147 Nail Disorders

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The nail is the biggest cutaneous appendage. It is an integral part of the functional unit tip of the digit, supports its sensory functions, and is an important tool for manual dexterity, for scratching, and for defense.

The nail develops from fetal week 9 onward and requires an intact terminal phalanx as well as a series of signaling proteins for its correct shape and size. At week 13, the nail field is fully developed and at week 14, the nail plate starts growing to cover the nail field almost completely by week 17. From week 20 onward, the digit and the nail grow synchronously, and at birth the nail plate has normally reached the distal ridge of the finger. If the nail is still shorter, the infant may be born with a distally ingrowing nail. Koilonychia is also frequently seen at birth, but usually disappears spontaneously.

The nail is a plate of keratin containing both hard "hair" keratins and soft epidermal keratins. It is transparent allowing the distal matrix – the lunula – and the nail bed to be seen. The nail is both physically and chemically resistant. Antimicrobial peptides in the nail protect it against bacterial and fungal attack even though it does not have any vascular supply.

The nail is formed in its entirety by the matrix (\bigcirc *Fig.* 147.1), which continuously produces nail substances during the whole life. Most of the matrix is covered by the proximal nail fold, the ventral surface of which forms a horny layer, the eponychium, that is firmly attached to the dorsal surface of the nail. When the nail grows out, it pulls part of this keratin layer with it forming the cuticle, which seals the nail pocket. When this attachment is lost, usually as a result of inflammation, infection, or trauma, the cuticle disappears and foreign material may enter under the proximal nail fold. This further irritates the tissue giving rise to paronychia. In newborns, this may be due to *Candida albicans*, probably acquired during the birth process.

The nail bed is seen as a pink structure through the normal nail. What gives it this pink color is still not completely understood, although it is thought to be due to the particular parallel arrangement of the capillaries running longitudinally in several layers one above the other. The nail is a window to the bed permitting the vascular supply and blood oxygenation to be evaluated. Even subtle color changes can be seen through the thin newborn nail and give important hints at general health indicators. The nail plate is very firmly attached to the nail bed, which produces just a thin layer of keratin allowing the nail to slide over it without losing its attachment.

The distal margin of the nail divides from the nail bed at the hyponychium. This, again, is a specialized structure preventing foreign substances from entering under the nail. Detachment of the nail from the nail bed is called onycholysis, usually, as a result of infection or trauma, and is mostly seen in girls due to overzealous manicure.

The lateral nail folds, together with the proximal one, form a frame for the nail ensheathing it on three sides.

The Newborn Nail

In the newborn, the nail is soft, thin, and pliable. If the big toenail has not yet overgrown the hyponychium, it may grow distal-laterally or laterally. This is easily treated by the mother: a lubricating ointment is applied on the tip of the toe, and in a warm bath, the distal and/or lateral nail folds are gently massaged away from the nail. Within a few weeks, the nail will have reached the tip of the toe.

Koilonychia or spoon nails are frequent and harmless. This minor abnormality disappears by itself within months or the first 2 years of life.

Some weeks after birth, transverse grooves running parallel to the lunula border may be seen. These Beau's lines are thought to be due to the trauma of birth that may temporarily slow down the nail growth rate.

A large number of congenital hereditary diseases and intrauterine damages can affect the nails (**Tables 147.1** and **147.2**). Ectodermal dysplasias (ED) may also affect the nails, particularly the group of tricho-onychotic EDs. Small and hypoplastic nails or fluting are the most common nail changes. Pachyonychia congenita type 1 is a disease of keratins 6a/16 and type 2 of 6b/17.

Nail Changes in Infants

Some infants develop a hypertrophic medial nail wall, which may cover up to one-half of the nail. Debris is



Figure 147.1

Schematic illustration of the nail. (a) Sagittal section through the distal phalanx of a finger. C Cuticle, DP Distal phalangeal bone, E Eponychium, ET Extensor tendon, FT Flexor tendon, HO Hyponychium, L Lunula, M Matrix, NP Nail plate, PNF Proximal nail fold. (b) Growth rate of a normal nail

Table 147.1

Nail alterations in chromosomal anomalies (Adapted from Haneke E (2006) Nagelkrankheiten. In: Hamm H (ed) Pädiatrische Dermatologie, 2nd edn. Springer, Heidelberg, pp 677–698)

Condition	Nail changes	Other symptoms
Trisomy 3q	Hypoplasia, anonychia	Hirsutism, synophris, eye anomalies, short neck
Monosomy 4p	Overconvex	Fish mouth, cheilopalatoschisis, head asymmetry, preauricular pits, retardation
Trisomy 7q	Overconvex	Facial asymmetry, retardation
Trisomy 8p	Hypoplasia, anonychia	Facial asymmetry, retardation
Monosomy 9p	Wide convex	Hypoplasia of middle face, trigonocephaly, upward slanted eyes, abnormal dermatoglyphics, retardation
Trisomy 9p	Dystrophic, claw-like	Downward slanted eyes, microcephaly, short digits, retardation
Trisomy 13	Narrow, convex, hypoplastic	Cheilopalatoschisis, aplasia cutis, polydactyly, hemangiomas, microcephaly, vitium cordis, genital hypoplasia
Trisomy 18	Hypoplasia of fifth finger and fifth toe	Cramped hands with overlapping fingers, short sternum, abnormal dermatoglyphics
Trisomy 21 (Down's syndrome)	Clubbing and macronychia	Upward slanted eyes, hypoplasia, short broad fingers, four-finger-fold, vitium cordis, cutis marmorata
X0 (Turner syndrome)	Small, convex, deep lying	Pterygium colli, short neck, congenital lymphedema, ovarial dysgenesis, cubitus valgus, nevi
Ring chromosome group C	Pachyonychia	Facial anomaly, retardation

collected in the deep fold leading to subacute inflammation. Massaging from the nail to the plantar surface of the tip of the toe is usually effective. When it does not respond, it may simply be removed and the small wound left for second intention healing.

Congenital malalignment, originally described as congenital dystrophy of big toenails, is another

abnormality of infant nails. Often, the distal phalanx of the big toe is deviated laterally and the nail even more. This may lead to oblique shearing trauma causing onycholysis and, as its consequence, the nail becomes oystershell-like distorted, triangular, discolored, thickened, and sharply bent medially. The condition may improve spontaneously up to the age of 2 years; however,

Table 147.2

Teratogens and their effects on nails (Adapted from Haneke E (2006) Nagelkrankheiten. In: Hamm H (ed) Pädiatrische Dermatologie, 2nd edn. Springer, Heidelberg, pp 677–698)

Teratogen	Nail changes	Other symptoms
Alcohol	Hypoplasia, anonychia of fifth finger, convex	Microcephaly, small lid opening, epicanthus, short nose, flat philtrum, retardation
Phenytoin	Hypoplasia, longitudinal pigmented streaks	Cheilopalatoschisis, deep-set ears, hirsutism, sunk-in nasal bridge, broad mouth, short neck, finger-like thumbs, short tapering fingers, wide distance between nipples
Trimethadione	Hypoplasia	Microcephaly, V-shaped merging eyebrows, ptosis, malformed ears, small flat nose, deafness
Warfarin	Hypoplasia	Hypertelorism, small nose, short neck, brachydactyly, spotted epiphyses

if this does not occur, the nail bed shrinks permanently, a distal nail wall develops, and the nail becomes more and more unsightly, sometimes painful. Unfortunately, there is no sign indicating spontaneous healing. Treatment is surgical by the age of 2 years with rotation of the entire nail organ into its correct axis.

Nail Changes in School Children

Ingrown nails are a common condition in children from around 12 years onward. There is a discrepancy between a relatively wide nail and a narrow nail bed distally. In most cases, the nails are markedly curved. Usually, the patient tends to cut the distal corner of the nail, leaving small spicules in the depth of the distal lateral nail groove, which pierce into the soft tissue when the nail grows out. Three stages of ingrown nails are distinguished: (1) Reddening and swelling of the lateral nail fold. (2) Secretion. (3) Granulation tissue. There may be fluctuations between stages 1 and 3 depending on foot care and hygienic measures. Treatment may be conservative in early stages with taping, packing, gutter treatment, artificial nails, or nail braces. This is a long-term treatment and requires good patient compliance. When conservative treatment is not successful or feasible, definitive surgical cure is achieved by selective lateral matrix horn resection or phenolization. Wedge excisions are obsolete.

Nail Signs

There are some nail changes that may be – to a certain degree – a marker of an internal or general condition.

Beau's lines are transverse, slightly curved furrows that run parallel to the lunula border. They indicate

a temporary slow down in the nail growth that had occurred some weeks ago, most commonly a disease with high fever. Beau's lines are more marked in fastgrowing fingernails than in slow-growing toenails. Single-digit Beau's lines are typical sequelae of a past surgical intervention.

Koilonychia, also called spoon nails, is typically seen in iron deficiency. It is a frequent finding in the big toenails of newborns and usually disappears spontaneously within a few weeks.

Leukonychia occurs as small dots, short transverse lines, or rarely as narrow longitudinal lines. Punctate and striate leukonychia is frequent in young girls, mostly due to overzealous manicure. Longitudinal leukonychia is caused by a narrow rim of keratin running from the lunula to the free nail edge; its cause is usually an onychopapilloma.

Melanonychia most commonly occurs as a longitudinal brown streak. In children, it is usually due to either a lentigo or a junctional melanocytic nevus of the matrix; nail melanomas are extremely rare in children. In warmer climate, some pathogenic fungi may produce a soluble melanin that may give rise to a grayish-to-blackish spike in the nail. In contrast to true melanonychia, subungual hematomas never reach the free nail margin and can easily be diagnosed dermatoscopically. Most hematomas have a history of a previous trauma.

Most other colorations are due to exogenous dyes or drugs.

Onycholysis is the distal separation of the nail from the nail bed. It has often a curved smooth border; this is called onycholysis semilunaris and is a result of manicuring the nail with sharp instruments.

Proximal onycholysis is called *onychomadesis*. It develops when the nail stops growing for a certain period. Single-digit onychomadesis typically develops after a

run-around or acute paronychia, whereas general diseases cause onychomadesis in a symmetrical fashion. It may be followed by nail shedding.

Characteristic Nail Conditions

Twenty nail dystrophy is characterized by the gradual development of rough nails of both the fingers and toes, a phenomenon called trachyonychia. The nails show fine depressions on their surface. One nail after the other may be affected, but very often one single nail remains spared. In the more common form, the nails lose their shine, but there is also a shiny variant. Skin lesions are not found. In most cases, histopathology exhibits a spongiotic dermatitis of the matrix; less frequently, a lichen planus, psoriasis, or eczema may be seen. Whether or not the spongiotic changes indicate an isolated alopecia areata of the nails remains to be clarified. The course is usually benign in children with spontaneous resolution around the age of 16. A combination of topical calcipotriol with a potent steroid accelerates healing.

Twenty nail dystrophy must be differentiated from nail lichen planus as this may cause permanent nail dystrophy even in children. The nails are rough, ridged longitudinally, and tend to break.

Nail Psoriasis

Nail involvement in children with psoriasis is not rare (see Papulosquamous and Related Disorders Including Psoriasis). Subungual hyperkeratosis, distal onycholysis, pits (**P***Fig. 147.2*), and so-called salmon or oil spots are the most frequent signs. Pits and small whitish spots



Figure 147.2 Psoriasis of the nails in a 13-year-old boy

develop from tiny psoriatic lesions of the most proximal part of the matrix. Salmon spots represent psoriatic plaques of the nail bed. Psoriatic leukonychia is due to a psoriatic affection of the middle of the matrix. Complete nail destruction indicates psoriatic involvement of the entire nail organ and is often seen in arthropathic psoriasis. In children, the nails may also become very thick and rough with huge subungual hyperkeratosis. Treatment of nail psoriasis is difficult as the nail wall and plate cover the diseased portions of the nail organ. Any systemic therapy resulting in clearance of the skin potentially also improves nail psoriasis.

Lichen Planus

Roughly one-tenth of the patients with lichen planus develop nail changes. In about one-quarter, the nail changes come after diagnosis of skin and mucosal lesions. Most characteristically, this condition affects the proximal and dorsal matrix with subsequent ridging, roughness, and loss of shine of the nail. The problem with nail lichen planus is its propensity to permanently destroy the nail. Early treatment is therefore mandatory. Topical vitamin D₃ analogues, best in combination with a steroid, should be started first. When this is ineffective over a course of >3 months, low-dose etretinate may be given. An alternative is intramuscular triamcinolone acetonide crystal injections, about 0.5 mg/kg bodyweight every 4–6 weeks.

Alopecia Areata

Alopecia areata often affects the nail. The more severe it is, the more likely are the nail changes. The nails become rough, nontransparent, and lose their shine. Thickening and brittleness occur when the nail plate contains a lot of serum inclusions. Histologically, alopecia of the nail represents a spongiotic dermatitis and it may be difficult to differentiate it from eczema. No specific treatment is known, but it appears that ungual alopecia areata runs a benign course.

Eczema

The different forms of eczema may cause variable nail changes. Rubbing with the surface of the nail in chronic atopic eczema is the main cause for the characteristic shiny nails. In dyshidrotic eczema, the periungual skin may be involved with small itching vesicles. Allergic contact dermatitis may cause nail deformation, particularly when the allergen gets under the proximal nail fold and induces an eczematous change in the matrix. In chronic eczema, a thickening of the proximal nail walls develops with rounding of its free margin and spontaneous loss of the cuticle. This leads to separation of the nail fold's undersurface with the nail, and more foreign substances, including allergens, microbes, and dirt, may enter the nail pocket. In girls, nail cosmetics may play a causative role.

Erythema Multiforme

Erythema multiforme is a skin disease characterized by round erythematosus lesions that develop a central blister, the roof of which often becomes necrotic and thus gray. The tricolor appearance make the lesions look like targets. When the nail organ itself is involved, onychomadesis, nail dystrophy, and later shedding may develop.

Infections and Infestations

Viral warts are common in periungual locations and are less frequent subungually. They are often rather oval than round, gravish hard lesions with a rough and sometimes clefted surface. The problem is not the diagnosis, but rather their treatment. A conservative approach with aggressive keratolysis and regular cautious curettage of the necrotized wart keratin has been adopted. Saturated monochloroacetic acid is applied sparingly on the wart and allowed to dry. It is then covered with 50% salicylic acid plaster and fixed with adhesive band. With all this on, the patient has to perform a hand or footbath as hot as tolerated twice a day for about 10 min. After a week, the adhesive tape and salicylic acid plaster is removed, showing a white soft surface of the wart. This is gently curetted and the procedure repeated until the wart is gone. The success rate is over 90%. However, after several debridements, the monochloroacetic acid tends to cause a nagging pulsating pain, and the treatment has to be interrupted. Whether duct tape is an alternative remains to be seen. Cryotherapy has about the same cure rate, but is very painful at the nail. Lasers are not more effective than other destructive methods. Limited evidence exists for the efficacy of topical 5-fluorouracil, intralesional interferons, photodynamic therapy, imiquimod, and a variety of vaccinations. Bleomycin, dinitrochlorobenzene, and other obligate topical sensitizers, 5-fluorouracil, interferons, and photodynamic therapy, are potentially hazardous or toxic treatments.

Digital herpes simplex is often underdiagnosed. It is commonly misdiagnosed as a recurrent paronychia or

felon. It presents with painful blisters around the nail or on the pulp of the digit that are barely visible under the thick horny layer of this region. Very often, a red stripe indicating lymphangitis is seen, sometimes even before any visible vesicle. Within a few days, the blisters may become putrid. When the diagnosis is made early enough, acyclovir may be used. Antibiotics are of no use.

Although *mollusca contagiosa* are probably transmitted with the scratching finger nail, their localization at the nail is very rare.

Cow pox, milker's nodule, and orf are clinically almost identical lesions caused by similar viruses. The first are usually acquired from wild living cats, the second from an infected udder, and the last from sheep. One to two weeks after infection, a nodule develops with sanguinolent secretion. Healing takes a few weeks.

The most common bacterial infection is *bulla repens*, also called run-around. This is often derived from hangnails that are torn out and leave a small wound. It is mostly due to staphylococcus aureus, less frequently streptococci. In the beginning, a clear blister develops, which soon becomes cloudy, and yellow pus can be seen. Pain is moderate to severe. Treatment of choice is opening of the blister and disinfective baths twice daily as well as dressings with antimicrobial ointments. Systemic treatment is rarely necessary.

A *whitlow* is a deeper infection due to pyogenic micrococci. Reddening, swelling, and pulsating pain are characteristic. A subungual felon may cause irreversible matrix damage within 24–48 h in children and should therefore be treated as an emergency. A swab has to be taken for bacterial culture and systemic treatment with a staphylococcus-specific antibiotic begun until the result of sensitivity testing has arrived.

Blistering dactylitis is a streptococcal infection that often remains undiagnosed as it usually causes no symptoms. Blisters occur on the tip of the digits and the horny layer later sheds off. They may represent a chronic streptococcal focus and require treatment. The blister roof is cut and the finger disinfected. Systemic penicillin treatment is recommended.

Onychomycoses are fungal infections, most commonly of the nail bed. Dermatophytes are responsible for more than 80% of the cases. They are rare in infants and increase in prevalence with age.

Distal lateral subungual onychomycosis is the most frequent type. The fungus infects the tip of the digit and then the hyponychium and nail bed, where it slowly grows proximally toward the matrix. The nail plate itself is rather an obstacle for the fungus than the site of infection; thus the nail plate covers the infection and makes it difficult to treat. The nail bed develops a subungual hyperkeratosis that is Nail Disorders

seen as a yellowish substance through the nail. Only in late advanced stages will the plate be destroyed (**•** *Fig. 147.3*).

In proximal subungual onychomycosis, the fungus breaks the barrier of the cuticle and grows along the underside of the proximal nail fold toward the matrix. Once this is reached, the fungus is included into the growing soft nail and penetrates it in virtually all layers. The nail bed does not produce a subungual hyperkeratosis. The nail that is invaded by the fungus takes on a whitish color; hence, the term proximal subungual white onychomycosis is often used (\bigcirc Fig. 147.4).

Superficial white onychomycosis is a peculiar infection of the surface of the nail plate. Several molds may be the



Figure 147.3Onychomycosis in a 12-year-old boy



Figure 147.4 Subungual exostosis of the big toe in an 11-year-old girl

causative pathogen, particularly in warm climate, whereas *Trichophyton mentagrophytes* is the common pathogen in temperate climate zones.

Endonyx onychomycosis is mainly caused by *T. soudanense* and *T. violaceum*. The pathogen grows entirely in the nail plate.

Any of these onychomycosis types may progress to *total dystrophic onychomycosis*. This, however, is also a common finding in *chronic mucocutaneous candidosis*.

Treatment of childhood onychomycoses is hampered by the fact that the potent systemic antifungals are often not approved for use in children. As long as the infection has not yet reached the matrix, an antimycotic lacquer may be used, but the treatment has to be performed with consistent compliance over many months. When the matrix is reached, in proximal subungual and total dystrophic onychomycosis, systemic treatment is advised. Depending of the age and weight, terbinafine, fluconazole, or itraconazole are the most potent drugs. In some countries, griseofulvin is still available. Chemical nail avulsion supports treatment in total dystrophic onychomycosis.

Nail involvement is not uncommon though often overlooked in *crustous scabies*. There is subungual hyperkeratosis and some nail roughness. There may be hundreds of mites under the nails, and failure to treat subungual scabies usually results in recurrence and spread of the infection. Treatment is with topical scabicide drugs, which have to be brushed in twice daily over several days. Ivermectin is an alternative.

Tungiasis is common in endemic regions and often causes some sort of paronychia. Under the nail in the region of the hyponychium, the female *tunga penetrans* digs itself into the skin and causes an itching nodule growing to the size of a pea. A small black dot represents the opening through which the sand flea lays its eggs. Treatment of choice is curettage of the hole in which the flea lives in order to prevent infectious complications such as erysipelas, cellulitis, or gangrene.

Tumors of the Nail Organ

Benign nail tumors are not exceptional in childhood. They often differ in their clinical appearance from the same tumors in other locations.

Koenen Tumors

Koenen tumors are round to sausage-shaped lesions that develop in about one-half of the patients with tuberous sclerosis from the age of 12 years onward. They are flesh

Recurrent Digital Fibromatosis of Reye

In early infancy, reddish dome-shaped lesions develop on the digits with the exception of the thumbs and big toes. When they grow in the periungual skin, they may distort the nail. Treatment is not necessary as they disappear spontaneously.

Keloids

Keloids in the nail region are rare, but may be monstrous. The most common cause is electrosurgery for periungual warts, which should therefore be abandoned. They may be painful and cosmetically embarrassing.

Angiomas

Pyogenic granuloma, an eruptive angioma, is sometimes seen around or even under the nail. It presents as a rapidly growing red nodule that tends to erode, superficially ulcerate, and bleed easily. It responds to potent topical steroids under occlusion, but may also be carefully removed at its base with cautery of the feeder vessel. The specimen has to be examined histopathologically in order to rule out other tumors.

Port-wine stains rarely affect the nail region. Whereas the periungual skin is deep violaceous red, the nail itself is usually white.

Glomus tumors are exceptional in children.

Exostosis

Subungual exostosis is a reactive lesion that may occur at any age. The most characteristic localization is the medial dorsal aspect of the distal phalanx of the big toe, but other digits may be involved as well. They present as a stonehard round lesion covered with skin that is extended and shiny, usually without the typical ridges of plantar skin. A radiograph confirms the diagnosis. Treatment of choice is the generous surgical removal at the base.

Diagnostic Measures

In most cases, nail alterations do not require many sophisticated and expensive tests. Careful clinical inspection of the digits in relaxed position and with pressure to check potential vascular changes is important. Probing can localize circumscribed pain. Transillumination is useful to check for a cystic lesion or a foreign body. Bacterial and fungal cultures are indicated when an infection is suspected. Radiographs have to be very soft in order not to overexpose the distal phalanx. Computed tomography and magnetic resonance imaging are sometimes helpful as is variable high-frequency ultrasound. Direct microscopy of subungual keratin allows to find fungi. However, the gold standard for diagnosis is histopathology, which requires a biopsy.

Nail Biopsy

In case of onychomycosis, a nail clipping stained with PAS is twice as sensitive as a fungal culture.

For an incisional biopsy, a local anesthesia is applied. This may be a distal or proximal digital block or a transthecal block. The biopsy site within the nail organ is crucial: Changes involving the nail surface require proximal matrix tissue, best as a lateral longitudinal nail biopsy. A punch may be taken for nail bed lesions but should not have a diameter greater than 3 mm.

Nail Avulsion

This is very rarely helpful and must never be done as a substitute for a diagnosis. Many conditions are worsened by avulsing the nail.

Under local anesthesia, a blunt, slightly curved nail elevator is inserted under the proximal nail fold to separate it from the underlying nail by careful back-and-forth movements from one side to the other. The elevator is then pushed through the hyponychium till the matrix to detach the plate from the nail bed and matrix. The proximal avulsion approach detaches the nail from the matrix; this is technically more demanding but less traumatizing.

When the nail plate is lacking after avulsion, the pulp of the tip of the digit may be dislodged dorsally; this is often the case in the big toenails. A distal nail wall develops that prevents the nail from growing out. Instead, it becomes thicker, yellowish, and intransparent. With time, it loses its attachment with the nail bed.

Subungual Hematoma

Children often traumatize a finger; most commonly it is a car door crash. The result is a hematoma that causes considerable pain. When it stretches over more than 50% of the nail field, a fracture is likely and requires adequate repair. In smaller hematomas, drilling a hole over it and releasing the blood immediately relieves the pain.

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