



What's New in Pediatric Nail Disorders?

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Abbreviations

HFMD	Hand, foot and mouth disease
LM	Longitudinal melanonychia
NAPSI	Nail Psoriasis Severity Index
NMN	Nail matrix nevus
NPS	Nail-patella syndrome
PASI	Psoriasis Area Severity Index
PC	Pachyonychia congenita
SE	Subungual exostosis
YNS	Yellow nail syndrome

Introduction

Nail disorders in children can be classified into seven categories: physiologic, congenital and/or hereditary, infectious, inflammatory, traumatic or mechanical, neoplastic, systemic, and/or iatrogenic [1]. Most pediatric nail conditions are benign but are a source of anxiety for parents or physicians inexperienced in onychology and may be a cosmetic issue or a cause of functional impairment [1, 2]. To date, very few stud-

ies have been conducted to determine the overall prevalence of nail disorders in children [3–5]. This prevalence has been estimated at 11.1% and 6.8% in two studies [3, 4]. A very recent study examined the span of nail conditions in a pediatric onychology center and highlighted the ten most frequent diagnoses (Table 14.1) [6].

This study also demonstrated that some diagnoses were more frequently encountered depending on the age category (Table 14.2). Another publication has classified nail diseases in children according to age [7].

This chapter collects the recent findings and updates found in the literature on children's nail disorders. This review includes the most relevant articles from the last 5 years. Search engines were Google Scholar and the Medline database through PubMed and MeSH. Keywords included "nails," "children," "infants," and "teenagers," as well as specific keywords for many nail disorders.

Table 14.1 Ten most frequent diagnoses in a pediatric nail consultation [6]

1. Fever-related Beau's lines or onychomadesis
2. Trachyonychia
3. Longitudinal melanonychia
4. Congenital malalignment of the great toenail
5. Podiatric-related chronic trauma
6. Onychomycosis
7. Acute trauma and <i>sequelae</i>
8. Congenital hypertrophy of the lateral nail folds
9. Acute paronychia
10. Juvenile ingrown nails

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Table 14.2 Most frequent diagnoses according to age [6]

0 to <2 years old	Congenital hypertrophy of the lateral nail Congenital malalignment of the great toenail
2 to <6 years old	Fever-related Beau's lines
6 to <12 years old	Trachyonychia Longitudinal melanonychia
12 to <18 years old	Juvenile ingrown nails Acute trauma and <i>sequelae</i> Podiatric-related chronic trauma Longitudinal melanonychia

Physiologic Alterations

Very few papers have described physiologic nail alterations in children, especially in newborns. A recent study from Chinazzo et al. described the normal nail features in newborns. It showed that koilonychia, onycholysis, and onychoschizia were frequent findings on toenails, while the absence of lunula was a usual feature of both finger- and toenails. The authors also reported an apparent hypertrophy of the proximal nail fold of the hallux in 1/3 of the toddlers and of the lateral folds of the hallux in 3/4 of the newborns. This hypertrophy was accentuated by koilonychia and a triangular shape of the nail plate. All dark-skinned newborns showed hyperpigmentation of the proximal fold of both hands and feet, especially between 2 and 6 months, that faded before the age of 1 [8].

One article reviewed the literature on koilonychia, which is a common feature on toenails in infants. Although mostly transient and idiopathic in young children, some familial and syndromic cases have been reported [9].

Key Points

- Koilonychia, onycholysis, and onychoschizia are frequent findings on toenails in newborns, as well as the absence of lunula on both finger- and toenails.

Congenital Disorders

Congenital Malalignment of the Great Toenails

This entity is probably underdiagnosed, and most of the recent publications are case reports [10–13]. Congenital malalignment usually appears at birth or during the first years of life [10]. Mean age for medical consultation is 3 years old [6]. However, a few studies reported cases of great toenail malalignment with onset in adolescence or young adulthood without any history of nail surgery or acute trauma, raising the possibility of a late-onset presentation of this condition [11, 14]. Congenital malalignment of the great toenail spontaneously improved in 58% of a recent case series of 25 patients, similar to the 50% rate quoted in the literature [6, 15]. Clinical observation with photographic follow-up along with podiatric care is the rule. For severe forms, or those that don't resolve spontaneously, surgical realignment may be considered to avoid unsightly and persistent nail dystrophy [10, 16]. In the most recent series, four patients underwent a whole nail unit rotation surgery, with an improvement of the nail dystrophy in three cases [6].

Congenital Hypertrophy of the Lateral Nail Folds

Congenital hypertrophy of the lateral nail folds affects children around the age of 1 (Fig. 14.1). Great toenails are mainly involved, both in 67% of cases. Spontaneous improvement occurs in around 3/4 of cases, reason for which conservative care and follow-up is usually sufficient. Surgery is infrequent and restricted to very prominent or recalcitrant cases [6].

Pachyonychia Congenita

Pachyonychia congenita (PC) is a group of autosomal dominant disorders resulting from mutations in one of five keratin genes [17, 18]. The last classification, validated by phenotype-



Fig. 14.1 Prominent hypertrophic medial fold in a toddler

genotype correlations, divides PC into subtypes according to the mutation found in the keratin-encoding genes *KRT6A*, *KRT6B*, *KRT6C*, *KRT16*, and *KRT17* [19]. PC prevalence in Western countries is 0.9 cases per million with a worldwide PC population estimated between five to ten thousand. In a large case-cohort study of 815 patients conducted by Samuelov et al., the earliest and most common clinical feature of PC was nail dystrophy, appearing during the first year of life in most cases. The median number of affected fingernails and toenails was ten, while the mean number was 6.4 and 8, respectively. Fifty-one percent of PC patients had all 20 nails affected [18]. Nail dystrophy may be responsible for pain, infection, and functional and psychosocial impairment [17, 18]. It deeply affects social interactions and function of adolescents. Among patients with a detectable mutation, PC manifests with nail thickening and plantar keratoderma before school age in more than 3/4 of affected children, allowing early diagnosis [19]. *KRT6A* mutations were associated with a younger age at diagnosis and a higher number of fingernails/toenails involvement. Conversely, a lack of fingernail involvement was most commonly associated with *KRT16* and *KRT6B* mutations [18, 20]. An increased number of affected toenails and earlier age of toenails involvement predicted fingernail dystrophy, while an increased number of affected fingernails correlated with toenails involvement. Earlier onset of toenails involvement correlated with the development of plantar keratoderma,

while age of fingernail dystrophy onset and walking aids use predicted palmar keratoderma. Finally, oral leukokeratosis and natal teeth correlated with an earlier toenail involvement, while hoarseness correlated with an increased number of involved fingernails [18]. No specific histopathological feature is identified in PC nails. Parakeratosis and plasma globules are the most prominent features in both clinically affected and unaffected PC nails. The presence of onychomycosis in a nail plate does not exclude a diagnosis of PC [20]. Nail removal remains a therapeutic option and seems to have an overall positive outcome [17].

Nail-Patella Syndrome

Nail-patella syndrome (NPS) is a rare autosomal dominant condition characterized by variable nail, skeletal, renal, and ocular anomalies [21]. The incidence of NPS is usually given as 1 per 50,000, but an epidemiologically based incidence estimate is still lacking [22, 23]. *LMX1B* is the major gene responsible for NPS and explains around 95% of all cases [24]. To date, over 180 heterozygous mutations of *LMX1B* have been reported in NPS [22]. A new mutation in *WIF1* gene has also been identified, suggesting a potential novel cause of NPS [24]. Mean age at the time of clinical diagnosis is 6.54 years [25]. Almost all patients have nail abnormalities, the most frequent being hyponychia. Other findings include triangular lunula, absence of lunula, anonychia, nail splitting, pterygium, koilonychia, as well as the loss of the dorsal creases in the skin overlying the distal interphalangeal joints [21, 25]. These nail changes may be present since birth and are commonly bilateral and symmetrical [25]. Each individual nail is usually more severely affected on its ulnar side [23, 26]. Thumbnails are the most affected, and the severity tends to decrease towards the little finger [23, 25, 26]. When toenails are involved, the abnormalities tend to be less severe, and the little toenail is most commonly affected [23]. Triangular lunula is a pathognomonic finding, but it is not encountered in all cases [25]. When present, it is

mainly associated with a dystrophy of the medial part of the nail, ranging from a simple longitudinal crease to pseudopterygium and median nail fracture. These findings are detectable only in forms of NPS with severe nail involvement [26]. The severity of the nail signs is unrelated to visceral involvement [25, 26].

Key Points

- Congenital hypertrophy of the lateral nail folds is a benign condition that spontaneously improves in 3/4 of cases. Surgery should be restricted to resistant cases.
- Congenital malalignment of the great toenail spontaneously improves in around 50% of cases. Clinical observation and photographic follow-up are a must. Severe cases and non-improving cases should be proposed surgery.
- Pachyonychia congenita is responsible for severe psychosocial impairment. By the age of 5, more than 3/4 of patients have nail alterations, allowing early diagnosis.
- Nail alterations are constant in nail-patella syndrome. Triangular lunula is pathognomonic but not always present.



Fig. 14.2 Viral warts on the finger and the upper lip of a nail biter

to a shorter time period required for a complete cure of the periungual warts with diphenylcyclopropenone immunotherapy [28]. Another type of immunotherapy that can be used is Vitamin D3 injections in an oily base, just beneath the warts, every 2 weeks for a maximum of four sessions. This new treatment seems promising. However, pain during injection is the limiting factor. It is a safe, inexpensive, and effective treatment for periungual warts [29]. Topical cidofovir is a possible alternative for recalcitrant periungual warts or those that fail to respond to the conventional treatment. Nonetheless, its high price and its limited availability are major limitations [30]. The application of a nitric-zinc complex solution (Verrutop®) twice a month on warts resulted in a complete clearance in 83.9% with excellent cosmetic results and tolerance, according to a recent study [31]. In a small case series of 11 patients, translesional bleomycin delivered via a multipuncture technique using an ablative carbon dioxide fractional laser before the application of bleomycin on the wart resulted in a complete clearance – recurrence-free for 6 months – in 70% of cases [32]. Finally, a combination of superficial shaving with photodynamic therapy also seems to be an effective and safe treatment for recalcitrant, multiple, and thick periungual warts [33].

Infectious Diseases

Periungual Warts

Periungual warts are a therapeutic challenge (Fig. 14.2). Treatment modalities include salicylic acid, cryotherapy, bleomycin, contact immunotherapy – with diphenylcyclopropenone or squaric acid dibutyl ester – and laser ablation [27]. A shorter sensitization period and a more severe sensitization reaction (characterized by a higher “erythema and blister index”) contribute

Nail Scabies

To date, only five cases of pediatric nail scabies have been reported in the literature [34–36]. In the study of Chinazzo et al. including 47 children with confirmed scabies, a nail involvement was found in 6.4% of children, all infants. In two of the three cases, scabies was surprisingly found in a great toenail rather than in fingernails. According to Chinazzo et al., the nail area may constitute a reservoir of mites and may be a risk factor for scabies recurrence. Nails should not be overlooked during scabies treatment [34]. Some authors recommend to cut, brush, and apply a topical scabicide on the nails, even in classic scabies [35].

Acute Paronychia

Neonates with oral self-soothing behaviors may be more at risk for developing paronychia of mixed anaerobic and aerobic infections. Initial therapy with broad-spectrum antibiotics amoxicillin/clavulanate or clindamycin is suggested [37].

Onychomycosis

Onychomycosis in childhood is rare and affects approximately 0.2–2.6% of all children [38, 39]. However, an increased incidence has been reported in the pediatric population [38]. *Trichophyton rubrum* is the most commonly isolated pathogen, as in adults [39, 40]. Children are more likely to respond to monotherapy since they have thinner and faster-growing nails [41]. New topical treatments for onychomycosis have an increased nail penetration and sometimes additional delivery routes to the site of infection [42, 43]. Tavaborole may be a good candidate to treat children with onychomycosis. It penetrates the nail plate very well and is effective when

used in monotherapy [41]. It was found to be safe and well tolerated in the pediatric population [44]. When a systemic treatment has to be prescribed, terbinafine is the first choice. It is recommended to perform a baseline transaminase monitoring. On a study with 261 children treated with terbinafine, the authors discovered that 12.5% of patients had grade 1 laboratory abnormalities – such as liver alterations – prior to (8.3%) or during therapy (4.2%). However, if the pretreatment routine laboratory is within normal limits, further monitoring is unnecessary, considering the low incidence of clinically significant adverse effects, costs of laboratory tests, and patient discomfort [45].

Key Points

- Periungual warts remain a therapeutic challenge.
- Nail scabies may be a risk factor for resistance to therapy and recurrence.
- Neonates with oral self-soothing behaviors may be more at risk for developing paronychia.
- New topical treatments with better nail penetration may be used in monotherapy for onychomycosis.

Inflammatory Disorders

Nail Lichen Planus

In Goettmann et al.'s study, among 67 patients with nail lichen planus, around 11% were children [46]. In a more recent study from Morocco, among the 20 cases of nail lichen planus, amazingly 40% were children, highlighting the frequency of pediatric forms in this country [47]. Nail alterations are reported in 13.9% of children presenting with skin lichen planus [48].

Nail Lichen Striatus

In a report of seven cases by Kim et al., four were children, between 4 and 11 years old. All patients had both typical skin lesions and nail abnormalities, although lichen striatus can be limited to the nail. Nail lichen striatus usually involved a single digit. The most common nail change was longitudinal fissuring (Fig. 14.3) [49]. Iorizzo et al. described the dermoscopic features of nail lichen striatus: involvement of only one part of the nail plate with linear longitudinal fissuring, ridging, and distal splitting, especially if seen with perionychial skin lesions, is characteristic. This dermoscopic pattern may help in the diagnosis of nail lichen striatus [50]. In the study of Kim



Fig. 14.3 Isolated fissure on the index nail due to lichen striatus

et al., most nail lesions resolved within a mean of 4 months after the initiation of a topical corticosteroid cream and 0.1% tacrolimus ointment for both skin and nail lesions [49].

Trachyonychia

In a recent series of 26 patients, trachyonychia mainly affected children between 6 and 12 years old, with a mean age of 8 years [6]. Biopsies for diagnostic purposes are not recommended because whatever the cause, trachyonychia doesn't lead to nail destruction [51]. A favorable evolution was observed in 75% of cases, confirming the good prognosis previously published, regardless of treatment (Fig. 14.4) [6, 51, 52]. Nonetheless, an annual photographic follow-up is usually suggested to reassure parents and their child. In a recent series, trachyonychia did not appear to regress faster in patients receiving topical treatment, in contrast with the study of Park et al., which reported a significant nail improvement in 98.6% of cases with calcipotriol/betamethasone ointment applied once daily for 6 months [6, 53].

Nail Psoriasis

The prevalence of nail involvement in psoriatic children varies widely between studies. In



Fig. 14.4 Trachyonychia. Spontaneous healing over 2 years without treatment

recent series, nail involvement was reported between 15.7% and 32.3% of psoriatic children [54–56]. This prevalence increases with age [55, 56]. Fingernails are involved more frequently than toenails [55]. The main clinical features are pitting on fingernails and onycholysis as well as pachyonychia on toenails. Other common features include Beau's lines, oil drop, paronychia, leukonychia, and splinter hemorrhages. All fingers are involved at similar frequencies, whereas the big toenails are involved twice as often as the others, suggesting a Koebner phenomenon. Nail involvement is significantly associated with male gender, palmoplantar psoriasis, and a higher PASI (Psoriasis Area Severity Index) score, indicating a more severe disease course over time [55, 56]. When comparing the mean NAPSI (Nail Psoriasis Severity Index) and the mean PASI according to nail lesions, a study found that subungual hyperkeratosis and nail fold psoriasis were significantly associated with the severity of both nail psoriasis and cutaneous psoriasis [57]. As in adults, nail psoriasis is closely associated with psoriatic arthritis [55]. Nail involvement in children is also associated with metabolic comorbidities such as abdominal obesity, overweight, or metabolic syndrome [58]. The microscopic nail features such as the presence of neutrophils and serous lakes in the nail plate have also been associated with higher PASI and NAPSI scores in psoriatic children. Nail clippings remain very useful to help the diagnosis and to rule out onychomycosis [59].

Key Points

- Nail lichen planus is rare in children, accounting for about 11% of all nail lichen planus cases.
- Nail lichen striatus is characterized by a linear longitudinal fissuring, ridging, and distal splitting of one part of the nail plate.
- Trachyonychia spontaneously improves in most cases and never leads to nail

destruction, whatever the cause. Biopsies are therefore unnecessary.

- Nail psoriasis is associated with psoriatic arthritis and with a higher PASI score, indicating a more severe course of the disease.

Traumatic and/or Mechanical Disorders

Pediatric Finger Injuries/Acute Trauma

Fingertip injuries are common in children, affecting mostly boys under 5 years old, probably because of an imbalance between gross motor abilities and cognitive development [60–63]. Younger children are also naturally curious and use their hands to probe and explore their environment [62, 64]. Among all finger injuries, nail bed injuries and fractures are the most prevalent [60, 62]. Nail bed injuries can be classified as subungual hematomas, simple or stellate lacerations, avulsions, and crush injuries [65]. Crushing injury of finger by door or window is still the most common mechanism of injury among younger children and accounts for a large number of hospital admissions. The door is often closed by another child, at home, in the presence of an adult. The right hand is usually involved and the most commonly injured finger is the middle finger (Fig. 14.5) [60, 62]. Imaging rules out bone fractures [64]. The management of subungual hematomas and nail bed lacerations remains controversial. Most hematomas caused by nail bed lacerations do not usually require nail removal and repair, if the nail/nail margins are intact. Small hematomas are often left to reabsorb without treatment [66]. For larger and painful subungual hematomas, there is much debate about removal of the nail plate for repair of a nail bed laceration compared with simple trephination – i.e., the creation of a hole in the nail plate to release trapped blood [65, 66]. Satku et al. compared the results of nail removal and



Fig. 14.5 Post-traumatic dystrophy. Horizontal duplication of the matrix

formal nail bed reconstruction with trephination of a subungual hematoma after a fingernail crush injury. They found that simple nail trephination was equal or superior to removal of the nail with nail bed repair, with significantly lower cost [61]. The rate of accidents is not reduced by the presence of adults, which highlights the need for preventive measures and health education [60, 63]. Satku et al. suggested techniques to prevent door crush injuries in children, including placing door stoppers and using plastic hinge protectors [61, 62]. The cost of prevention of fingertip injuries by using safety equipment is less than the cost of treating them [61, 63]. Sports injuries are most common among the 13- to 18-year-old age group [62].

Podiatric Disorders

Podiatric abnormalities remain underestimated and should be considered as potential causes of nail abnormalities in children. It includes toe malposition, difference in shape/size between the two feet, abnormal gait, or improper shoes. Toe malpositioning results in nail thickening, nail bed hyperkeratosis, or onycholysis, which may mimic other nail disorders such as onychomycosis or nail psoriasis. If not treated, it can eventually lead to permanent dystrophies in adulthood and *sequelae* related to feet malposition, like back pain. It is therefore crucial to examine the whole foot and

recognize the toe position. Medical treatment is usually not necessary. Conservative measures are recommended, such as nail debridement or orthodigital devices. Referral to a podiatric practitioner is useful. Surgery is not recommended in children [67].

Onychotillomania and Onychophagia

Onychophagia affects all age groups but is more prevalent in children and adolescents, predisposing to psychosocial issues and complications including paronychia and dental problems. According to the study of Winebrake et al. including 281 patients, the prevalence of nail biting for more than 1 month was 37%, with a median age of onset of 5 years old. Only fingernails were involved in 88% of cases. There was a history of nail biting in 63% of cases. Biters were 3.34 times more likely to have a diagnosis of a psychiatric disorder than non-biters [68]. The study of Lynch et al. showed that children who suck their thumbs or bite their nails were less likely to have atopic sensitization in childhood and adulthood. On the contrary, no association was found with asthma or hay fever [69]. The application of bitter nail lacquers produced significant improvement in terms of odds of frequent nail biting [68]. Management of onychophagia includes a combination of pharmacotherapy, such as N-acetylcysteine and antidepressant therapy, stimulus control, habit reversal training, as well as cognitive behavioral and aversion therapy [68, 70]. N-Acetylcysteine warrants consideration as an efficacious pharmacological intervention due to its relative safety [68, 71]. Auricular acupressure appears to improve the efficacy of habit reversal treatment, likely by reducing anxiety [72]. Halteh et al. discourage practitioners to tell children that no treatment is needed and that they will outgrow it [73].

Retronychia

Retronychia is mostly described in adults, but pediatric cases have also been reported, with



Fig. 14.6 Retronychia in a teenager

the same clinical features (Fig. 14.6) [74]. Until recently, nail avulsion was the treatment of choice. A study from Lencastre et al. evaluated the efficacy of potent topical steroids in retronychia. The youngest patient included in the study was 12 years old. Topical steroids were used under occlusion for an average duration of 8 weeks, with a complete or partial response in 41.1% and 28.5% of cases, respectively. Response correlated with milder paronychia and longer treatment durations. Topical steroids should therefore be the first-line treatment of retronychia, especially in milder forms, associated with footwear and gait biomechanics assessment [75, 76]. Nail avulsion should still be performed if there is no improvement after 10 weeks [75].

Ingrown Nails

In the study of Arica et al., a positive family history of ingrown nail was present in 15.7% of patients. High prevalence of incorrect nail cut-

ting (72.1%), trauma (36.1%), poorly fitting shoes (29%), hyperhidrosis (12.9%), and obesity (9.7%) were determined among the patients with ingrown toenails. The mean age at presentation was 15 years [77]. Ingrown toenails in children and adolescents should be treated in first instance by nonoperative methods, such as taping and nail brace application [78–80]. Operative options can be considered for resistant cases or in case of recurrence [80]. Partial nail avulsion with chemical matrix cauterization is considered as the technique of choice [81]. However, Livingston et al. suggested to consider the Vandembos procedure – a large debulking of the lateral folds with secondary intention healing – which is associated with a low recurrence rate in children, adolescents, and young adults with ingrown toenails. Patient-reported recovery time, complication rate, functional outcomes, and satisfaction are excellent [82].

Key Points

- Fingertip injuries are common in children, mostly crushing by a door or a window. This should be preventable with adequate measures.
- Topical steroids are the first-line treatment of retronychia, especially in milder forms.
- Nail biting is common in children. It is associated with psychiatric disorders but reduces the risk of atopic sensitization.
- Ingrown toenails in children and adolescents should be first treated conservatively.

Nail Tumors

Subungual Exostosis

Subungual exostosis (SE) is an uncommon, benign osteocartilaginous tumor of the distal phalanx [83]. Half of the patients are under 18, with an equal female/male ratio [83–85]. The hallux is the most common location [83, 84].

Fingernails are rarely affected. Pain is a common complaint [83]. A history of trauma is present in about 30% of cases [84]. One study suggested that trauma, including surgeries, may have a dual function of induction and regression of SE [86]. Infections are also frequently implicated in the etiology of the disease [83]. A study focusing on dermoscopic features of subungual exostosis showed that vascular ectasia (Fig. 14.7) was the most common dermoscopic finding, followed by hyperkeratosis, onycholysis, and ulceration [87].

Longitudinal Melanonychia

The mean age at time of consultation for longitudinal melanonychia (LM) is 8 years old [6]. In a recent histopathological study of 30 childhood cases of melanonychia conducted in the USA, Cooper et al. reported subungual lentigo in 20 cases, subungual nevus in 5 cases, and atypical melanocytic hyperplasia in 5 cases [88, 89].



Fig. 14.7 Subungual exostosis of the hallux in a teenager

Childhood LM due to a benign nail matrix nevus (NMN) displays more melanoma-associated features compared with those of adults [90, 91]. Those features include a sharply demarcated pigment band of even width, a Hutchinson's sign – usually observed at the hyponychium and/or proximal nail fold cuticles – and a longitudinal brush pigmentation [91]. A pseudo-Hutchinson sign, a triangular sign, an irregular pattern, as well as dots and globules are also detected more commonly in children. NMN in children are darker and multicolored. Pigmentation also tends to be broader in children, but without statistical significance [90]. There is still no consensus on how to manage or follow up NMN in children. It has been shown that the predictive scoring model for dermoscopy of nail melanoma in situ couldn't be applied to the pediatric population (Fig. 14.8) [92–94]. Nail melanoma is exceedingly rare in children, with less than 20 cases of pediatric subungual melanoma in the literature, without any deaths reported to date [89, 92, 95]. Only two cases were subungual invasive melanomas that subsequently metastasized to regional lymph nodes without distant metastases, providing evidence that subungual melanoma with

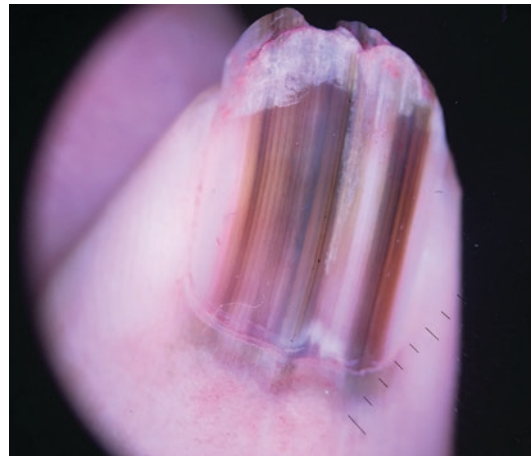


Fig. 14.8 Longitudinal melanonychia in a child. The dermoscopic aspect would be extremely worrying in an adult. These types of features are very common in LM in children and do not account at all for a malignant process. The lesion was removed after several years of follow-up and showed a junctional nevus

metastatic potential does occur in children, albeit exceptionally rarely [92]. Therefore, the vast majority of excised lesions are in fact benign, even if they display worrisome clinical, dermoscopic, or sometimes histopathological features [88, 96]. Considering the morbidity associated with surgical excision of the nail matrix and the rarity of nail unit melanoma in children, the overwhelming majority of cases can be managed conservatively with photographic and dermoscopic follow-up, especially under 14 years old, with biopsy only required in selected cases [88, 89, 92, 96]. When LM appears in older teenagers, a closer follow-up is required [89]. Piraccini et al. decide to biopsy only when the band rapidly enlarges and involves the whole nail and when its color is dark black. This management remains difficult, especially when dealing with anxious parents [94, 97].

Onychopapilloma

Onychopapilloma is a benign nail unit tumor that is usually seen in adults. Two cases of onychopapilloma in children – aged 9 and 10 years old – and one in a teenager have been recently described [98, 99].

Key Points

- Subungual exostosis affects mainly young patients; half of them are <18 years old.
- Longitudinal melanonychia is benign in children in the vast majority of cases, even if it frequently displays worrisome features. A wait-and-see policy is most of the time appropriate.
- Onychopapilloma is a benign longitudinal nail tumor which can occur in children and teenagers.

Systemic and/or Iatrogenic Disorders

Fever-Related Beau's Lines or Onychomadesis

A temporary cessation in matrix growth from a systemic shock like a thermal peak may result in a Beau's line or an onychomadesis, according to the duration of the injury [100]. This condition is observed mainly in the group 2 to 6 years old, most probably because of the fragile nail matrix in young children [6, 101]. Thirty percent of children with hand, foot and mouth disease (HFMD) have such nail abnormalities [102]. Fingernails are more commonly involved than toenails. Nail changes typically occur synchronously [101]. It is still debated whether the inhibition of the nail matrix proliferation results from direct inflammation spreading from skin lesions of HFMD around nails or a coxsackievirus-specific nail matrix involvement, or HFMD's severe systemic impact on the general condition of the small children [100]. In HFMD, nail changes usually occur within 1 or 2 months after onset and last for 1 to 8 weeks [101].

Yellow Nail Syndrome

YNS is a rare disorder and is even more rarely reported in children with around 20 cases described in the literature. As in adults, yellow nails and recurrent respiratory infections are the most common presentations. Lymphedema was also noted in 12 of 19 cases in the literature. There seem to be two age peaks in the incidence of YNS. Most pediatric cases presented at birth, maybe because of a transplacental crossing of titanium from the mother to the fetus. The second peak appears between the age of 6 and 10 and may be related to the high exposure to candy or children's toothpastes that contain titanium dioxide at this age. Avoiding titanium exposure could alleviate the symptoms of YNS [103]. The prognosis of YNS in children is not known [104].

Key Points

- Beau's lines and onychomadesis occur in 30% of children with hand, foot and mouth disease, within 2 months after the infection.
- Yellow nail syndrome is an extremely rare disorder in children, although severe. It may be related to titanium absorption from candies and toothpastes.

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