

Hair and scalp disorders.

The serial Book of Pediatric Dermatology is finished. Now we review Pediatric Dermatology from another side. The latter will be more practical for those physicians, who are not yet skilled in pediatric dermatology. In the previous book the thread consisted of the primary skin lesions. Now the thread will be the site of the lesions, even with different references according to the age.

The new handling is aimed at the differential diagnosis of dermatological disorders. Regarding the other topics, from etiology to treatment, we will refer to the previous book, only talking about what is new and about peculiar problems of the involved site.

The site of cutaneous lesions is important due to two apparently contrasting reasons as follows:

1- some skin disorders have a preference for particular sites. This preference, together with history and epidemiological data, makes the diagnosis easier even for physicians, who do not know very well the dermatological alphabet;

2- many sites, especially the scalp, oral cavity, nail, folds, regions with thicker horny layer such as palmar-plantar regions and extensor aspect of elbows and knees, have peculiar anatomic characteristics. The latter obscure the differences between lesions of different nature, making their differential diagnosis much more difficult. The scalp is the first example of these peculiar skin sites.

Anatomic and dermatological characteristics of the scalp. From an anatomic point of view the scalp is characterized by well developed pilo-sebaceous follicles with terminal hairs. Moreover, the skin of the scalp is scarcely extensible and more adherent to the underlying structures. From a dermatological point of view, due to the presence of hair, all the lesions affecting the scalp, mainly erythematous and dyschro-

mic flat lesions, are less visible as compared with those ones affecting the so-called glabrous skin, namely the skin covered by thin and barely visible hairs.

Moreover, sebum, which is here more abundant than in other sites, conglutinates the horny cells -the latter are usually continuously eliminated by an invisible process in other sites- and makes the cells visible, usually as dandruff.

From a practical point of view, a moderate erythematous inflammation well visible in the glabrous skin, level with the scalp is scarcely visible or manifests itself only with scaling. The latter is more evident on the scalp as compared with the glabrous skin. In conclusion, when a skin disorder affects both the scalp and the glabrous skin, it is less hazardous to do the diagnosis on the glabrous skin, choosing among the skin disorders more frequently affecting the scalp.

Hair in the neonatal period

Characteristics of the hair in the neonatal period. The hair has a growing phase (anagen) lasting about three years. In this phase the deepest portion of the hair follicle embraces the dermal papilla, establishing with the latter a close relationship. Later on, the hair enter the telogen phase through an intermediate phase (catagen). During the latter, the keratinocytes of its matrix do not enter the mitotic cycle and undergo apoptosis. Its bulb goes up towards the surface, losing the contact with the dermal papilla, around which another hair starts its growing phase.

In the adult about 10-15% of hair are normally in telogen and this percentage is randomly distributed throughout the scalp. On the other hand, in the fetus and in the first months of life



Fig. 1168: Physiological band-like hair loss of the nape of the neck in a 3-month-old baby.

the hair enter the telogen phase in a synchronic way with waves starting from the forehead and going towards the occipital region. The latter phenomenon is almost never clinically evident. The only clinical feature reminiscent of the synchronic wave is the loss of hair occurring between the 2nd and 4th month of post-natal life in the nape of the neck (Fig. 1168). This phenomenon, which is popularly attributed to the supine decubitus, can be detected also in babies not sleeping that way and can be observed only in this period of life, although the child sleeps in the same way even in the subsequent periods of time. Sometimes, waves of hair loss can be observed even in other sites besides the nape of the neck.

Moreover, the discussion is open about the time when the synchronic hair loss subsides. Moreover, this theory cannot explain the diffuse hair loss occurring around the 3rd-4th month of life. This physiological entity is the most frequent form of telogen effluvium (see later). It is not due to the trauma of delivery, because its frequency did not decrease in the towns, where the percentage of cesarean section is over 50% of deliveries.

Newborn devoid of hair. The less rare causes of diffuse rarefaction of hair at birth are keratosis follicularis spinulosa decalvans, ectodermal dysplasia and monilethrix. *Keratosis fol-*

licularis spinulosa decalvans (Fig. 1169) is characterized by generalized follicular keratosis with severe rarefaction of the hair.

There are numerous forms of *ectodermal dysplasia*, essentially the hypohidrotic form (Fig. 1170-4), which is transmitted by a recessive trait, X-linked or autosomal, and the hidrotic form, which is transmitted by an autosomal dominant trait. In the hypohidrotic form there can be hyperthermic crises during summertime or even in the incubator, tooth (Fig. 1172) and nail abnormalities and gross face.

In *monilethrix*, which is transmitted by an autosomal dominant trait, the newborn may present with a more or less severe hair rarefaction, but usually more marked on the nape of the neck (Fig. 1174). In an adult relative, a rarefaction of the hair can be detected only in this region. Monilethrix is associated to follicular keratosis, which is mainly visible on the nape of the neck (Fig. 1175). On light microscopy, the characteristic alternation of nodes (swelling) and internodes (thinning) of the hairs (Fig. 1176).

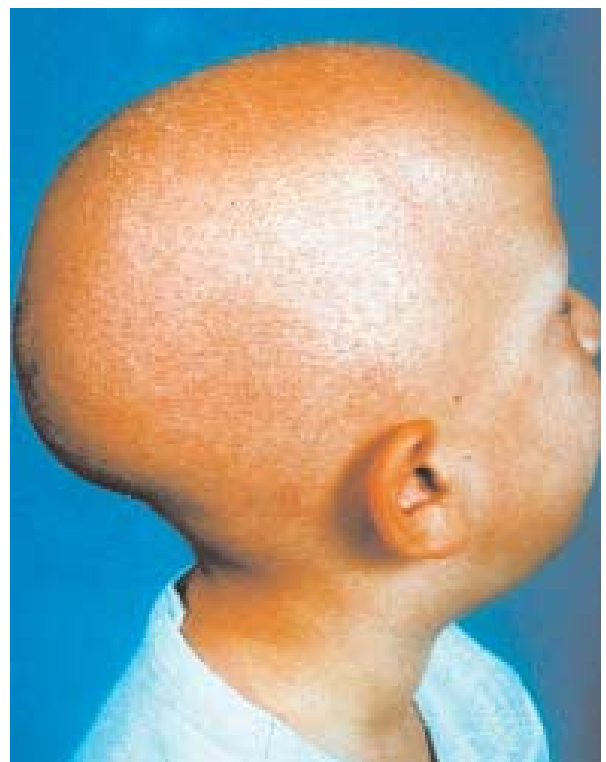


Fig. 1169: Keratosis follicularis spinulosa decalvans with severe hypotrichosis and diffuse follicular keratosis.



Fig. 1170



Fig. 1171



Fig. 1172



Fig. 1173

Fig. 1170, 1171, 1172, 1173: Gross face (Fig. 1170), with severe hypotrichosis of the hair, eyebrows and eyelashes when aged 40 days (Fig. 1170), 2 years (Fig. 1171) and 8 years (Fig. 1172). You can notice (Fig. 1172) the lack of 3 incisors. You can also notice atopic dermatitis of the face (Fig. 1171), of the perioral region and hand (Fig. 1172). On scanning electron microscopy (260x) the hairs are flat with a longitudinal groove ("pili canaliculi").



Fig. 1174

Fig. 1174, 1175, 1176: Monilethrix in two siblings with prevalent involvement of the nape of the neck. Follicular keratosis is clearly evident in the elder sister (Fig. 1175) on the nape of the neck. Fresh examination of the hair on light microscopy (Fig. 1176, 40x) shows the characteristic alternated nodes (arrows) and internodes (asterisks) of the hair, with a shape reminiscent of a jewel (aplasia moniliforme).



Fig. 1175

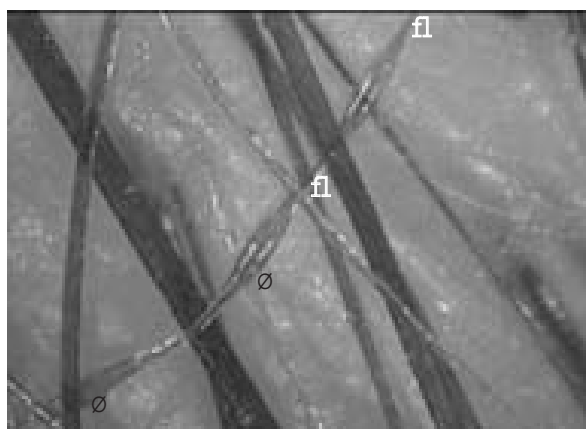


Fig. 1176

An **area devoid of hairs in the newborn** can be observed in aplasia cutis, sebaceous nevus and congenital triangular alopecia.

Aplasia cutis is present at birth (see p. 577 of Book, EJPD, vol.12, n. 3) as an ulcer which rapidly repairs with scarring or since birth with a scar, in both cases devoid of hairs.

Sebaceous nevus (Fig. 1179) presents with a barely infiltrated plaque, pink-yellow or chamois in color, with a finely granular or even warty surface (see p. 526 of the Book, EJPD, vol. 11, n. 2)

Congenital triangular alopecia (Fig. 1177) can be evident at birth or get visible later (see p. 586 of the Book, EJPD, vol.12, n. 3). It is characterized by an area of absolutely normal skin,

but devoid of hair or with very fine and short hairs.

A **tuft of thicker and darker hairs** can be seen in congenital melanocytic nevus (see p. 529 of the Book, EJPD, vol. 11, 2001). In fact, this tuft is usually the only sign of melanocytic nevus (Fig. 1182), which gets visible only after trichotomy.

The hair collar sign (Fig. 1183) consists of a ring of thicker and darker hairs surrounding a plaque of infiltrated and soft skin (see p. 580 of the Book, EJPD, vol.12, 2002). This sign is a clue to the diagnosis of aplasia cutis with an underlying closure defect. The latter can consist of a defect of the underlying bone with heterotopy of brain and sometimes communication

Fig. 1177, 1178, 1179: An area devoid of hair in the newborn period is more frequently due to sebaceous nevus (Fig. 1179), aplasia cutis (1178) or congenital triangular alopecia (Fig. 1177). The superficial appearance of the skin is a clue to the diagnosis. In congenital triangular alopecia (Fig. 1177) the skin is normal, in aplasia cutis (Fig. 1178) the skin is scarred or ulcerated and in sebaceous nevus (Fig. 1179) it is granular or velvety.



Fig. 1177



Fig. 1178



Fig. 1179



Fig. 1180



Fig. 1181

Fig. 1180, 1181, 1182, 1183: At birth congenital melanocytic nevus of the scalp is characterized by a very variable visibility, ranging from evident lesions (Fig. 1180) to lesions partially obscured by hypertrophic hairs (Fig. 1181) till to forms unveiled only by a tuft of darker and thicker hairs (Fig. 1182). However, a tuft of darker and thicker hairs (Fig. 1183) may be also expression of incomplete closure of the skin, bone and meningeal structures, as in deep aplasia cutis (hair collar sign).



Fig. 1182



Fig. 1183

with the encephalon. MR imaging is indicated when this sign is present and even in case of dermal sinus, manifesting itself with exsudation of liquor coming from a punctiform orifice, usually on the vertex.

A **tuft of white hair** can be seen in different disorders such as piebaldism (Fig. 1185), more rarely in hypopigmented nevus (Fig. 1184, see EJPD, vol. 10, 145-54, 2000) or in congenital

melanocytic nevus. *Piebaldism* or partial albinism is usually transmitted by an autosomal dominant trait and is associated to achromic areas of the skin, with isles of pigmented skin inside the white areas.

More rarely, poliosis is due to *hypopigmented nevus* (Fig. 1184) with segmental distribution or to precocious regressive phenomena inside a *congenital melanocytic nevus*.

Fig. 1184, 1185, 1186: A tuft of white hairs in the first months of life can be due to hypopigmented nevus (Fig. 1184, arrows), to piebaldism (Fig. 1185) -in the latter case it is associated to white patches of the glabrous skin and often to a positive family history- or to congenital melanocytic nevus with early regressive phenomena (Fig. 1186, arrows).

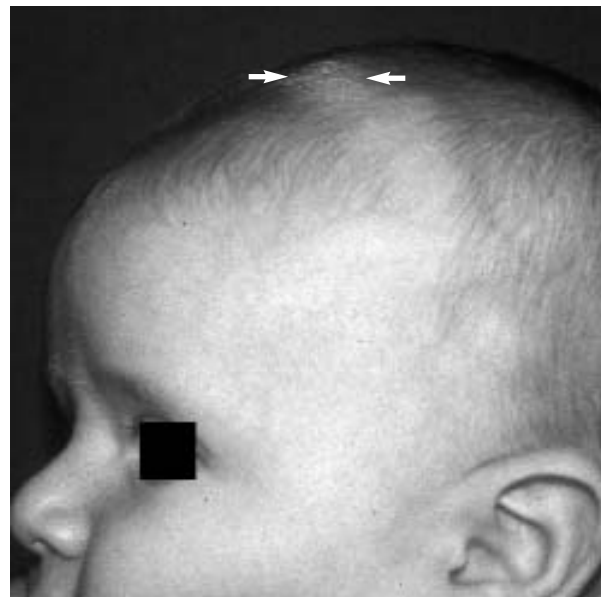


Fig. 1184



Fig. 1185



Fig. 1186

2-6-month-old children

Hair loss at this age can be due to congenitale causes such as acrodermatitis enteropathica or, more frequently, to atopic dermatitis.

Acrodermatitis enteropathica (see EJPD, vol. 2, 13-23, 1992) is due to a zinc deficiency. It can be congenital (Fig. 1187) and transmitted by an autosomal recessive trait -in this case it is due to the lack of a pancreatic ligand of zinc- or acquired and transitory, mainly in the premature with low gestational age (Fig. 1188). In both cases the hair loss occurs at the beginning of the third month, in the full term baby simultaneously with the withdrawal of breastfeeding -the maternal milk contains a zinc ligand similar to the pancreatic one- and in the premature with low gestational age, due to the imbalance between the scarce zinc supply -zinc as other oligomine- rals is transferred to the fetus in the last three months of pregnancy- and the increased request due to the role played by zinc in many enzymes of the proteic synthesis, which is very active in the premature.

Fig. 1187, 1188: Acrodermatitis enteropathica -congenital (Fig. 1187) or acquired in the premature with low gestational age (Fig. 1188)- associates burn-like, periorificial and acrolocated lesions, diarrhea and hair loss.



Fig. 1187

In acrodermatitis enteropathica the hairs under polarized light present a typical swan neck (2) appearance.

Atopic dermatitis is the most frequent disorder of the scalp at this age and should be differentiated from seborrheic dermatitis (p. 243 of the Book, EJPD, vol. 6, 1996).

The latter, which is often erroneously called "cradle cup", is a not well defined entity, because there are not characteristic clinical features or laboratory findings. We are dealing with a modest dermatitis of the scalp, more frequent in the second-third month, asymptomatic, not exudative and mainly affecting the hairline (Fig. 1189) and the glabellar region.

When the lesions are diffuse or exudating or symptomatic or associated with scaling in other sites, the diagnosis of atopic dermatitis is much more probable. In atopic dermatitis the scalp is usually affected for 3-6 months and in the most severe cases makes the physiological hair loss more evident.

The treatment is based on the manual removal of the scales and crusts. The latter is not much



Fig. 1188

appreciated by the child and consequently by the mother.

When after manual removal of the crusts exudating lesions are present, manifesting themselves as shiny or even humid skin, or intensely erythematous lesions heralding imminent exuda-

tion appear, an anti-inflammatory, corticosteroid cream should be applied. Alcoholic corticosteroid lotions should be avoided, because unnecessary, given the scarce hair at this age, and responsible for burning sensation and non-compliance by the children.



Fig. 1189



Fig. 1190

Fig. 1189, 1190, 1191, 1192: Between the 2nd and 6th month atopic dermatitis (AD) can affect the scalp, deteriorating the physiological hair loss. AD is characterized by diffuse, also in other sites, lesions (Fig. 1191, 1192), sometimes exudative and symptomatic. The elder sister of the infant as in Fig. 1190 with AD of the scalp had presented the same lesions in the first months. Seborrheic dermatitis (Fig. 1189) is asymptomatic and only slightly scaling.

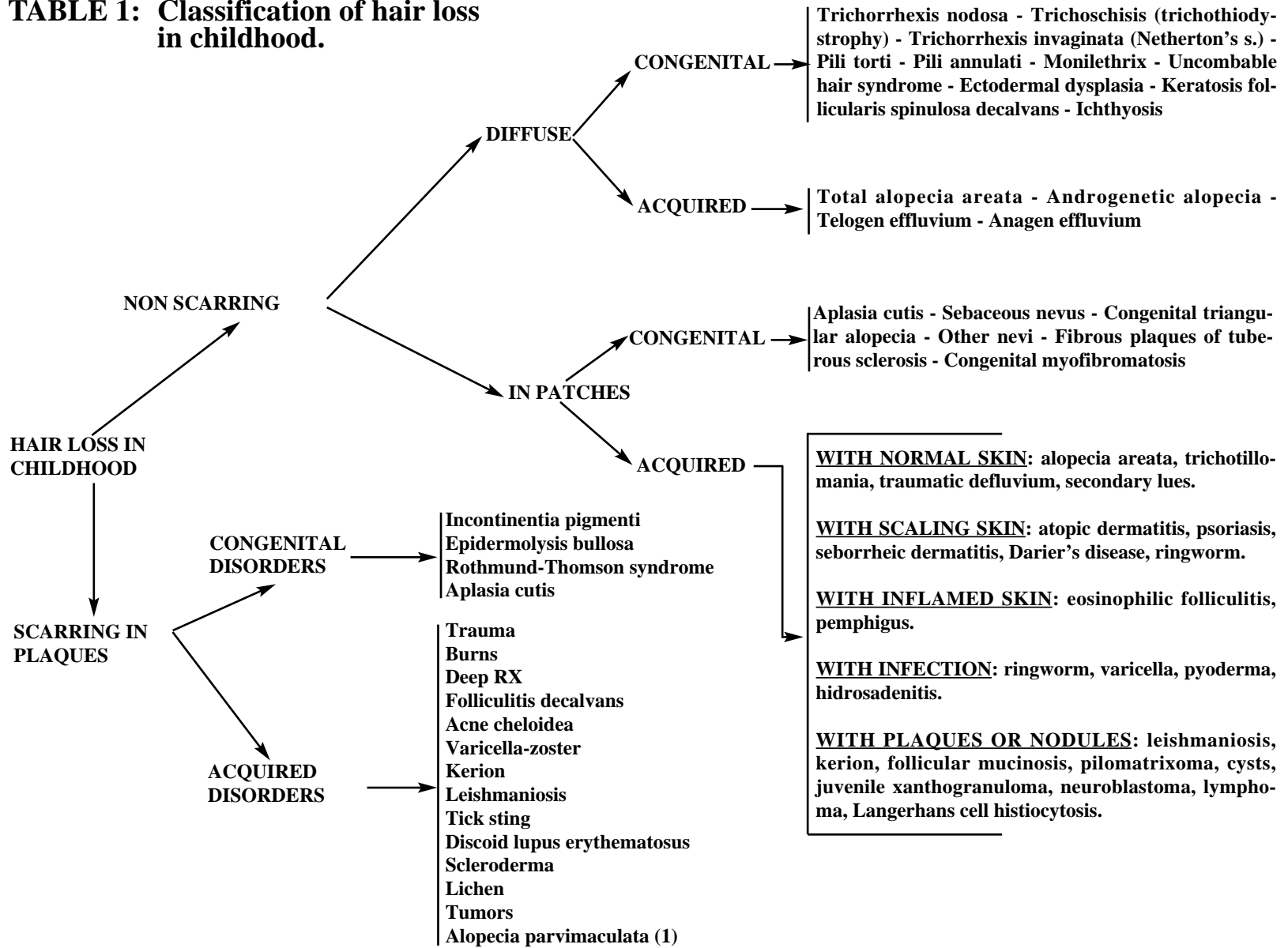


Fig. 1191



Fig. 1192

TABLE 1: Classification of hair loss in childhood.



All the pediatric age

The lack of hair can be grossly divided into scarring and non scarring, diffuse and localized, congenital and acquired. In scarring alopecia the hair loss is irreversible, the skin is altered due to the actual inflammation or scarring. Inside the plaque there can be isolated hairs, irregularly distributed. In case of scar, the follicular orifices cannot be detected.

Non scarring, diffuse, congenital hair loss. The most frequent forms have been already mentioned in the newborn. Physicians should also remember that some acquired forms can start very early in the neonatal period. On the other hand, although sometimes present at birth, the disorders of this group can get evident at the end of the first year or even later on with short and brittle hair.

The disorders of this group can be sporadic or genetically transmitted. In this group there are forms with follicular keratosis and forms lacking such alteration.

Among the forms with follicular keratosis, we should mention *keratosis follicularis spinulosa decalvans*, which can be associated also to inflammatory and scarring lesions, follicular ichthyosis, KID syndrome (keratitis, erythroder-

mic ichthyosis, deafness) and pachionichia congenital.

Monilethrix is included in the group of disorders with dysplasia of the hair shaft, although being associated with follicular keratosis.

In this group are included the disorders with microscopic and ultramicroscopic alterations of the hair shaft and clinically by short, brittle and irregular hair such as Menkes syndrome, trichothiodystrophy, Netherton's syndrome, pili torti, monilethrix, uncombable hair syndrome and woolly hair nevus.

Trichorrhexis nodosa is microscopically characterized by the presence of nodes. Level with the node the hair breaks, with fan-shaped frayed cortical fibres. It can occur as an isolated defect or associated to metabolic disorders such as argininosuccinic aciduria and citrullinemia. This alteration is also present in *Menkes syndrome*, due to a defect of copper metabolism.

Trichoschisis, namely a fracture of the hair independent from the presence of nodes, is characteristic of *trichothiodystrophy* (Fig. 1193), an autosomal recessive disorder, due to altered metabolism of sulfur. On polarized light the hair shows light and dark alternated bands (Fig. 1194), which are probably expression of alternating sulfur content.



Fig. 1193



Fig. 1194

Fig. 1193, 1194: Trichothiodystrophy has short and sparse hair (Fig. 1193) with alternated bands on polarized light (Fig. 1194).

Trichorrhexis invaginata or bamboo hair is due to invagination of the fully keratinized distal shaft into the incompletely keratinized proximal portion of the shaft, with consequent fracture level with the invagination.

This abnormality of the hair shaft is characteristic of *Netherton's syndrome*, an autosomal recessive disorder associating atopy, linear ichthyosis (Fig. 1195, 1197) and trichorrhexis invaginata (Fig. 1196). The alteration of the shaft gets evident after some months or years and even later appears the typical linear ichthyosis, which is usually preceded by ichthyosiform erythroderma.

"*Pili torti*", namely presenting torsions on their longitudinal axis, can be isolated or associated to other abnormalities of the shaft such as



Fig. 1195

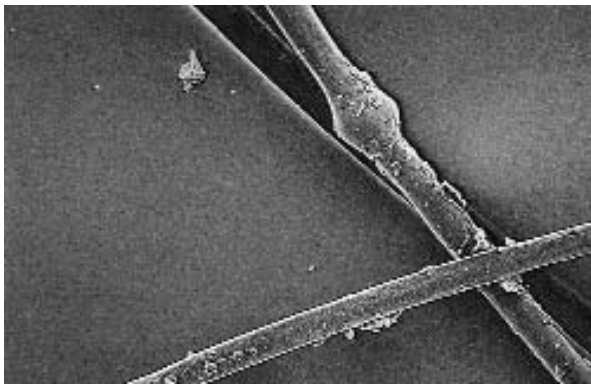


Fig. 1196



Fig. 1197

Fig. 1195, 1196, 1197: Netherton's syndrome in two sisters. Erythematous, linear lesions can appear precociously. The typical, bamboo-like invagination (Fig. 1196, courtesy of dr. Selvaag) of the hair occurs after the first years.



Fig. 1198

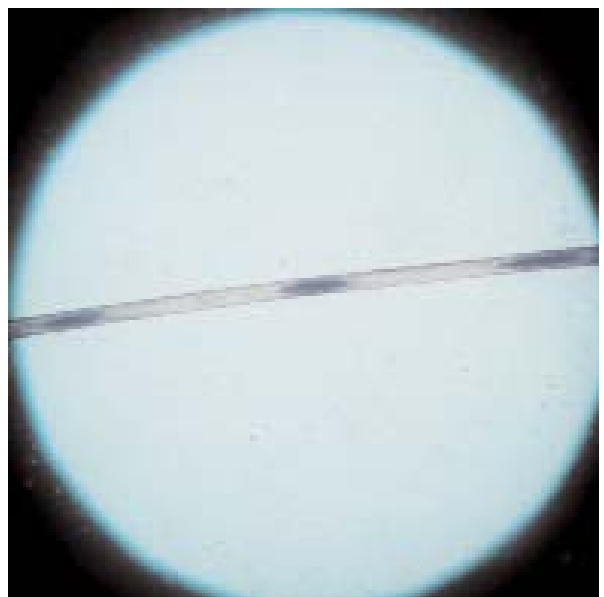


Fig. 1199

Fig. 1198, 1199: Pili anulati: all'esame obiettivo (Fig. 1198) e a luce ottica (Fig. 1199, 40x) bande chiare e scure alternate.

“pili canaliculi”, namely flat hairs with longitudinal grooves, or “pili annulati”, namely hairs presenting clinically (Fig. 1198) and on light microscopy (Fig. 1199) light and dark alternated bands, in numerous disorders.

We already mentioned *monilethrix* in the neonatal age.

In “*uncombable hair syndrome*” (Fig. 1200) can be observed hairs with triangular section and longitudinal grooves. This syndrome is probably due to a precocious keratinization of the internal epithelial sheath. It is clinically characterized by spun-glass, silvery-blond hair.

Woolly hair nevus is microscopically characterized by “pili torti et canaliculi” (Fig. 1202) and clinically by a more or less large tuft of curly hairs (Fig. 1201), which are lighter of the other smooth or wavy hair. Woolly hair nevus is often the continuation of an epidermal nevus of the face.



Fig. 1200: Uncombable hair syndrome with spun-glass hair. On light microscopy, the hair have a triangular section and longitudinal grooves.



Fig. 1201



Fig. 1202

Fig. 1201, 1202: The girl as in Fig. 1202 has an epidermal nevus of the left cheek. Level with the scalp, the latter is responsible for a woolly hair nevus, characterized (Fig. 1202) by “pili torti et canaliculi”.

Non scarring, diffuse, acquired hair loss. A diffuse hair loss can be due to telogen effluvium, generalization of alopecia areata, androgenetic alopecia and anagen effluvium.

Telogen effluvium (Fig. 1203) is the most frequent form of diffuse hair loss, occurring 2-3 months after the triggering cause. It is due to an early stop of the anagen phase. Due to the latter, many hairs simultaneously enter the telogen phase and fall out after the usual obligatory time in telogen. Usually, no more than 100 hairs fall out daily, whereas in telogen effluvium more than 1,000 hairs can fall out daily. Besides neonatal physiological effluvium, the most frequent causes in children are fever, infectious diseases, surgery, some drugs such as retinoids and valproic acid, nutritional defects such as biotin, iron and zinc deficiency, and psychological stress. When telogen effluvium is moderate with a loss lower than 25% of hair, the problem, which is much evident for the mother, is barely evident for the physician, because the residual hair resist to traction and because hair loss spontaneously regresses within 3-6 months when the responsible cause is eliminated.

Total (Fig. 1204) -namely complete loss of hair- and universal -namely loss of all hairs of the body- *alopecia areata* starts almost always



Fig. 1203: Various findings of the hairs in telogen effluvium on light microscopy, 40x.

in patches (Fig. 1206). The latter usually within weeks or months get confluent giving raise to a generalized loss. The involved skin is clinically normal with kept follicle orifices, as visible with a 10x magnifying lens.

Androgenetic alopecia (Fig. 1205, 1206) starts in the severe cases at the peripuberal age, localizing in the central portion of the scalp, where the hair undergo the miniaturization process till disappearing.

Acute *anagen effluvium*, which is less rare in children, is characterized by easy, non painful, extractability of hairs, maybe due to scarce adherence of the hair shaft to the surrounding epithelial sheath. Normal children have a few - less than 5 or 6- hairs with these characteristics (6). The latter should be taken into account when diagnosing loose anagen syndrome, which is characterized by apparently normal or thin, but not brittle, hair, often in children with red hair (4). Easily extractable hairs have an irregular bulb and an irregularly ruffled cuticula (5) and lack the epithelial sheath. The idiopathic form of this syndrome tends to improve with time (3). Besides the idiopathic form, there are inherited forms and forms associated to ectodermal dysplasias.

Anyway, before taking into account the diagnosis of loose anagen syndrome, physicians should rule out the possibility that anagen effluvium is due to an acute event, such as poisoning due to mercury, arsenic, thallium, boric acid and selenium. When in the history there is chemotherapy, the diagnosis is easy. The stopping of mitoses induced by the latter leads to the sudden interruption of the growing phase of the hair and to its easy extractability. The withdrawal of chemotherapy is followed by regrowth of the hair. However, this event does not always occur after chemotherapy, depending of the type of radiations and their penetrating capacity. After both chemo- and chemotherapy the color and structure of hair can change.

We remember in the prgriseofulvin era the iatrogenic, X-ray or thallium acetate induced hair loss, aimed at treating ringworm of the scalp.

Systemic disorders can be responsible, besides telogen, even for anagen effluvium. We

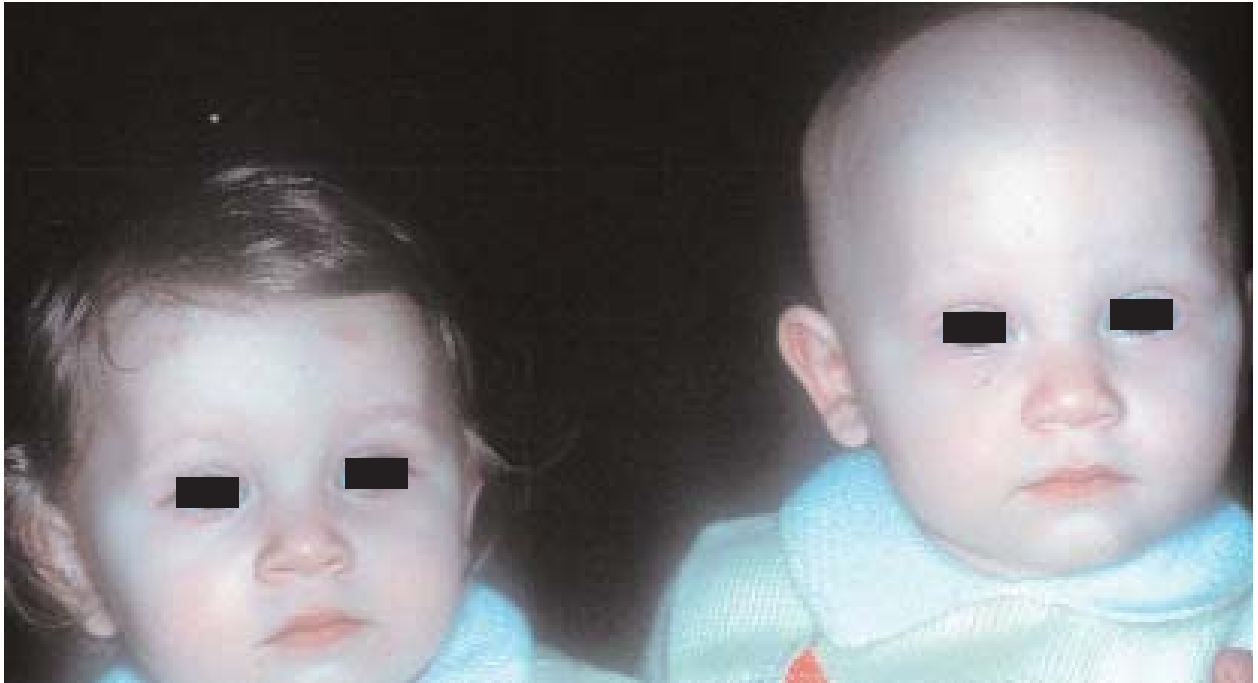


Fig. 1204: Total alopecia areata in one of two dizygotic sisters.



Fig. 1205: Severe androgenetic alopecia starts in peripubertal age.



Fig. 1206: Three types of alopecia, nuchal area celsi, linear traumatic and androgenetic alopecia on the vertex.



Fig. 1207

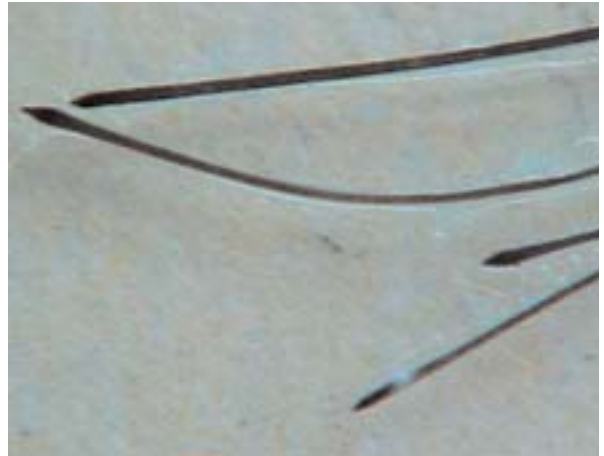


Fig. 1208

Fig. 1207, 1208: Anagen effluvium after cis-platinum (Fig. 1207) with characteristic arrow-shaped hairs (Fig. 1208, 10x).



Fig. 1209



Fig. 1210

Fig. 1209, 1210: Loose anagen syndrome (Fig. 1209), with typical microscopic appearance of the hairs (Fig. 1210, 10x).

should mention endocrinopathies such as hyperhypothyroidism, hypoparathyroidism, hypopituitarism, diabetes mellitus, Cushing and adrengo-

nital syndrome), deficiency of oligominerals, biotin, systemic lupus erythematosus, chronic renal failure and malignancy.

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