi's granuloma and tinea incognito can result from treatment of tinea capitis with potent topical steroids. Typically with tinea incognito, the raised margin is lost, scaling and itch are absent, and the inflammation is reduced to a few nondescript nodules. Majocchi's granuloma on the scalp is rare and presents as a granulomatous folliculitis with inflammatory nodules bordering flat, scaly patches.

Yeast infection of the scalp with *Candida* is uncommon and generally occurs only in the setting of immunodeficiency.

The Wood's lamp is an invaluable tool for diagnosing fungal conditions. Green fluorescence is seen when a fluorescent substance, possibly pteridines, is formed when hair keratin is invaded. Not all dermatophytes cause fluorescence, and scalp lotions and creams can mask fluorescence. These should be washed off before examination with the Wood's lamp. Fluorescence may confirm fungal etiology, give an indication to the species involved, and define the extent of the infection. Six to eight hairs should be plucked for fungal microscopy and culture to first, confirm the diagnosis and then to identify the species involved.

The standard treatment for all ringworm of the scalp is still griseofulvin. Given late in inflammatory infections (which have a tendency to spontaneously heal), it may not alter the course of existing lesions, but it will prevent the development of new ones. Gray-patch ringworm requires the full dose, which is 10 mg/kg daily of microsize griseofulvin for children for at least 6 weeks. Cultures should be repeated after 4 weeks and every 2 weeks thereafter until mycological cure. Black dot ringworm may require slightly longer treatment and should be continued for 2 weeks beyond clinical and mycological cure. Kerion generally requires 6 weeks of therapy.

Headache from the drug may be overcome by reducing the dose and slowly increasing it again. An "id" eruption, consisting of multiple, small papules on the side of the face and the trunk or vesicles on the hands, may occur after commencing therapy with griseofulvin for tinea capitis; however, a true drug hypersensitivity to griseofulvin is rare. Secondary bacterial infection

occasionally occurs, and if suspected, swabs should be taken and a broad-spectrum oral antibiotic such as erythromycin given.

Alternatives to griseofulvin are ketoconazole, itraconazole, fluconazole, and terbinafine. Cure rates are similar, shorter treatments are required, and there are fewer drug interactions. The drawbacks are that ketoconazole has problems with hepatotoxicity (1 in 10,000), and many of these newer drugs are not licensed for use in children.

The source of infection should be traced. With *M. canis,* cats and dogs should be examined with a Wood's light by a veterinary surgeon and infected animals treated with griseofulvin.

Griseofulvin treatment of kerion does not tend to prevent the development of scarring alopecia, so it is prudent to combine the griseofulvin with prednisolone 0.5 mg/kg body weight daily for the first week or 2, and this may also hasten resolution of the symptoms. Oral antibiotics are generally not required unless secondary bacterial infection is proven. Selenium sulfide or ketoconazole shampoo may also decrease the period of fungal shedding. In very young children unable to swallow tablets, the griseofulvin can be crushed and mixed with ice cream, or a griseofulvin suspension can be used.

Children with infection due to anthropophilic fungi and *M. canis* should be kept at home from school until treatment has been commenced. In school epidemics, classmates should be examined with a Wood's light; however, in recent infections the fluorescent part of the hair may not yet have emerged from the follicle, and fluorescence will only be detected if the hair is plucked and the root examined.

Pediculosis

Lice feed on the skin and deposit their eggs (nits) on the hair. Two species of louse infest humans, *Pediculosis humanis* and *Phthirus pubis*. There are two subspecies of *P. humanis*: *P. humanis capitis*, the head louse, and *P. humanis corporis*, the body or clothing louse. Head lice and body lice look almost identical and are capable of interbreeding, but on the host they tend to maintain their territorial preferences. Although the head louse occasionally wanders onto the body, the body louse rarely ventures onto the scalp.

The female head louse is 3–4 mm long. The male is slightly smaller and banded across the back. During her 40-day life span, the female lays approximately 300 eggs at a rate of eight per day. The eggs are oval, white capsules with a lid (operculum) and are firmly cemented to the side of the hair shaft adjacent to the scalp. After about a week, the larvae hatch close to the scalp. Larvae resemble small adults and begin feeding on the blood of the host soon after hatching. After undergoing three molts in 10 days, the louse reaches maturity and commences mating.

In most established infestations of head lice, there are <10 adult lice and counts of >100 are uncommon. Most infections are acquired by direct head-to-head contact, but combs, brushes, and hats are important in some cases. Pruritus is variable. It is often intense and only rarely absent. It is usually worst in the occipital region, where the infestation is heaviest. Scratching leads to impetiginization, and the hair may become matted down by exudate to produce "plica polonica." Nits can be seen with the naked eye and are very easily seen with a Wood's light. This is useful for screening in schools during epidemics. Head lice are rare in the Negroid race, as the lice appear not to clasp tightly curled hair as well.

The different phases of the skin's reaction to infestation has been described:[31]

Phase 1: no clinical symptoms

Phase 2: pruritic papules

Phase 3: immediate wheal formation after bites, followed by an intensely itchy delayed papular eruption

Phase 4: papular reaction with diminished skin reactivity and mild pruritus

Difficulties with treatment are threefold. There is now biological resistance to most of the chemical insecticides,[32,33] there are practical and social problems with the application of physical therapies, and in schoolchildren or in cases where untreated, infested family members are present, reinfestation poses a real problem.

In the presence of severe itch, scratching may cause secondary bacterial infection, for which systemic antibiotics are usually required.

Malathion and carbaryl became the mainstays of therapy for head louse infection after emergence of resistance to the organochloride antiseptics. They were required to be left in the scalp for 12 hours before being washed off. Both agents effectively kill lice; however, they do not kill all of the eggs and a second application after 7–10 days is usually recommended. Malathion coats the hair, making it resistant to reinfection for 6 weeks. Increasing resistance prompted the use of new agents, and now permethrin is the most widely used agent. This generally only requires a 10-minute scalp application after shampooing. Blow drying should be avoided, as heat degrades these insecticides.

Multiple resistance has now become a problem, with one report of resistance to permethrin measured at 87%, with a coincident resistance to malathion of 64%.[34] In vitro testing of the same population of lice showed that carbaryl was the most effective agent in this case, reducing the live louse count to $<5\%$.

Ivermectin as an oral agent has been shown to be

effective,[35] but its use has been limited owing to reports of increased susceptibility to neurological problems in children.[36] Topical use of ivermectin may prove successful in the future, but once again resistance is likely to become an issue.

None of these treatments remove the dead nits. Combing with a fine-toothed comb is tedious and painful. Nits will eventually wear away after repeated washing, but 8% formic acid in a cream rinse can be used as a nit remover.

Head lice epidemics are common, and in general, malathion, carbaryl, and permethrin are rotated periodically by local health authorities to prevent resistance. Infected children should be kept at home from school until the first treatment is completed, and in some countries, this is a statutory requirement aimed at preventing epidemics. There is no recent evidence, however, that exclusion is effective in controlling head lice in school children.[37]

Conclusions

A significant part of managing pediatric hair conditions is being able to educate the patient and parents about the etiology of the condition, the natural history, what treatment options are available, how successful they are, and what constitutes a realistic end point. Knowledge of basic hair biology not only allows a description of normal but also enhances the assessment of the child with a pathological condition. Each patient should be assessed in a systematic way, first deciding whether the disorder lies with hair volume or the hair shaft. Infective causes are important to recognize, as they are amenable to treatment. A hair deficit that may be an isolated feature or the only visible sign of an associated multisystem disorder, and therefore, it is important to recognize, as it will direct the clinician to the appropriate investigations to unmask more serious underlying or associated deficits. Sometimes genetic counseling or a clear description of the condition is all that can be offered to the patient and family, but even this is of great help and will allay anxiety or many unfounded concerns. Keeping up to date with therapeutic options in this rapidly progressing field is vital.

References

1. Bertolino AP, Klein LM, Freedberg IM. Biology of hair follicles. In: Fitzpatrick TB, Eisen AZ, Wollf K, Freedberg IM, Austen KF, eds. Dermatology in general medicine. 4th ed. Boston: McGraw-Hill, 1993:291.
2. Dawber RP, de Berker D, Wojnarowska F. Disorders of hair. In: Champion RH, Burton JL, Burns DA, Breathnach SM, eds. Rook/Wilkinson/Ebling textbook of dermatology. 6th ed. Oxford: Blackwell Science, 1998:2885.
3. Reynolds EL. The appearance of adult patterns of body hair in man. Ann NY Acad Sci 1951;53:576–84.
4. Rafaat M. Hypertrichosis pinnae in babies of diabetic mothers. Pediatrics 1981;68:745–6.
5. Oliver GL, McFarlane DC. Congenital trichomegaly with associated pigmentary degeneration of the retina, dwarfism, and mental retardation. Arch Ophthalmol 1965;74: 169–74.
6. Coleman R, Harper JI. Hairy elbows syndrome (familial hypertrichosis cubiti). Clin Exp Dermatol 1994;19:86.
7. Harris HW, Miller OF. Midline cutaneous and spinal defects. midline cutaneous abnormalities associated with occult spinal disorders. Arch Dermatol 1976;112:1724–8.
8. Brownstein MH, Helwig EB. Subcutaneous dermoid cysts. Arch Dermatol 1973;107:137–9.
9. Sinclair RD, Darley C, Dawber RP. Congenital inclusion dermoid cysts of the scalp. Australas J Dermatol 1992;33: 135–40.
10. Ruge JR, Tadanori T, Naidich TP, et al. Scalp and cranial masses of infants and children. Neurosurgery 1988;22: 1037–42.
11. Conley FK. Epidermoid and dermoid tumors: clinical features and surgical management. In: Wilkins RH, Rengachary SS, eds. Neurosurgery. New York: McGraw-Hill, 1985:668–73.
12. Baxter JW, Netsky MG. Epidermoid and dermoid tumors: pathology. In: Wilkins RH, Rengachary SS, eds. Neurosurgery. New York: McGraw-Hill, 1985:655–61.
13. Tymen R, Forestier J-F, Boutet B, Colomb D. Naevus tardif de Becker. Ann Dermatol Venereol 1981;108:41–6.
14. Schauder S, Hanefeld F, Noske UM, Zoll B. Depigmented hypertrichosis following Blaschko's lines associated with cerebral and ocular malformations: a new neurocutaneous, autosomal lethal gene syndrome from the group of epidermal naevus syndromes? Br J Dermatol 2000;142: 1204–7.
15. Hamm H. Cutaneous mosaicism of lethal mutations. Am J Med Genet 1999;85:342–5.
16. Felgenhauer W-R. Hypertrichosis lanuginosa universalis. J Genet Hum 1969;17:1–44.
17. Burton JL, Marshall A. Hypertrichosis due to minoxidil. Br J Dermatol 1979;101:593–5.
18. Bencini PL, Montagnino G, Sala F, et al. Cutaneous lesions in 67 cyclosporin-treated renal transplant recipients. Dermatologica 1986;172:24–30.
19. Van Dijke CP, Heydendael RJ, de Kleine MJ. Methimazole, carbimazole and congenital skin defects. Ann Intern Med 1987;106:60–1.
20. Trakimas CA, Sperling LC. Temporal triangular alopecia acquired in adulthood. J Am Acad Dermatol 1999;40:842–4.
21. Sinclair RD, Banfield CC, Dawber RP. Hereditary alopecias and hypotrichosis. In: Handbook of diseases of the hair and scalp. Oxford: Blackwell Science, 1999:139–40.
22. Roberts JL, Whiting DA, Henry D, Basler G, Woolf L. Marie Unna congenital hypotrichosis: clinical description, histopathology, scanning electron microscopy of a previously unreported large pedigree. J Invest Dermatol 1999; 4:261–7.
23. Roberts JL, Wong CQ, Stenn KS. Marie Unna hereditary hypotrichosis gene maps to human chromosome 8p21 near. Hairless J Invest Dermatol 2000;114:595–7.
24. van Steensel M, Smith FJ, Steijlen PM, et al. The gene for hypotrichosis of Marie Unna maps between D8S258 and

D8S298: exclusion of the hr gene by cDNA and genomic sequencing. Am J Hum Genet 1999;65:413–9.
25. Jones LN. Investigation of structural proteins in human hair defects using anagen follicles. Br J Dermatol 1996;135: 80–5.
26. McGowan KM, Coulombe PA. Keratin 17 expression in the hard epithelial context of the hair and nail, and its relevance for the pachyonychia congenita phenotype. J Invest Dermatol 2000;114:1101–7.
27. de Berker D, Sinclair RD. Defects of the hair shaft. In: Dawber RP, ed. Diseases of the hair and scalp. 3rd ed. Oxford: Blackwell Science, 1997:286.
28. Price VH, Thomas RS, Jones FT. Pili annulati: optical and electron microscopic studies. Arch Dermatol 1968;98:640–7.
29. Korge BP, Hamm H, Jury CS, et al. Identification of novel mutations in basic hair keratins hHb1 and hHb6 in monilethrix: implications for protein structure and clinical phenotype. J Invest Dermatol 1999;113:607–12.
30. Hamm H, Traupe H. Loose anagen hair of childhood: the phenomenon of easily pluckable hair. J Am Acad Dermatol 1989;20:242–8.
31. Mumcuoglu KY, Klaus S, Kafka D, et al. Clinical observations related to head lice infestation. J Am Acad Dermatol 1991;25:248–51.
32. Goldsmith JM. Head louse treatment: is there an insecticide resistance problem? Med J Aust 1990;153:233–4.
33. Picollo MI, Vassena CV, Casadio AA, et al. Laboratory studies of susceptibility and resistance to insecticides in pediculus capitus. J Med Entomol 1998;35:814–7.
34. Downs AM, Stafford KA, Harvey I, Coles GC. Evidence for double resistance of permethrin and malathion in head lice. Br J Dermatol 1999;141:508–11.
35. Burkhart CG, Burkhart CN, Burkhart KM. An assessment of topical and oral prescription and over the counter treatments for headlice. J Am Acad Dermatol 1998;38: 979–82.
36. Burkhart CN, Burkhart CG. Another look at ivermectin in the treatment of scabies and head lice. Int J Dermatol 1999;38:235.
37. De Berker D, Sinclair RS. Getting ahead of head lice. Austral J Dermatol 2000;41:209–12.

The Liberty nickel was introduced in 1883 replacing the Shield Nickel. Originally the Liberty nickel did not have "five cents" written anywhere on the coin. Unscrupulous people gold plated the nickels and passed the coins as five dollar gold pieces, since they were about the same size. A particular con man named Josh Tatum was especially well known for passing gold plated nickels. He was caught tried and found not guilty because he did not ask for change and no laws were on the books to prevent gold plating coins. This is how the term "Just Joshing" came about. Later in 1883, "cents" was added in the obverse of the coin and laws were passed to prevent gold plating coins to prevent this from reoccurring.

The third most expensive coin in United States history is the 1913 version of the Liberty nickel, it sold for $1.485 million in 1996. The Liberty nickel was officially produced between 1883 to 1912. In 1919 Samuel Brown, a collector, speculator of coins and former employee of the mint began to advertise for 1913 Liberty Nickels. Mysteriously in the 1920's five examples of 1913 Liberty nickels turned up in the collection of one individual, Samuel Brown's.[2,3] It was hypothesized that the 1913 examples were clandestinely coined at the mint by an insider and sold to Mr. Brown.

From the collection of Raymond T. Kuwahara, MD, Memphis, TN.

REFERENCES:

1. Yeoman RS. A guide book of United States coins. 53rd ed. New York, NY: Whitman, 2000:8,9,172.
2. Mishler C. Coins Questions and answers. 4th ed. New York, Golden Books, 1998:57-8.
3. www.pngdealers.com