Nail Disorders

Antonella Tosti and Bianca Maria Piraccini

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ANATOMY (Fig. 71.1)

Key features

- Nail matrix → nail plate production
  proximal matrix → dorsal nail plate
  distal matrix (lunula) → ventral nail plate
- Proximal nail fold → nail matrix protection
- Nail bed and hyponychium → nail plate adhesion and distal detachment
- Nail growth rate:
  fingernails: 3 mm/month
  toenails: 1 mm/month

The nail plate is a fully keratinized structure produced by the germinative epithelium of the nail matrix. As it grows, the nail plate emerges from the proximal nail fold and progresses distally, lying across and strictly adhering to the nail bed. As the nail plate approaches the tip of the digit, it detaches from the underlying tissues, forming the hyponychium. Proximally and laterally, the nail plate is surrounded by the nail folds. The proximal nail fold consists of two layers of epithelium – a dorsal layer, which is the distal continuation of the skin of the dorsum of the digit, and a ventral layer, which is continuous with the nail matrix.

The nail matrix is responsible for the production of the nail plate and consists of an epithelium that keratinizes without the formation of a granular layer. Nail matrix keratinization occurs along an oblique axis, reflecting the upward and distal movement of cells during the process of maturation and differentiation. For this reason, the proximal portion of the nail matrix produces the dorsal portion of the nail plate, while the distal matrix is responsible for the production of the ventral nail plate. The distal portion of the nail matrix is visible through the transparent nail plate as a white, distally convex half moon known as the lunula. The matrix contains melanocytes that are normally quiescent; however, they may become activated and synthesize melanin, which is transferred to the surrounding keratinocytes. Distal migration of melanin-containing keratinocytes then gives rise to a pigmented nail plate.

Nail plate production occurs continuously, proceeding from the 15th week of embryonic life until death. Under normal conditions, the mean growth rate of a fingernail is 3 mm/month and that of a toenail is 1 mm/month. Nail growth rate can be influenced by several factors, including age, systemic diseases, and medications.

NAIL SIGNS

By clinical examination, it is possible to identify which site within the nail apparatus is affected by a particular disease process (Table 71.1 &

![Schematic drawing of the nail apparatus in longitudinal section.](image)

Table 71.1 Correlation of nail findings with anatomic site of nail damage.

<table>
<thead>
<tr>
<th>Affected site</th>
<th>Clinical manifestation</th>
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<tbody>
<tr>
<td>Proximal matrix</td>
<td>Beau's lines, Pitting, Longitudinal ridging, Longitudinal fisurining, Trachyonychia</td>
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<tr>
<td>Distal matrix</td>
<td>True leukonychia</td>
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<td>Proximal + distal matrix</td>
<td>Onychomadesis, Koilonychia, Nail thinning</td>
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<tr>
<td>Nail bed</td>
<td>Onycholyis, Subungual hyperkeratosis, Apparent leukonychia, Splinter hemorrhages</td>
</tr>
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</table>
Fig. 71.2. Recognizing specific nail signs is very important in understanding and diagnosing nail dystrophies. Nail signs can be schematically divided into three major categories:

- Signs due to abnormal nail matrix function
- Signs due to nail bed disorders
- Signs due to deposition of pigment within the nail plate.

**Nail Signs due to Abnormal Nail Matrix Function**

**Beau's lines**

<table>
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<th>Key features</th>
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<tr>
<td>Transverse depressions</td>
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<td>More evident in the central portion of the nail plate</td>
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<tr>
<td>Most often traumatic</td>
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<tr>
<td>Involvement of multiple digits suggests a systemic cause</td>
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</tbody>
</table>

First described by Beau in 1846, these transverse depressions of the nail plate surface result from a temporary interruption of the mitotic activity of the proximal nail matrix. The depth of the depression indicates the extent of the damage within the matrix; the width of the depression (along the longitudinal axis) indicates the duration of the insult. Beau's lines grow distally with the nail plate, with multiple lines indicating repeated damage. Most commonly, Beau's lines are caused by mechanical trauma (e.g. manicures, onychotillomania) or dermatologic disease of the proximal nail fold (e.g. eczema, chronic paronychia). The presence of Beau's lines at the same level in all nails suggests a systemic cause (e.g. severe or febrile illness, erythroderma, drugs).

**Onychomadesis (nail shedding)**

<table>
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<th>Key features</th>
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<tr>
<td>Proximal detachment of the nail</td>
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<tr>
<td>Most often traumatic</td>
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<tr>
<td>Involvement of multiple digits suggests a systemic cause</td>
</tr>
</tbody>
</table>

Fig. 71.2. Nail signs and nail disorders.
The term “onychomadesis” describes the detachment of the nail plate from the proximal nail fold and is due to a severe insult that produces a complete arrest of nail matrix activity [Fig. 71.3]. Onychomadesis appears as a sulcus that replaces the proximal nail plate. The causes of onychomadesis are the same as those for Beau’s lines⁴.

**Pitting**

**Key features**
- Punctate depressions of the nail plate surface
- Migrate distally with nail growth
- Proximal nail matrix damage by psoriasis, alopecia areata, eczema

Pitting describes the presence of small depressions on the nail plate surface [Fig. 71.4]. Pits are due to foci of abnormal keratinization of the proximal nail matrix that result in clusters of parakeratotic cells in the dorsal nail plate. These clusters are easily detached, leaving the pits. Diseases that produce nail pitting include psoriasis, alopecia areata and eczema.

**Onychorrhexis**

**Key features**
- Thinning
- Longitudinal ridging and fissuring
- Diffuse nail matrix damage by lichen planus is a possible cause
- Mild disease is a common manifestation of aging

Onychorrhexis produces longitudinal ridging and fissuring of the nail plate. Often associated with nail thinning, it indicates diffuse damage to the nail matrix. Diseases commonly responsible for onychorrhexis include lichen planus, impaired vascular supply, trauma, and tumors that compress the nail matrix. Mild longitudinal ridging of the nails is a common feature of aging.

**Trachyonychia (twenty-nail dystrophy, sandpapered nails)**

**Key features**
- Nail roughness
- Often associated with thinning
- Conditions that may cause trachyonychia include alopecia areata (common), lichen planus (rare), psoriasis (rare) and eczema (very rare)

Trachyonychia is a specific nail plate surface abnormality characterized by diffuse homogeneous roughness [Fig. 71.5]. In most patients, the affected nails are opaque, lusterless and rough; the nail plate surface has longitudinal ridging due to fine superficial striations distributed in a regular, parallel pattern (sandpapered nails). A less common variant, referred to as shiny trachyonychia, is characterized by multiple small punctate depressions distributed in a geometric pattern within parallel, longitudinal lines. Conditions that may cause trachyonychia include alopecia areata, lichen planus, psoriasis and eczema. Some authors use the term “twenty-nail dystrophy” for idiopathic cases of trachyonychia in children [see below], even though the disease does not necessarily involve all 20 nails.

**True leukonychia**

**Key features**
- White opaque discoloration
- Punctate, striate or diffuse
- Most often traumatic
- Distal nail matrix damage
- Needs to be distinguished from apparent leukonychia (nail bed discoloration)
- Needs to be distinguished from pseudoleukonychia (nail plate invasion by fungi), which is typical of superficial white onychomycosis. In this case the nail plate surface is friable due to the presence of keratin debris and fungal elements.
The nail plate has a normal surface but loses its transparency and looks white because of the absence of parakeratotic cells within its ventral portion. True leukonychia is caused by diseases that disturb distal nail matrix keratinization and it presents with three morphologic variants:

- **Punctate leukonychia.** The nail plate shows small opaque white spots that move distally with nail growth and sometimes appear before reaching the distal nail. It is caused by trauma and is most commonly observed in the fingernails of children.
- **Striate leukonychia.** The nail plate shows one or more transverse white opaque parallel lines. It is frequently observed in the fingernails of women, due to matrix trauma secondary to manicures. It may also occur in great toenails as a consequence of trauma from shoes. It is also typical of Mees’ lines, the white transverse bands seen in arsenic and thallium poisoning.
- **Diffuse leukonychia.** The nail plate is completely or almost completely opaque and white. Total leukonychia is rare and sometimes hereditary. It may be associated with keratodermas and other congenital defects such as deafness.

**Koilonychia (spoon nails)**

**Key features**
- Thinned concave nails
- Physiologic in the toenails of children

The nail plate is thinned, flat and spoon-shaped due to upward eversion of its lateral edges. Although it is physiologic and resolves spontaneously in children's toenails, in adults it is most commonly occupational in origin or associated with severe iron deficiency.

**Nail Signs due to Nail Bed Disorders**

### Onycholysis

**Key features**
- Distal nail plate detachment
- Detached nail looks yellow–white
- Commonly due to environmental trauma, psoriasis or onychomycosis

The nail plate is detached from the nail bed and appears white because of the presence of air in the subungual space. Pigmentation of the onycholytic area may occur as a consequence of microbial colonization or blood extravasation. In addition to environmental trauma, the nail bed disorders that most commonly produce onycholysis are psoriasis and onychomycosis (Fig. 71.6). Photo-onycholysis may be precipitated by UV light exposure, either alone or in combination with medications such as tetracycline.

### Onychauxis

**Key features**
- Nail thickening
- Subungual hyperkeratosis
- Commonly due to psoriasis or onychomycosis

The nail plate appears thickened due to the presence of subungual scales. Causes of subungual hyperkeratosis include psoriasis, onychomycosis and eczema.

**Apparent leukonychia**

**Key features**
- White discoloration that fades with pressure
- Nail plate transparency maintained
- Often due to drugs (chemotherapeutic agents) or systemic diseases (e.g., hypoaalbuminemia)

The nails are white because of abnormalities in the color of the nail bed and this is usually due to nail bed edema. Apparent leukonychia does not move distally with nail growth and the white discoloration fades with pressure (see below for different types).

**Splinter hemorrhages**

**Key features**
- Thin longitudinal dark-red subungual lines
- Most often traumatic
- Damage to longitudinally oriented nail bed capillaries

Splinter hemorrhages appear as dark-red, thin longitudinal lines, usually localized to the distal portion of the nail. The shape of the hemorrhages is due to the longitudinal orientation of nail bed capillaries. The most common causes of splinter hemorrhages are trauma, psoriasis and onychomycosis. Proximal splinters are rare and possible indicators of systemic diseases, including endocarditis (infectious and marantic), vasculitis [including septic vasculitis, trichinosis and the antiphospholipid antibody syndrome]

A recently described vascular lesion of the nail bed is the “red comet”, which occurs in ~30% of patients with tuberous sclerosis complex (Fig. 71.6). Red comets appear as longitudinal short red streaks, partially blanchable with compression, with an enlarged distal end; they extend from the mid to the distal third of the nail and never reach the free edge of the nail. Red comets are thought to represent telangiectasias plus extravasated blood.

**Nail Signs due to Deposition of Pigment**

**Key features**
- Exogenous → convex proximal border
- Endogenous → concave proximal border (distally convex)
- Subungual deposition → often associated with onycholysis

Causes of nail pigmentation include staining from external pigment, presence of pigment under the nail plate, and deposition of pigment within the nail plate. Nail pigmentation from external staining typically follows the shape of the proximal nail fold. Common examples are yellow-brown nail discoloration due to nicotine in smokers or darkening of the nail plate due to hair dyes in hairdressers. Subungual deposition of pigment is often seen with *Pseudomonas* colonization, where the green discoloration of the nail results from the production of pyocyain by the bacteria. Dermatophytes usually cause a yellow–white subungal pigmentation. The nail plate may be pigmented secondary to melanin deposition or deposition of other pigments such as iron or gold. A Fontana–Masson stain of a nail clipping is useful in differentiating melanin from other pigments.

**Longitudinal melanonychia**

**Key features**
- Longitudinal brown to black band
- Commonly seen in darkly pigmented individuals
- Single band may be a sign of nail melanoma
- Multiple bands often due to drugs or systemic disease

The nails are white because of abnormalities in the color of the nail bed and this is usually due to nail bed edema. Apparent leukonychia does not move distally with nail growth and the white discoloration fades with pressure (see below for different types).
Longitudinal melanonychia is very commonly observed in individuals with darkly pigmented skin, and up to 90% of adult African-Americans have one or more pigmented bands. It is seen much less frequently in Caucasians. Longitudinal melanonychia is due to the presence of melanin within the nail plate, and it may be caused by simple activation of nail matrix melanocytes or by melanocyte hyperplasia as in a lentigo, nevus or melanoma.

Clinically, melanonychia presents as one or more longitudinal pigmented bands extending from the proximal nail fold to the distal margin (Fig. 71.7). The band of melanonychia can vary in color from light brown to black and the pigmentation may be homogeneous or variable. The width ranges from a few millimeters to the whole nail width. Multiple bands are usually due to melanocyte activation (Table 71.2).

A range of dermoscopic patterns has been described for longitudinal melanonychia (e.g. regular, thin, parallel light brown lines reflecting melanocytic activation), but their accuracy with regard to the diagnosis of subungual melanoma has not been established (see below).

Nonetheless, epiluminescence microscopy is increasingly being performed and further studies correlating dermoscopic patterns with histopathologic findings are anticipated. In addition, intraoperative dermoscopy is being employed to assist in determining margins for excision.

### Hutchinson's sign

**Key features**

- Brown–black periungual pigmentation
- Possible sign of nail melanoma
- Needs to be distinguished from pseudo-Hutchinson’s sign (dark bands producing “illusory” pigmentation of the proximal nail fold due to cuticle transparency)

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### Causes of onycholysis

<table>
<thead>
<tr>
<th>Most common etiologies</th>
<th>Clues to diagnosis</th>
<th>Less common etiologies</th>
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<tbody>
<tr>
<td>Environmental</td>
<td>History</td>
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<td>Fingernails vs. toesails</td>
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<td>Minimal, if any,</td>
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<td>nail bed changes</td>
<td>cyanacrylates</td>
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<td>Associated hemorrhage</td>
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<td>Primary skin disorders</td>
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<td>EPP</td>
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<td>Metabolic/systemic disorders</td>
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<td>Tumors</td>
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Fig. 71.6 Causes of onycholysis. *Due to secondary colonization with Pseudomonas aeruginosa. EPP, erythropoietic protoporphyria; HPV, human papillomavirus infection; NSAIDs, nonsteroidal anti-inflammatory drugs; PCT, porphyria cutanea tarda; PRP, pityriasis rubra pilaris; SCC, squamous cell carcinoma; TFTs, thyroid function tests.
First described by Hutchinson in 1886, this eponymous sign represents pigmentation of the proximal nail fold or the hyponychium in association with longitudinal melanonychia. Hutchinson's sign, in the setting of nail melanoma, is due to horizontal growth of the tumor. Although pigmentation of the proximal and lateral nail fold or hyponychium is a feature of congenital melanocytic nevus, the development in adults of longitudinal melanonychia, is due to horizontal growth of the tumor. Although pigmentation of the proximal and lateral nail fold or hyponychium is a feature of congenital melanocytic nevus, the development in adults of longitudinal melanonychia is very suggestive of melanoma.

**Green nail syndrome**

**Key features**
- Greenish-black or greenish-blue nail plate discoloration due to pyocyanin
- Predisposing factors include exposure to water, detergents and soaps
- Needs to be distinguished from a hematoma, melanocytic nevus, melanoma or an Aspergillus infection

In green nail syndrome, the nail has a greenish-black or greenish-blue color due to the deposition of pyocyanin, a blue-green pigment produced by *Pseudomonas aeruginosa*, underneath the onycholytic nail plate (see Ch. 74). It is often accompanied by paronychia. Predisposing factors include prolonged exposure to water, use of detergents and soaps, nail trauma and other causes of onycholysis. Not surprisingly, this disorder is seen in barbers, dishwashers, bakers and medical personnel. Green nail syndrome is important because affected hospital staff may spread *Pseudomonas aeruginosa* to debilitated patients. The diagnosis is usually clinical, and, if necessary, can be confirmed by Gram stain and culture of exudate and nail fragments. Treatment involves avoiding predisposing factors, wearing protective gloves, clipping back of the nail, and the use of topical solutions of 2% sodium hypochlorite or acetic acid for 1–4 months.

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**Table 71.2 Causes of longitudinal melanonychia.**

<table>
<thead>
<tr>
<th>Causes of Longitudinal Melanonychia</th>
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<tbody>
<tr>
<td><strong>Melanocyte activation</strong></td>
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<td>Trauma</td>
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<td>Nail biting/onychotillomania</td>
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<td>Drugs</td>
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<td><strong>Non-melanocytic tumors</strong></td>
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<td><strong>Nail matrix nevus</strong></td>
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<td><strong>Nail matrix melanoma</strong></td>
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**CONGENITAL AND HEREDITARY NAIL DISEASES**

The congenital and hereditary nail diseases include a number of conditions in which nail abnormalities are present at birth or develop during infancy. In some cases, the nail abnormalities are key features for the diagnosis of syndromes or hereditary diseases.

**Congenital Malalignment of the Great Toenails**

**Key features**
- Lateral deviation of the nail plate
- Often bilateral
- Predisposing factor for ingrown toenails

Congenital malalignment of the great toenails was first described by Samman in 1978 and well characterized by Baran et al. in 1979. It is common, but no prevalence data are available. Congenital malalignment of the great toenails is possibly caused by an abnormality in the ligament that connects the matrix to the periosteum of the distal phalanx. The nail plate of the hallux is laterally deviated with respect to the longitudinal axis of the distal phalanx. This results in nail matrix damage with Beau’s lines and onychomadesis [Fig. 71.8]. The nail plate...
is often thickened and transversely overcurved. Congenital malalignment of the great toenails is frequently bilateral and is the most common cause of ingrown toenails in children and adolescents. The condition often improves spontaneously.\(^{10}\)

**Congenital Hypertrophy of the Lateral Fold of the Hallux**

**Key features**
- Hypertrophic lateral nail fold
- Newborns and infants

Hypertrophic lateral nail folds in newborns were first described by Martinet et al. in 1984. The hypertropy is usually present at birth; it is characterized by an overgrowth of the soft tissue of the internal portion of the lateral nail fold, resulting in a hypertrophic lip that partially covers the nail plate. This abnormal growth may deviate the nail laterally and/or cause nail embedding with an acute inflammatory reaction and pain. The abnormality is usually bilateral and may regress spontaneously after a few years.\(^{11}\)

**Racquet Thumbs (Brachyonychia)**

**Key features**
- Congenital
- Broad and short thumbnail

Racquet thumb is a common malformation due to shortening of the distal phalanx. It is usually inherited as an autosomal dominant trait. The nail is shortened and abnormally wide. Racquet nails are usually an isolated finding and radiologic examination demonstrates a short distal phalanx.

**Nail–Patella Syndrome (Onycho-osteodysplasia; Fong Disease)**

**Key features**
- Hypoplasia of the radial side of the thumbnails
- Triangular lunulae
- Presence of bone abnormalities

Nail–patella syndrome is an autosomal dominantly inherited condition first described by Chatelain in 1820. It has been associated with mutations in the LMX1B gene, which encodes a transcription factor that regulates collagen synthesis. The condition most frequently involves the thumb and may also involve the other fingers but to a lesser extent.\(^{12}\) [Fig. 71.9]. The nails are absent or hypoplastic, and the dystrophy is usually more marked on the radial side of the digit. A triangle-shaped lunula is commonly observed. The nail changes are typically associated with bony abnormalities, including absent or hypoplastic patellae, radial head dysplasia, and iliac crest exostoses (“horns”). In children, the diagnosis is best confirmed by the presence of iliac horns on pelvic X-ray. Nephropathy develops in approximately 40% of patients. This leads to renal insufficiency in up to 8% of cases.

**Epidermolysis Bullosa**

**Key features**
- Periungual/subungual blistering
- Periungual granulation tissue
- Onycholysis
- Nail thickening and shortening
- Pterygium and nail atrophy

Nail abnormalities are common in all forms of epidermolysis bullosa (EB). Repeated blistering produces onycholysis with shortening and thickening of the nail due to nail bed scarring. Involvement of the matrix can result in nail thinning and atrophy. Dystrophic or absent nails with periungual granulation tissue are characteristic of junctional EB, Herlitz type and laryngo-onycho-cutaneous syndrome. Nail abnormalities may precede the development of skin blistering, as in “late onset” junctional EB and pretibial dominant dystrophic EB, or the nail dystrophy may be an isolated finding, as in some families with dominant dystrophic EB.

**Ectodermal Dysplasias**

**Key features**
- Abnormal development of hair, teeth and/or eccrine glands
- Nail shortening and thickening
- Nail hypoplasia

Ectodermal dysplasias were first described by Weech in 1929 and classified by Freire-Maia in 1977. Nail abnormalities are an important sign in a large number of these syndromes, where they are associated with hair, teeth and/or eccrine gland abnormalities (see Ch. 63). Most frequently, the nails are short and thickened with onycholysis. All the fingernails and toenails are usually affected.
Pachyonychia Congenita

Key features
- Thickened and extremely hard nails
- Subungual bed hyperkeratosis
- V-shaped indentations (notching) of the distal margin
- Painless keratinous cysts

Pachyonychia congenita was first described by Jadasson and Lewandowski in 1906. Pachyonychia congenita is due to mutations in keratin genes KRT6a and KRT16 in type I and KRT6b and KRT17 in type II. Nail abnormalities are present in both of the two major forms of pachyonychia congenita. The nails are thickened with an increased transverse curvature due to severe nail bed hyperkeratosis. Nail trimming is extremely difficult. Associated findings include hyperhidrosis, oral leukokeratosis, follicular hyperkeratosis and palmoplantar keratoderma, with pain on ambulation. Premature dentition and pilosebaceous cysts are observed more frequently in type II [see Table: 58.4].

Darier Disease (Follicular Dyskeratosis)

Key features
- Red and white longitudinal streaks
- V-shaped indentation (notching) of the distal margin
- Multinucleated giant cells in the nail bed epithelium

Nail abnormalities of Darier disease are common and diagnostic, and were first described by Ronchese in 1965. The nail plate has multiple red and white longitudinal streaks (Fig. 71.10). Distally, wedge-shaped subungual hyperkeratosis and fissuring of the free margin of the nail plate are also seen. The latter occurs in conjunction with the white and red bands. Histologically, there is hyperplasia of the nail bed epithelium with multinucleated epithelial giant cells, but not suprabasal clefts\(^1\). Nail changes do not improve with oral retinoid therapy. Similar nail abnormalities may be seen in patients with Hailey-Hailey disease. A single, red longitudinal band (longitudinal erythronychia) with distal subungual hyperkeratosis is not sufficient for diagnosing Darier disease of the nails, since single bands may be due to a subungual benign tumor (onychopapilloma) and, less often, Bowen’s disease\(^2\,3\). Nail abnormalities are present in up to 50% of patients with psoriasis and may be the only manifestation of the disease. Nail psoriasis is often associated with psoriatic arthritis and enthesitis\(^4\,5\). Clinical findings that are diagnostic for nail psoriasis include irregular pitting, salmon patches of the nail bed, and onycholysis with an erythematous border. These signs are often seen together in the same patient and are localized to the fingernails. Psoriatic pits are large, deep, and irregularly scattered within the nail plate (see Fig. 71.4), they may be covered by whitish, easily detachable scales. The “oil drop” sign (salmon patches) appears as an irregular area of yellow-orange discoloration visible through the nail plate (Fig. 71.11). Onycholysis surrounded by an erythematous border is also typical in nail psoriasis. Patients with psoriasis often have other nail abnormalities that are not diagnostic, as they are commonly seen in other conditions. These include splinter hemor­rhages, subungual hyperkeratosis, nail plate thickening and crumbling, and paronychia.

Diseases that should be differentiated from nail psoriasis are listed in Table 71.3.

Treatment
Nail psoriasis rarely responds to topical treatment and is often aggravated by sun exposure. Treatment measures are reviewed in the therapeutic ladder (Table 71.4).

Acrodermatitis Continua of Hallopeau
(Hallopeau’s Acrodermatitis, Dermatitis Repens)

Key features
- Recurrent, self-limited episodes of painful inflammation
- Pustules involving the nail bed and periungual skin
- Fingernails most commonly affected, often one digit

Nail abnormalities include subungual hyperkeratosis, painful transverse streaks (Fig. 71.11), and onycholysis with an erythematous border.

THE NAIL IN DERMATOLOGIC DISEASES

Several dermatologic diseases often have associated nail abnormalities that may prove helpful in confirming the clinical diagnosis.

Fig. 71.10 Darier disease. Alternating longitudinal red and white streaks with wedge-shaped subungual hyperkeratosis, V-shaped notching of the distal margin, and fissuring.

Fig. 71.11 Nail psoriasis. The nails show salmon patches (“oil drop” changes) and onycholysis with an erythematous border.
DIFFERENTIAL DIAGNOSIS OF NAIL PSORIASIS

- Onychomycosis – infection of the toenail may be difficult to distinguish from onychomycosis; hout mycologic examination
- Idiopathic onycholysis and traumatic onycholysis – area of onycholysis is not surrounded by erythema and there is an absence of subungual hyperkeratosis
- Alopeia areata – pits are small and geometrically distributed
- Acrokeratosis paraneoplastica (Bazex syndrome; paraneoplastic acrokeratosis) - the psoriasiform eruption has a typical acral distribution with involvement of the nose and ears

Table 71.3 Differential diagnosis of nail psoriasis.

DIFFERENTIAL DIAGNOSIS OF ACRODERMATITIS CONTINUA OF HALlopeau

- Acute contact dermatitis and dyshidrotic hand eczema – lesions are usually vesicular rather than pustular, with palm and/or plantar involvement
- Bacterial or viral paronychia – a relapsing course is not typical of bacterial infection; relapses are seen in HSV infection, but the nail heals completely between episodes
- Onychomycosis due to non-dermatophyte molds – may be associated with periungual or subungual inflammation and a purulent discharge. Toenails are most commonly affected and the nail plate contains fungal organisms; fungal cultures are required

Table 71.5 Differential diagnosis of acrodermatitis continua of Hallopeau. HSV, herpes simplex virus.

THERAPEUTIC LADDER FOR NAIL PSORIASIS

- Avoid trauma (3)
- Topical vitamin D3 analogues: calcipotriene (calcipotriol) – nail bed psoriasis (2)
- Topical tacrolimus 0.1% gel: nail bed psoriasis (2)
- Topical calcipotriene plus betamethasone dipropionate ointment – nail bed psoriasis (2)
- Intranasal corticosteroid (e.g. 2.5–5.0 mg/ml triamcinolone acetonide in saline) – nail matrix psoriasis (1)
- Acitretin (0.2–0.3 mg/kg/day) – nail matrix and nail bed psoriasis (1)
- Methotrexate – if indicated for additional manifestations (2)
- Cyclosporine – if indicated for additional manifestations (2)
- Targeted immunomodulators (“biologic therapeutics”) – if indicated for additional manifestations (2)

Table 71.4 Therapeutic ladder for nail psoriasis. Key to evidence-based support: (1) prospective controlled trial; (2) retrospective study or large case series; (3) small case series or individual case reports.

### Treatment

- Topical vitamin D3 analogues (calcipotriene [calcipotriol], calcitriol) or the combination of calcipotriene plus betamethasone dipropionate. Distal nail bed disease is most easily treated with a solution or lotion as the vehicle.
- Acitretin (0.3 mg/kg/day for 4–6 months) in severe cases.

### Parakeratosis Pustulosa

**Key features**

- Exclusively seen in children
- Psoriasiform lesions
- One fingernail usually affected

Parakeratosis pustulosa was first described by Sabouraud in 1931 and better characterized by Hjorth and Thomsen in 1967. It is seen exclusively in children, in whom it is usually limited to one digit, most often the thumb or the index finger. In most patients, nail changes are preceded by erythema, scaling, and vesicles of the fingertip. The affected digit shows mild psoriasiform changes with onycholysis and subungal hyperkeratosis. The nail abnormalities are usually more marked on one side of the nail. The disease usually regresses spontaneously, but some children develop psoriasis.

**Treatment**

Treatment is with emollients, topical corticosteroids and/or calcipotriene.

### Lichen Planus

**Key features**

- Longitudinal fissuring of the nail plate (onychorrhexis)
- Possible cicatricial outcome (dorsal pterygium)
- Several nails usually affected
- Histologic examination is required for diagnosis when nail involvement is isolated

Nail abnormalities are present in approximately 10% of patients with lichen planus (see Ch. 11). However, nail lichen planus is most frequently seen in the absence of skin, scalp or mucosal involvement. Clinical findings that are diagnostic include nail thinning, ridging and fissuring, and dorsal pterygium [Fig. 71.13]. These signs indicate matrix involvement and require prompt treatment to avoid scarring. Dorsal pterygium results from adhesion of the proximal nail fold to the nail bed due to matrix destruction and disappearance of the nail plate. Nail bed lichen planus produces nonspecific nail changes such as onycholysis, nail thickening and yellow discoloration.

Fig. 71.12 Acrodermatitis continua of Hallopeau. Recurrent pustular eruption of the nail bed and distal finger.

Acrodermatitis continua was first described by Crocker in 1888 and better classified by Hallopeau in 1890. Nail involvement is a typical feature of Hallopeau’s acrodermatitis, which is usually limited to one digit. It can also occur in patients with palmoplantar or generalized pustular psoriasis. Patients complain of relapsing episodes of acute painful inflammation with pustules around and under the nail plate [Fig. 71.12]. Other symptoms include onycholysis, onychomadesis, and scaling of the nail bed and perungual skin. Acrodermatitis continua of the nail is, in most cases, not associated with cutaneous plaques of psoriasis vulgaris. Proximal spread of the disease is unusual.13

The differential diagnosis of acrodermatitis continua is outlined in Table 71.5.
Fig. 71.13 Nail lichen planus. Note the nail thinning with longitudinal ridging and fissuring.

**DIFFERENTIAL DIAGNOSIS OF NAIL LICHEN PLANUS**

- **Systemic amyloidosis** – thinning and fissuring are associated with splinter hemorrhages; histologic examination demonstrates amyloid deposits within the nail matrix and nail bed dermis
- **Lichen striatus** – the lichenoid nail changes are restricted to one or two digits and limited to one side of the nail plate
- **Dyshidrosis congenita** – lichenoid nail changes are associated with oral leukoplakia and reticulated hyperpigmentation
- **Nail pterygium due to bullous disease** – the history differs and there are often associated cutaneous and/or mucosal lesions
- **Nail pterygium due to digital ischemia** – the digits are typically cold and there is a history of Raynaud’s phenomenon
- **Graft-versus-host disease** – similar lichenoid nail changes
- **Additional entities** – psoriasis, onychomycosis and the yellow nail syndrome

**Table 71.6 Differential diagnosis of nail lichen planus.**

When isolated, this diagnosis may be difficult to establish and it requires histologic evaluation. The differential diagnosis of nail lichen planus is outlined in Table 71.6.

**Treatment**

Treatment with systemic therapies may be required to avoid pterygium formation (see Ch. 11). IntraleSIONAL injections of corticosteroids (2.5–5.0 mg/ml triamcinolone acetonide in saline) may be utilized when the disease involves a few nails.

**Trachyonychia (Twenty-Nail Dystrophy, Sandpapered Nails)**

**Key features**

- Nail roughness due to excessive longitudinal ridging
- Several nails are usually affected
- More common in children
- Often associated with alopecia areata
- Benign outcome

Trachyonychia was first described by Allkiewiez in 1950. Identical nail abnormalities were given the name “twenty-nail dystrophy” by Hazeldigg et al. in 1977. Twenty-nail dystrophy or trachyonychia describes a spectrum of nail plate surface abnormalities that produce nail roughness. Since the nail changes do not always involve all 20 nails, the term “trachyonychia” (from the Greek word meaning rough) is preferred over twenty-nail dystrophy by most authors. It may occur in association with alopecia areata (12% of children and 3% of adults with severe alopecia areata) or it may be idiopathic. The latter is more common in children.

The nails are thin, opaque and lusterless and give the impression of having been sandpapered in a longitudinal direction (i.e. vertically striated sandpapered nails) (see Fig. 71.5). The cuticles are often hyperkeratotic. Trachyonychia is fairly asymptomatic and patients complain only of brittleness and the cosmetic appearance. In addition to alopecia areata, this finding is occasionally caused by several inflammatory diseases that mildly disturb nail matrix keratinization, including lichen planus, eczema and psoriasis. Determination of the underlying inflammatory disease responsible for the trachyonychia requires a nail biopsy. However, this is generally not recommended because of the benign nature of the disease and trachyonychia improves spontaneously in the majority of patients.

When used to treat hair and cutaneous manifestations, systemic corticosteroids and systemic retinoids can also improve trachyonychia in patients with alopecia areata and psoriasis, respectively.

**Alopecia Areata**

**Key features**

- Geometric pitting
- Children are most commonly affected

Nail abnormalities are present in approximately 20% of adults and 50% of children with alopecia areata. Signs that are characteristic of nail alopecia areata include geometric pitting and trachyonychia. The pits are small, superficial and regularly distributed in a geometric pattern (grid-like). Additional nail abnormalities observed in alopecia areata include punctate leukonychia, erythema of the lunula, and onychomadesis. Trachyonychia is more common in children and most frequently seen in male patients with alopecia totalis or universalis.

**Eczema (Dermatitis)**

**Key features**

- Irregular pitting and Beau’s lines
- Subungual hyperkeratosis
- Chronic paronychia

Hand eczema is often associated with nail changes. In acute eczema, there are vesicles and erythema of the proximal nail fold and the hyponychium. Nail matrix damage produces irregular pitting and Beau’s lines; onychomadesis can occur in severe cases. Chronic eczema frequently localizes to the hyponychium, resulting in subungual hyperkeratosis, onycholysis and fissuring of the hyponychium. Chronic eczema of the proximal nail fold can lead to chronic paronychia. In atopic dermatitis, the nail plate frequently shows mild superficial abnormalities such as irregular pits and Beau’s lines. Controlling the skin disease results in gradual improvement of the nails.

**THE NAIL IN SYSTEMIC DISEASES**

Abnormalities of the nails have been anecdotally reported in a number of systemic diseases, but most of these abnormalities are nonspecific. In this chapter, we will discuss only those nail changes that are useful in the diagnosis of systemic disorders.

**The Nail Plate in Forensic Medicine**

**Key features**

- Drug exposure
- Poisoning
- Genetic analysis
The slow growth rate of nails makes it possible to utilize nail clippings to detect previous exposure to metals or toxins (e.g., heroin). Compounds that can be measured in the nails include heavy metals and drugs.

**Clubbing (Hippocratic Fingers, Watch-Glass Nails, Drumstick Fingers)**

**Key features**
- Bulbous digits
- Watch-glass nails
- Greater than 180° angle between the proximal nail fold and nail plate (Lovibond’s sign)

First described by Hippocrates in the first century BC, clubbing may be congenital or acquired. Acquired clubbing is uncommon and in 80% of cases is associated with pulmonary diseases.

Clubbing is caused by enlargement of the soft tissue of the distal digit. The nail plate is enlarged and excessively curved, with a greater than 180° widening of the angle between the proximal nail fold and the nail plate (see Fig. 71.2). In hypertrophic osteoarthropathy, clubbing is associated with hypertrophy of the extremities and painful pseudo-inflammatory joint disease. Systemic conditions associated with clubbing are outlined in Table 71.7.

**Yellow Nail Syndrome**

**Key features**
- Arrest in nail growth
- Yellow-green nails
- Absence of the cuticle
- Overcurvature and thickening
- All or most nails are affected

The yellow nail syndrome was first described by Samman and White in 1964. It is not common and its pathogenesis is still unknown. Linear nail growth is arrested or greatly reduced. The nails are thickened and transversely and longitudinally overcurved with disappearance of the cuticle. The nail color varies from pale yellow to dark yellow-green (Fig. 71.14). Onycholysis is frequently seen, as is nail plate shedding. In most patients, all 20 nails are involved. Characteristically, the nail abnormalities are associated with lymphedema and respiratory tract involvement, including chronic bronchitis, bronchiectasis, sinusitis and pleural effusions. The diagnosis of yellow nail syndrome, however, requires only the presence of the typical nail changes.

**Treatment**

Treatment is not effective in all cases and must be prescribed for several months:
- Vitamin E 1200 IU/day
- Pulse itraconazole (400 mg daily for 1 week a month) or fluconazole (150 mg daily for 1 week a month); theoretically, the drugs act by accelerating nail growth.

**Apparent Leukonychia**

**Key features**
- White discoloration that fades with pressure
- Nail plate transparency is maintained

On the basis of the pattern of the nail discoloration, three different types of apparent leukonychia can be distinguished. Nail bed edema may be a common factor.

### Table 71.7 Systemic conditions associated with clubbing.

<table>
<thead>
<tr>
<th>Systemic Conditions Associated with Clubbing</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>CONGENITAL/GENETIC</strong></td>
</tr>
<tr>
<td>Cardiovascular disorders</td>
</tr>
<tr>
<td>Congenital heart disease (usually cyanotic)</td>
</tr>
<tr>
<td>Pulmonary arteriovenous malformations</td>
</tr>
<tr>
<td>(often in the setting of hereditary hemorhagic telangiectasia)</td>
</tr>
<tr>
<td>Bronchopulmonary disease</td>
</tr>
<tr>
<td>Cystic fibrosis</td>
</tr>
<tr>
<td>Other</td>
</tr>
<tr>
<td>Pachydermoperiostosis (see Ch. 98)</td>
</tr>
<tr>
<td><strong>ACQUIRED</strong></td>
</tr>
<tr>
<td>Bronchopulmonary diseases</td>
</tr>
<tr>
<td>Neoplasms (primary or metastatic cancers, pleural tumors)</td>
</tr>
<tr>
<td>Chronic infections (abscesses of the lungs, tuberculosi)</td>
</tr>
<tr>
<td>Bronchiectasis</td>
</tr>
<tr>
<td>Pulmonary fibrosis, sarcoidosis</td>
</tr>
<tr>
<td>Cardiovascular diseases</td>
</tr>
<tr>
<td>Aneurysm or dialysis fistula (unilateral clubbing); congestive cardiac failure; bacterial endocarditis</td>
</tr>
<tr>
<td>Gastrointestinal diseases</td>
</tr>
<tr>
<td>Inflammatory bowel disease</td>
</tr>
<tr>
<td>Carcinomas</td>
</tr>
<tr>
<td>Infestations, e.g. amebiasis, ascarisis</td>
</tr>
<tr>
<td>Liver disorders, e.g. chronic active hepatitis, cirrhosis</td>
</tr>
<tr>
<td>Infection</td>
</tr>
<tr>
<td>HIV infection</td>
</tr>
<tr>
<td>Arterial graft sepsis (clubbing limited to perfused extremities)</td>
</tr>
<tr>
<td>Endocrine diseases</td>
</tr>
<tr>
<td>Thyroid disease (primarily hyperthyroidism)*</td>
</tr>
<tr>
<td>Secondary hyperparathyroidism</td>
</tr>
<tr>
<td>Other</td>
</tr>
<tr>
<td>POEMS syndrome</td>
</tr>
<tr>
<td>Laxative abuse</td>
</tr>
<tr>
<td>Hemiplegia (unilateral)</td>
</tr>
<tr>
<td>Trauma, median nerve injury (unidigital)</td>
</tr>
</tbody>
</table>

*Most often in the setting of thyroid acropachy (clubbing together with digital swelling and persontal reaction) in patients with autoimmune thyroid disease.

**Fig. 71.14 Yellow nail syndrome.** Note the overcurvature of the nail and loss of the cuticle, in addition to the yellow discoloration.

- Terry’s nails were described by Terry in 1954 as a common sign of liver cirrhosis, occurring in up to 80% of patients. The leukonychia affects the whole nail except for a 1 to 2 mm distal band. Terry’s nails are also frequently seen in normal individuals.
- Muchcrck’s nails were described by Muchcrck in 1956 in patients with hypoaluminemia (nephrotic syndrome). They are a very common finding in patients receiving combination chemotherapy. The nail has multiple transverse whitish bands, parallel to the lunula (Fig. 71.15).
Hair, Nails and Mucous Membranes

Pseudo-inflammatory Nail Changes

Key features
- Cold painful whitlow: digital ischemia
- Painless whitlow and normal temperature: bone metastases

The distal portion of the digit is greatly enlarged and red, simulating an acute paronychia, but the skin temperature is cold or normal. This picture may be caused by digital ischemia or metastases to the distal phalanges. In digital ischemia, the digit is cold and painful. The most common causes include arterial obstruction and autonomic neuropathies causing ischemia (including diabetes). Digital metastases are usually painless, metastases in the fingers are most commonly associated with pulmonary neoplasms and those in the toes with genitourinary neoplasms. X-ray examination is required for the diagnosis19.

Peripheral Neuropathies

Key features
- Nail abnormalities limited to the first three digits: carpal tunnel syndrome
- Onychomadesis with pseudopyogenic granuloma: mild peripheral nerve injury

Examination of the nail may suggest the diagnosis in carpal tunnel syndrome, since nail abnormalities are limited to the first three fingers and are associated with fingertip paresthesias. Depending on the severity of nerve damage, nail changes vary from Beau’s lines to onychomadesis and necrosis. Neurologic examination and electromyography confirm the diagnosis.

Onychomadesis associated with pseudopyogenic granuloma of the proximal nail fold is a sign of mild and transitory peripheral nerve injury. It is typically seen in patients with phalanx, metacarpal or wrist bone fractures during or after cast immobilization. Patients often report a history of pain or paresthesias of the hand during the period of immobilization. The condition spontaneously resolves over a few weeks20.

Autoimmune Connective Tissue Disorders

Key features
- Nail fold capillary abnormalities
- Cuticular hemorrhages
- Ventral pterygium
- Nail beaking

Abnormalities in the proximal nail fold capillaries are frequently seen in autoimmune connective tissue disorders, as they occur in most patients21. Roughness, hemorrhages and necrosis of the cuticles are common in dermatomyositis and scleroderma, where capillaroscopy shows reduced capillary density and avascular areas alternating with dilated capillary loops. In systemic lupus erythematosus, capillaroscopy shows a normal capillary density with dilated tortuous capillaries.

Ventral pterygium (pterygium inversus unguis) is characterized by the adhesion of the distal nail plate to the hypnonychium, resulting in pain during nail trimming. Ventral pterygium is a distinctive sign of scleroderma related to impaired peripheral perfusion22. In scleroderma, ischemic changes and bone resorption may result in nail beaking with bending of the nail plate around the shortened fingertip (see Fig. 43.2).

HIV Infection

Key features
- Proximal subungual onychomycosis due to Trichophyton rubrum
- Candida onychomycosis
- Longitudinal melanonychia
- Squamous cell carcinoma

Onychomycoses are common in HIV-infected patients, occurring in up to 25% of affected individuals. Dermatophytes are most commonly responsible, but Candida spp. and molds are also often isolated23. Although a large number of nail abnormalities have been reported in association with AIDS24, only a few clinical entities can be considered typical of this disease.

- Proximal subungual onychomycosis due to T. rubrum. This variety of onychomycosis is considered a marker for HIV infection and immunodeficiency. The proximal nail plate is opaque and white due to the presence of fungi in its ventral portion.
- Candida onychomycosis. Candida does not invade the nail plate of immunocompetent individuals, and the diagnosis of true Candida onychomycosis indicates immunosuppression – often, HIV infection.
- Longitudinal melanonychia. Usually, several nails are involved, and it is often associated with skin hyperpigmentation.
- HPV-induced squamous cell carcinoma. Longstanding perungal warts in HIV patients should always raise the suspicion of squamous cell carcinoma and be examined histologically. HPV types 16 and 35 have been detected in such lesions25.

Drug-Induced Nail Abnormalities (Table 71.8)

Key features
- Involvement of most or all nails
- Beau’s lines and onychomadesis
- Painful hemorrhagic onycholysis
- Multiple bands of melanonychia
- Paronychia with pyogenic granulomas

Drug-induced nail changes usually involve several or all nails. The pathogenesis is most often toxic in nature26.
Cancer chemotherapeutic agents, due to their cytotoxic properties, are the most frequent cause of nail changes. These include Beau’s lines, onychomadesis, fragility, pigmentation, onycholysis, paronychia, and vascular problems, including subungual hemorhages, hematomas and ischemia. Painful subungual hemorrhages and abscesses are typically seen with taxanes [paclitaxel and docetaxel].

Paronychia associated with pyogenic granulomas has been reported with epidermal growth factor receptor (EGFR) inhibitors used as chemotherapeutic agents [see Ch. 21]. Nail changes appear from 1 to 3 months after starting treatment and disappear after interruption of treatment.

Cyclic retinoids are particularly nail fragility, paronychia and pyogenic granulomas. Antiretroviral drugs often cause nail pigmentation, paronychia and pyogenic granulomas. Oral retinoids are frequently responsible for nail abnormalities, especially nail fragility, paronychia and pyogenic granulomas (Fig. 71.16). Although uncommon, photo-onycholysis is a characteristic side effect of tetracyclines, taxanes, psoralesns and photodynamic therapy (see Fig. 71.6).

Certain drugs (e.g. anticoagulants and anticonvulsants) taken during pregnancy may impair the development of the digits, leading to congenital nail hypoplasia.

### Table 71.8 Drug-induced nail abnormalities.

<table>
<thead>
<tr>
<th>Nail abnormality</th>
<th>Responsible agents</th>
</tr>
</thead>
<tbody>
<tr>
<td>Beau's line and onychomadesis</td>
<td>Chemotherapeutic agents</td>
</tr>
<tr>
<td>True leukonychia</td>
<td>Chemotherapeutic agents</td>
</tr>
<tr>
<td>Nail thinning and brittleness</td>
<td>Chemotherapeutic agents, Retinoids</td>
</tr>
<tr>
<td>Onycholysis/photo-onycholysis</td>
<td>Chemotherapeutic agents, particularly taxanes Tetracyclines Psoralesns NSAIDs</td>
</tr>
<tr>
<td>Apparent leukonychia (e.g. Muehrcke's nails)</td>
<td>Chemotherapeutic agents, particularly polychemotherapy including anthracyclines, vincristine</td>
</tr>
<tr>
<td>Melanonychia</td>
<td>Chemotherapeutic agents Psoralesns Zidovudine (AZT)</td>
</tr>
<tr>
<td>Discoloration (non-melanin)</td>
<td>Minocycline Antimalarials Gold</td>
</tr>
<tr>
<td>Paronychia and periungual pyogenic granulomas</td>
<td>Retinoids Antitretroviral drugs (indinavir, efavirenz, lamivudine) Tetracyclines Psoralesns Methotrexate Sirolimus</td>
</tr>
<tr>
<td>Ischemic changes</td>
<td>Beta-blockers Bleomycin</td>
</tr>
</tbody>
</table>

### Warts [see Ch. 79]

**Key features**
- Periungual keratotic papules
- Hyperkeratosiis of the cuticle
- Onycholysis and subungual hyperkeratosis
- In recalcitrant lesions, consider the possibility of squamous cell carcinoma

Periungual warts are common in nail biters, in whom they are multiple and involve several nails. The warts appear as keratotic papules, and when localized to the proximal nail fold, they frequently produce periungual hyperkeratosis simulating a hyperkeratotic cuticle. Nail bed warts lead to uplifting of the nail plate with onycholysis. Squamous cell
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**Carcinoma**

Carcinoma can arise in or mimic a verruca; therefore, suspicious lesions must be further evaluated.

**Treatment**

See therapeutic ladder (Table 71.9).

**Onychomycosis (Tinea Unguim)**

See Chapter 77.

**ENVIRONMENTAL NAIL DISORDERS**

**Brittle Nails (Fragility, Onychoschizia)**

- **Key features**
  - Lamellar exfoliation (onychoschizia)
  - Fingernails affected
  - Nail dehydration due to environmental factors

Brittle nails are very common and usually affect women. Nail brittleness results from dehydration of the nail plate as a result of environmental factors such as frequent handwashing. In lamellar onychoschizia, the distal nail plate splits horizontally into multiple layers (see Fig. 71.2). Other signs of nail brittleness include splitting, softening and onychorhrosis.

**Treatment**

- Avoidance of water and chemical exposure.
- Biotin [2.5–5 mg/day].
- Topical moisturizers and humectants.

**Chronic Paronychia**

- **Key features**
  - Proximal nail fold inflammation
  - Absence of the cuticle
  - Fingernails affected
  - Nail plate surface abnormalities

Chronic paronychia commonly involves the fingernails of adult women. Although the pathogenesis is still debated, there is accumulating evidence that the condition represents a contact reaction to irritants or allergens. Occupational chronic paronychia is common in food handlers. Chronic paronychia is clinically characterized by inflammation of the proximal nail fold with erythema, edema and absence of the cuticle. One or several fingernails (especially the thumb and second or third fingers of the dominant hand) are affected. Damage to the nail matrix results in nail plate surface abnormalities such as Beau's lines. Chronic paronychia usually assumes a prolonged course with superimposed, recurrent, self-limited episodes of acute exacerbation. Secondary infections with *Candida* spp. and *Pseudomonas aeruginosa* are common.

**Treatment**

- Avoidance of water and chemical exposure.
- Topical corticosteroids.
- Topical imidazoles.
- Topical antifungals such as moominate [e.g. 4% thymol in 95% ethanol].
- Systemic antifungals are not useful.

**Idiopathic Onycholysis**

- **Key features**
  - Limited to fingernails
  - White or green discoloration
  - Absence of subungual hyperkeratosis

Idiopathic onycholysis frequently involves the fingernails as a consequence of repetitive water immersion and exposure to irritants (see Fig. 71.6). The affected nail is detached from the nail bed and often shows an abnormal color due to secondary microbial contamination of the subungual space.

**Treatment**

- Avoidance of water and chemical exposure.
- Clipping of the detached nail plate.
- Topical antifungals.

**TRAUMATIC NAIL ABNORMALITIES** (Table 71.10)

**Onychotillomania**

- **Key features**
  - Cuticle absent and proximal nail fold inflamed
  - Nail plate surface abnormalities, e.g. longitudinal central depression
  - Melanonychia
  - Hemorrhages and crusts

Self-induced nail abnormalities are common, but often not recognized due to the wide variety of nail signs and the difficulty in obtaining a correct clinical history.

- Nail biting may affect the nail plate and/or the proximal nail fold; the trauma often produces nail matrix damage with secondary nail plate abnormalities, including surface irregularities and longitudinal melanonychia.
- Habit-tic deformity affects the thumb and is due to the nervous habit of rubbing and pushing back the mid-portion of the cuticle of the thumb with the index finger. The nail plate of the thumb shows multiple midline Beau's lines with a prominent longitudinal central depression. Median caliliform dystrophy of Heller, also known as dystrophia unguis mediana caliliformis, is characterized by an inverted fir tree-like split or caliliform defect in the nail plate (Fig. 71.18) and is possibly a variant of habit-tic deformity.
- Nail destruction associated with psychiatric disorders. The clinical picture is variable, with nail plate destruction, hemorrhages, and periungual crusts and erosions. Instruments (scissors, clippers, etc.) are often utilized to destroy the nails.
Nail changes associated with a manicure or pedicure.

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Table 71.10 Nail changes associated with a manicure or pedicure.

Acute hematomas occur after trauma severe enough to lead to accumulation of blood under the nail plate. Toenails are most commonly affected. Acute hematomas are typically associated with pain, and compression of the matrix may cause secondary nail plate dystrophy. An X-ray of the digit may be advisable to exclude a bone fracture.

Repeated microtrauma or hemorrhagic conditions produce asymptomatic small subungual hematomas of the nails that are incorporated within the ventral nail plate and migrate distally with nail growth. The color of subungual hematomas ranges from purple–red to black. Dermoscopy is useful for differentiating hematomas from melanin deposition.

**Treatment**

Acute hematomas require drainage of the blood by creating a hole through the nail plate.

**Traumatic Toenail Abnormalities**

**Onychogryphosis**

- History of acute trauma
- Purple–red to black in color
- Typical dermatoscopic features
- Discoloration moves distally with nail growth

Onychogryphosis is common in elderly persons and almost exclusively affects the toenails, usually the hallux. The nail acquires a typical ram’s horn shape due to asymmetric growth. The nail plate is thick, hard and yellow–brown. Multiple transverse striations are often present. Nail hardness and self-neglect may lead to monstrous deformities.

**Pincer Nails (Trumpet Nails)**

- Nail bed pinching by overcurved nail plate
- Toenails most frequently affected
- Severe pain
- Subungual exostosis should be excluded by X-ray examination

Traumatic toenail abnormalities are most commonly seen in women who wear high-heeled pointed shoes.

- Traumatic onycholysis of the hallux is the most common clinical presentation. The onycholysis may be distal, where the detachment is caused by compression of the distal pulp by poorly fitting shoes, or lateral, when there is overlapping of the second toe onto the first. Clipping of the detached nail plate reveals a normal nail bed. The differential diagnosis includes onychomycosis, but here the onycholysis is associated with subungual hyperkeratosis.
- Transverse leukonychia of the hallux results from repeated microtrauma by shoes to untrimmed, long, great toenails. Multiple bands of true leukonychia move distally with nail growth.
- Frictional melanonychia affects the toenails of the fourth and/or fifth digits of women. It is due to activation of nail matrix melanocytes by friction from shoes or from the adjacent digit. The bands may be multiple and black in color.
- Retroneychia is caused by embedding of the nail into the proximal nail fold following trauma and may present with inflammation of the proximal nail fold.
Pincer nails are usually seen on the toes and may be hereditary or acquired; the latter is more commonly due to ill-fitting shoes. The nail plate displays an excessive transverse overcurvature, especially in its distal portion (see Fig. 71.2). This results in compression of the distal nail bed with severe pain.

**Treatment**

Surgical correction is necessary when a subungual exostosis is present.

**Ingrown Toenails (Onychocryptosis)**

Lateral ingrowing usually affects the hallux of young adults with congenital malalignment of the great toenails. Precipitating factors include improper or aggressive nail cutting and trauma. Penetration of nail plate spicules into the lateral nail fold epithelium causes painful inflammation. Chronic onychocryptosis is characterized by the growth of granulation tissue that eventually undergoes epithelialization.

Distal embedding is a common complication of nail avulsion. The nail plate growth is blocked by the hyponychium, which forms a distal rim. Retronychia represents the ingrowth of the proximal nail plate into the proximal nail fold, with one to three nail plates misaligned beneath the uppermost nail plate. There may be associated proximal periungual pyogenic granulomas.

**Key features**
- Teensagers and young adults
- Congenital malalignment often present
- Precipitated by improper nail cutting and hyperhidrosis
- Painful inflammation of the lateral fold
- Growth of granulation tissue

**Treatment**

- Prevention through patient education.
- Removal of the embedded spicule.
- Uplifting of the lateral nail plate with cotton or dental floss.
- Granulation tissue may be prevented by topical antibiotics and topical corticosteroids or treated by cryotherapy, chemical cautery or electrodesiccation.
- Chemical (88% phenol), laser or excisional surgical removal of the lateral matrix is advisable in severe cases.
- In retronychia, surgical avulsion of the proximally embedded nail plate as well as the underlying plate[s] is necessary.

**NAIL TUMORS**

Nail surgery is discussed in Chapter 149.

**Benign Tumors**

**Pyogenic granuloma (botryomycoma)**

- **Key features**
  - Bleeding angiomatous papulonodule
  - Periungual or subungual
  - Often traumatic

Pyogenic granulomas commonly appear within the nail apparatus, where they may be periungual or subungual, and often follow penetrating trauma. Other common causes of pyogenic granuloma of the nail include ingrown toenails, systemic drugs (e.g. retinoids, EGFR inhibitors), peripheral nerve damage, and frictional onycholysis due to prolonged walking. The tumor appears as a bleeding, friable, soft red papulonodule (see Fig. 71.16). When subungual, it is associated with onycholysis. The differential diagnosis includes amelanotic melanoma. Treatment is surgical.

**Fibromas/Fibrokeratomas**

- **Key features**
  - Filiform growth
  - Nail plate furrow
  - Possible sign of tuberous sclerosis

Isolated periungual and subungual fibromas are not rare in the general population. Multiple lesions occur in 5% of patients with tuberous sclerosis (Koenen’s tumors). Periungual fibromas appear as pink or skin-colored papules originating from the proximal nail fold. The fibroma may compress the nail matrix and produce a longitudinal groove in the nail plate. Subungual fibromas that grow underneath the nail plate produce longitudinal erythronychia or onycholysis. A fibrokeratoma is characterized by a hyperkeratotic tip and may be surrounded by a collarette of raised skin.

**Subungual exostosis**

- **Key features**
  - Subungual hard nodule
  - Onycholysis
  - Toenails (especially hallux)
  - X-ray examination is diagnostic

Subungual exostoses, first described by Dupuytren in 1847, are the most common benign bony proliferations associated with nail abnormalities. Subungual exostoses are commonly precipitated by trauma and usually seen on the great toe of young patients (Fig. 71.19A). An exostosis produces a firm, tender subungual nodule that elevates the nail plate. The nodule may ulcerate or become hyperkeratotic. The diagnosis is confirmed by X-ray examination (Fig. 71.19B).

**Myxoid cysts (mucous cyst)**

- **Key features**
  - Proximal nail fold swelling
  - Nail plate depression and grooves
  - Periodic fluid drainage

First described by Hide in 1883, myxoid cysts are the most common nail tumor, often occurring in middle-aged women. Typically located in the proximal nail fold of the fingernails, they appear as small soft nodules that often spontaneously drain a viscous jelly-like fluid. Compression of the matrix produces nail plate depression and grooves (Fig. 71.20). Occasionally, the cysts are subungual. These cysts are connected to the distal interphalangeal joint by a tract, and osteoarthrits of the distal joint is a frequent association.

**Treatment**

Possible treatments for myxoid cysts include sclerotherapy, cryosurgery, and intralesional corticosteroid injections. Caution must be taken because septic arthritis is a potential complication of surgical procedures that allow access of surface bacteria into the underlying synovial space. All of these procedures are associated with a high frequency of relapses. Definitive cure of myxoid cysts can be obtained with surgical procedures that include ligation of the pedicle that connects the cyst to the joint.
Nail Disorders

Fig. 71.19 Subungual exostosis. A Skin-colored subungual nodule elevating the nail plate. B Radiograph of the digit demonstrating the subungual bony proliferation.

Fig. 71.20 Myxoid cyst. The longitudinal nail groove is a result of the compression of the nail matrix by the cyst.

Fig. 71.21 Onychomatricoma. The nail is thickened and overcurved with longitudinal yellow streaks. The distal border shows several holes.

Glomus tumor

Key features
- Severe pain
- Temperature sensitivity
- Subjective symptoms contrast with minimal clinical signs

First described by Wood in 1812, a glomus tumor arises from the neuro-myrioarterial glomus cells of the nail bed dermis. It is associated with severe pain that radiates proximally and is often aggravated by physical or thermal stimuli (especially cold). Subjective symptoms typically exceed clinical signs. The tumor appears as a red–bluish nail bed macule visible through the transparent nail plate (see Ch. 149). MRI allows the diagnosis in doubtful cases (see Ch. 114).

Onychomatricoma

Key features
- Localized longitudinal thickening with transverse overcurvature
- Yellow–white discoloration
- Multiple, longitudinal, tumor-containing hollows
- Holes in the distal nail

First described by Baran and Kint in 1992, onychomatricoma is a rare neoplasm that produces localized or diffuse thickening of the nail plate with perforations consisting of multiple longitudinal hollows that contain the digiting tumor. Most commonly, onychomatricoma arises within the fingernails of middle-aged individuals (Fig. 71.21). A frontal view of the nail typically reveals multiple holes in the thickened free margin. The affected nail is thickened and yellow–white in color, with multiple splinter hemorrhages. Histologically, the tumor is characterized by multiple fibroepithelial projections that extend into the thickened nail plate. The tumor epithelium is identical to that of the normal nail matrix and keratinizes without a granular layer.

Melanocytic nevi of the nail matrix

Key features
- Longitudinal melanonychia
- Onset in childhood

Nail matrix nevi are an uncommon cause of longitudinal melanonychia, especially in comparison with melanocyte activation (see Table 71.2). They usually develop during childhood and often involve the fingers, especially the thumb. The color, width and pigment
distribution may vary considerably and it is not unusual to observe fading or darkening of the pigmentation over time. Pigmentation of the periungual tissues is also possible.

**Treatment**

Optimal management of nail matrix nevi is still debated. The authors’ approach is immediate excision of rapidly growing lesions and excision of lesions after puberty [see Ch. 149].

**Malignant Tumors**

Malignant tumors of the nail are summarized below. For further details of the disorders and their treatment, see Chapters 108 and 113.

**Bowen's disease (squamous cell carcinoma in situ)**

**Key features**
- Verrucous lesion
- Onycholysis
- Melanonychia

Bowen's disease of the nail is uncommon and is seen most often in middle-aged men. Fingers of the left hand are most commonly involved. Clinically, it may be difficult to differentiate Bowen's disease from warts. The affected digit shows periungual or subungual verrucous lesions with onycholysis and longitudinal melanonychia (Fig. 71.22). Predisposing factors include HPV infection and chronic X-ray exposure.

**Keratoacanthoma**

**Key features**
- Painful subungual nodule
- Rapid growth
- Osteolysis on radiography

Keratoacanthoma is an extremely rare tumor that usually affects the thumb or the index or middle fingers. It appears as a painful subungual keratotic nodule that grows rapidly over a period of weeks. Deep invasion with bone destruction is frequent. A history of trauma is often reported. In contrast to cutaneous keratoacanthomas, keratoacanthomas of the nail do not regress spontaneously.

**Squamous cell carcinoma**

**Key features**
- Verrucous lesion
- Ulcerated nodule
- Onycholysis
- Nail plate destruction

Squamous cell carcinoma is the most frequent malignant tumor of the nail apparatus. It most commonly affects the fingernails of middle-aged men. A number of studies have confirmed a causative role for HPV, mostly HPV-16, in the development of this tumor. Clinically, there is a slowly growing periungual or subungual mass that may ulcerate and bleed. Periungual swelling and inflammation are often seen. Bony involvement may occur but metastases are extremely rare.

**Verrucous carcinoma (carcinoma cuniculatum, epithelioma cuniculatum)**

**Key features**
- Rapidly growing verrucous nodule
- Nail destruction

Verrucous carcinoma is a rare, low-grade variant of squamous cell carcinoma characterized by locally aggressive clinical behavior but low potential for metastasis. It rarely involves the nail apparatus. Clinically, the tumor appears as a rapidly growing nodule that often destroys the nail. Bone resorption is common.

**Melanoma**

**Key features**
- Longitudinal melanonychia
- Thumb is the most frequent site
- Nail plate destruction
- Hutchinson's sign
- Amelanotic in 25% of cases

Nail melanoma is rare, accounting for 0.7–3.5% of all melanomas. It most frequently involves the thumb of middle-aged individuals, and patients often report a history of trauma. Diagnosis is often delayed, and in 25% of cases the tumor is amelanotic. The 5-year survival is only 15%.

From a clinical standpoint, nail melanoma may have the following features:
- Longitudinal melanonychia. The pigmented band is usually dark brown or black in color, with blurred margins. Nail plate abnormalities result from nail matrix damage. The dermoscopic


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18. McGonagle D. Enthesitis: an autoimmune inflammatory lesion linking nail and joint involvement in psoriatic disease. J Eur Acad Dermatol Venereol. 2009;23(Suppl 1):9–13. pattern that has been associated with melanoma is a brown background with longitudinal lines that are irregular in color, thickness and spacing. However, a recent study indicated that these dermoscopic patterns were not useful in the early detection of nail melanoma, as dermoscopy did not improve accuracy in the clinical diagnosis of melanoma39.

- A subungal pigmented lesion that may have ulcerated and is associated with onycholysis and nail destruction.
- An amelanotic nodule that frequently ulcerates and bleeds, resembling a pyogenic granuloma (Fig. 71.23). The presence of Hutchinson’s sign [pigmentation of the periungual tissues due to superficial spreading of the tumor] should raise the suspicion of nail melanoma. An ABCDEF of nail melanoma has been proposed, to facilitate diagnosis (Table 71.11)40.