Original paper

# Childhood longitudinal melanonychia: case series from Poland

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## Abstract

**Introduction:** Longitudinal melanonychia (LM) is characterized by a tan, brown or black longitudinal streak within nail plate caused by the presence of melanin. LM is relatively common in dark-skinned population, infrequent in Caucasian population, and rare in children.

**Aim:** We report epidemiological, clinicopathological and dermoscopic analysis of 8 cases of childhood LM from Poland, which is the largest series in the Central and Eastern European population.

**Material and methods:** Three hundred and forty-eight patients presenting with various nail pigmentation (in 2010–2016) were analysed. 72 cases of LM have been identified, including 8 cases of childhood LM (< 16 years of age), which were included in further analysis.

**Results:** Seven patients were boys and one girl, with mean age of 9 years (range: 6–13). More than a half ($n$ = 5) presented skin phototype II. The most common location of melanonychia was the first left fingernail. Dermoscopy revealed heterogeneity of longitudinal lines colour in 5 cases. The irregularity of longitudinal line thickness in 5 cases and irregularity of parallelism in 5 cases was observed. Histopathological evaluation was performed in 4 patients, in 3 cases it revealed the presence of nail matrix nevus, in one case the presence of melanocytic proliferation of the lentiginous pattern along the dermoepidermal junction.

**Conclusions:** Despite the fact that melanoma was not recognised in any case, such a possibility should always be considered as the cause of LM, even in the paediatric population. Dermoscopy seems to be useful in patient follow-up and management.

**Key words:** longitudinal melanonychia, nail apparatus melanoma, children, dermoscopy.

## Introduction

Longitudinal melanonychia (LM) also known as melanonychia striata is defined as a grey to black pigmentation of the nail plate due to the presence of melanin caused by hyperplasia or activation of nail matrix melanocytes [1]. The most important clinical aspect of LM is the association with the nail apparatus melanoma (NAM). It is estimated that approximately 76% of cases of this neoplasm presents initially as LM [1, 2]. NAM represents from 0.7% to 3.4% of all diagnosed melanomas in the Caucasian population. Rarity of this entity and non-specific clinical presentation contribute to the delay of the treatment due to inaccurate initial diagnosis. This determines worse prognosis in comparison to cutaneous melanoma [3].

The incidence of LM depends on genetically determined differences in the number and activity of melanocytes localized in nail apparatus. LM is relatively common in Afro-American, Japanese and Latino population. According to literature data, the occurrence of LM in Caucasian population does not exceed 1%. LM is rarely observed in children [1–5]. In the Polish literature this medical issue has not been reported yet.

## Aim

To present the largest series of childhood LM in the Central and Eastern European population.

## Material and methods

We analysed 348 patients who have been diagnosed with various nail pigmentation in 2010–2016. In adults, 64 cases of LM have been identified (including 3 cases

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**Table 1.** Clinical and dermoscopic features of studied cases

| Patient | Sex | Age of diagnosis [years] | Age of onset [years] | Phototype | Location | Hutchinson sign/pseudo-Hutchinson sign | Width of the pigmented band [mm] | Nail dystrophy | Colour | Irregularity of thickness | Irregularity of parallelism | Patient management | Histopathology |
|---|---|---|---|---|---|---|---|---|---|---|---|---|---|
| 1 (Figure 1) | M | 13 | 9 | IV | 2nd LT | (+)/(–) | 8 | (–) | Heterogenous | + | + | Total excision of the nail apparatus | Nail matrix nevus |
| 2 (Figure 2) | M | 6 | 4 | II | 5th RF | (–)/(+) | 3 | (–) | Homogenous | – | – | Follow-up (spontaneous regression) | – |
| 3 | M | 8 | 7 | III | 1st LF | (–)/(+) | 2 | (–) | Homogenous | – | + | Follow-up | – |
| 4 | M | 12 | 10 | II | 5th RF | (–)/(+) | 3 | (–) | Heterogenous | + | – | Follow-up | – |
| 5 | M | 7 | 4 | II | 1st RF | (–)/(+) | 1 | (–) | Homogenous | – | – | Follow-up | – |
| 6 (Figure 3) | M | 8 | 6 | II | 1st LF | (–)/(–) | 8 | (–) | Heterogenous | + | + | Partial excision of the nail apparatus | Nail matrix nevus |
| 7 (Figure 4) | M | 12 | 9 | III | 1st LF | (–)/(+) | 5 | (–) | Heterogenous | + | + | Follow-up and total excision of the nail apparatus | Melanocytic proliferation of lentiginous pattern along the dermoepidermal junction |
| 8 | F | 6 | 4 | II | 2nd RF | (–)/(+) | 4 | (–) | Heterogenous | + | + | Partial excision of the nail apparatus | Nail matrix nevus |

*M – male, F – female, RF – right fingernail, LF – left fingernail, RT – right toenail, LT – left toenail, (+) – presence, (–) – absence.*

of NAM; 4.7%), whereas 8 cases of LM have been identified in children (< 16 years of age). The latter were included in the further detailed analysis. Final diagnosis was established according to clinical, dermoscopic (onychoscopic) and histopathological evaluation.

## Results

Clinicopathological and dermoscopic features of the analysed cases are summarized in Table 1 (Figures 1–4). Mean age at diagnosis was 9 years (range: 6–13), whereas the mean age of LM onset was 6.6 years (range: 4–10). There was no personal or family history of melanoma in the studied group. All children were otherwise healthy. No preceding trauma, history of medications intake or pigmentation-related disorders were reported.

The most common location of melanonychia was the left first fingernail. Mean width of the pigmented band was 4.3 mm. In 2 cases, pigmentation involved > 50% of the nail plate; in 3 cases, > 30% of the nail plate. The Hutchinson sign was present in one case, and pseudo-Hutchinson sign in 6 cases. Nail dystrophy has not been observed. Dermoscopically, the colour heterogeneity of longitudinal lines within a band was present in 5 cases; the irregularity of longitudinal lines thickness was present in 5 cases and irregularity of parallelism in 5 cases.

In 3 patients, the histopathological examination was performed after initial evaluation. In 1 patient (patient 1, Figure 1 A), after considering the doubtful character of the lesion, mother's anxiety and size of the lesion, a decision of total excision of the nail apparatus was made. The procedure was performed in conduction anaesthesia. The nail apparatus was excised in one piece, the wound was covered with full-thickness skin graft taken from the inguinal area. The postsurgical period was uncomplicated. Further aesthetic results were satisfying (Figure 1 B). Histopathological examination revealed the presence of nail matrix nevus.

In 5 patients, the regular follow-up was recommended. In patient 2, spontaneous regression of LM was observed. Clinical examination at the first visit revealed 3 mm wide brown longitudinal streak of the right fifth fingernail; after 6 months, previously observed pigmentation was absent (Figures 2 A, B).

In patient 7, after 13-month follow-up, based on the worrying change in clinical and dermoscopic presentation (Figures 4 A, B), a decision of total excision of the nail plate was

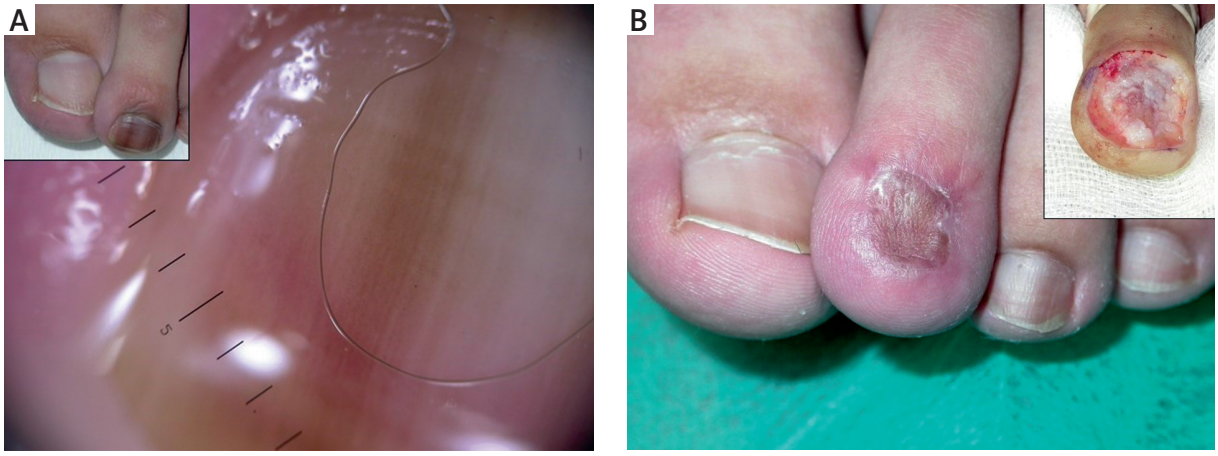


**Figure 1.** Patient 1. **A** – LM involving most of the second toenail plate with the presence of Hutchinson sign (clinical and dermoscopic presentation). Heterogeneity in colour and irregularity of thickness and parallelism of the longitudinal lines. **B** – Clinical presentation after wide local excision of the nail apparatus and well-healed toenail after skin graft

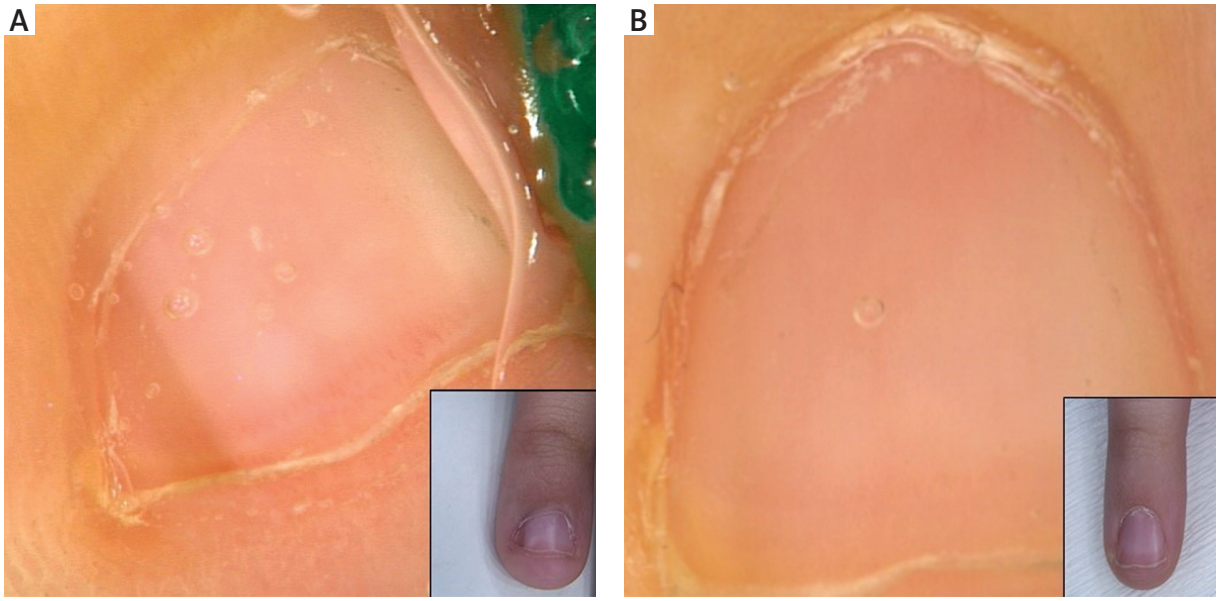


**Figure 2.** Patient 2. **A** – LM of the fifth right fingernail (clinical and dermoscopic presentation). Longitudinal, homogenous in colour, regularly distributed pigmented bands. **B** – Spontaneous regression of LM after 6 months (clinical and dermoscopic presentation)

made. Histopathological examination revealed the presence of melanocytic proliferation of the lentiginous pattern along the dermoepidermal junction (Figures 5 A, B).

## Discussion

LM incidence in the Caucasian population is estimated to be approximately 1% [3]. Nevertheless, authors' observations indicate a much lower frequency of this lesion. The overstatement of the data may result from inconsistency of nomenclature. Some researchers classify lesions with non-melanocytic origin (e.g. subungual haemorrhage) into a group of melanonychia [5, 6]. Inde-

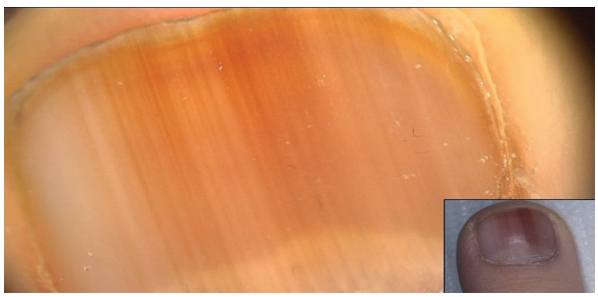

**Figure 3.** Patient 6. LM of the left thumb (clinical and dermoscopic presentation). Heterogeneity in colour and irregularity of thickness and parallelism of the longitudinal lines

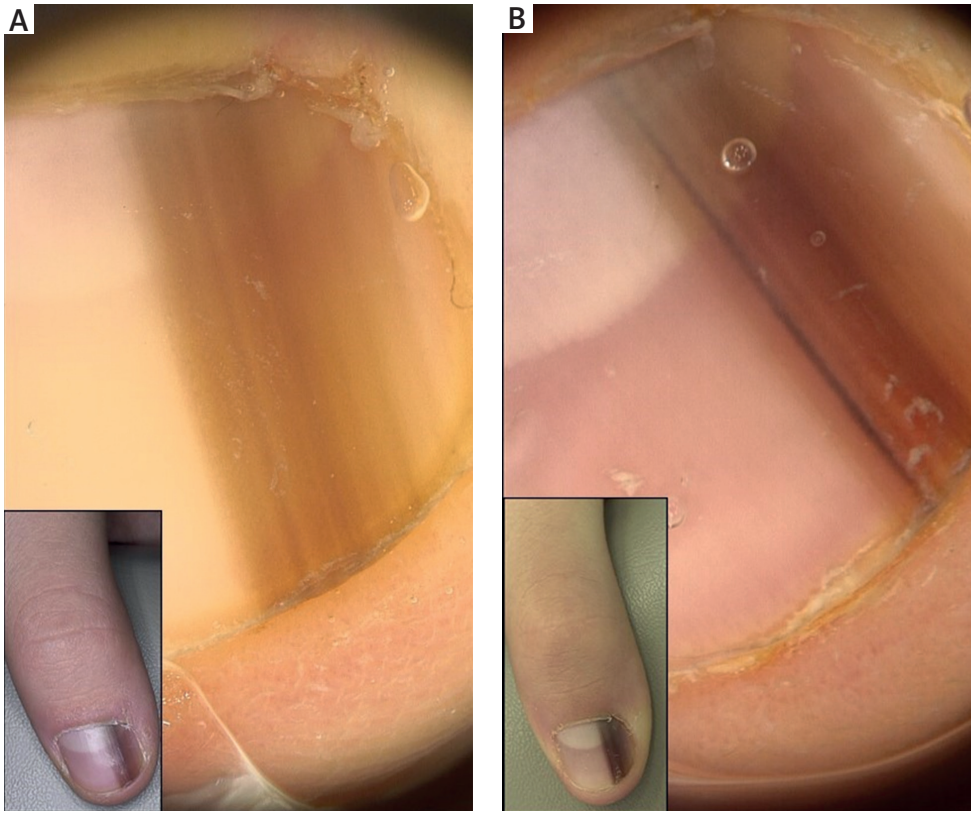


**Figure 4.** Patient 7. **A** – LM of the left thumb (initial clinical and dermoscopic presentation). Moderate heterogeneity in colour and irregularity of thickness and parallelism of the longitudinal lines. **B** – LM of the left thumb – a significant change in clinical and dermoscopic presentation after 13-month follow-up. Remarkable heterogeneity in colour and irregularity of thickness and parallelism of the longitudinal lines

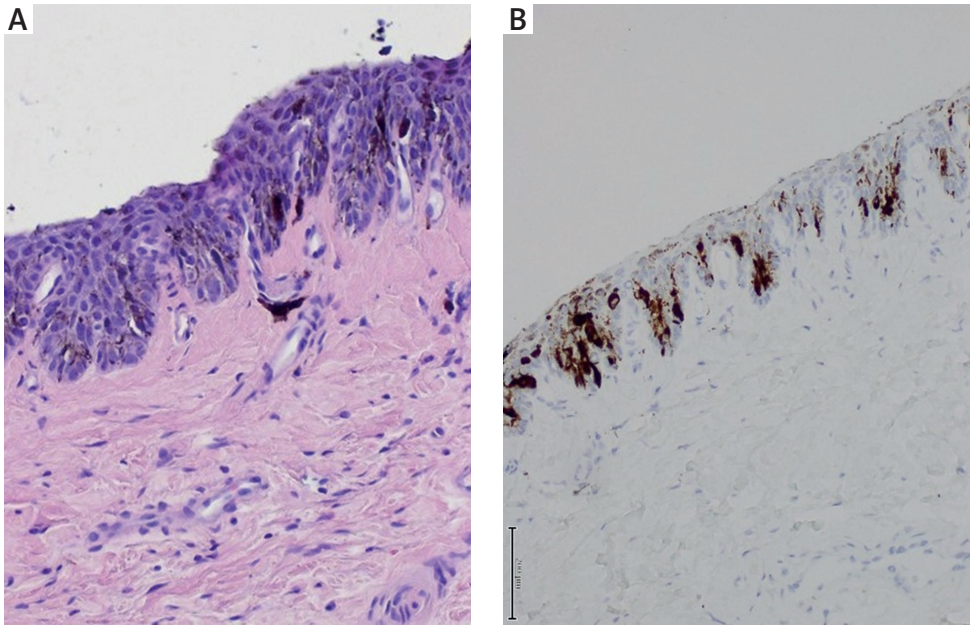


**Figure 5. A** – Melanocytic proliferation of the lentiginous pattern along the dermoepidermal junction with focal individual epithelioid melanocytes revealing architectural and cytologic atypia (hematoxylin and eosin). **B** – Melan-A staining of melanocytes at the dermoepidermal junction

**Table 2.** Clinical characteristics of cases of childhood nail apparatus melanoma: a literature review

| No. | | Age of onset [years] | Age of diagnosis [years] | Sex | Origin country | Phototype | Location | Clinical presentation | Histopathological diagnosis | Comments |
|---|---|---|---|---|---|---|---|---|---|---|
| 1 | Lyall, 1967 [19] | At birth | 1 | M | ND | ND | 3rd RF | ND | Invasive MM | Lymph node metastases |
| 2 | Uchiyama, 1979 [20] | 1 month | 7 | ND | ND | ND | 3rd RF | ND | Invasive MM | Lymph node metastases |
| 3 | Hori, 1988 [21] | ND | 3 | F | Japan | ND | 5th LF | ND | MM *in situ* | |
| 4 | Kato, 1989 [22] | 1 | 4 | M | Japan | ND | 3rd LF | Diffused pigmentation involving the entire nail plate and proximal nailfold | MM *in situ* | Diagnosis debatable |
| 5 | | 1.5 | 2 | F | Japan | ND | 2nd RF | LM | MM *in situ* | Diagnosis debatable |
| 6 | | 0.5 | 1 | F | Japan | ND | 1st RT | LM | MM *in situ* | Diagnosis debatable |
| 7 | Kiryu, 1998 [23] | 3 | 5 | F | Japan | ND | 5th LF | LM | MM *in situ* | Diagnosis debatable |
| 8 | Antonovich, 2005 [24] | 13 months* | 7 | F | Philippines | ND | 4th LF | LM | MM *in situ* | |
| 9 | Motta, 2007 [25] | 3 | 12 | F | Spain | III | 1st RF | Initial presentation: brown spot under the lunula | MM *in situ* | |
| 10 | Iorizzo, 2008 [26] | 1 | 14 | F | Argentina | IV | 3rd RF | LM | MM *in situ* | |
| 11 | | 5.5 | 6 | M | Brazil | III | 1st RT | LM | MM *in situ* | |
| 12 | Tosti, 2012 [27] | At birth | 0.5 | M | Italy | II | 1st RT | LM | MM *in situ* | |
| 13 | | 1 | 11 | F | Italy | II | 2nd RF | LM | MM *in situ* | |
| 14 | Bonamonte, 2014 [28] | 2 | 9 | M | Italy | II | 5th LF | LM | MM *in situ* | |
| 15 | Haddock, 2014 [29] | ND | 5 | F | USA | ND | 2nd RF | ND | MM *in situ* | |

*ND – not described, *the child was adopted at the age of 13 months, the precise onset of the LM was not known.*

pendently of the studied population, LM in children occurs very rarely. In Chinese research performed by Leung *et al.* [7], among 461 examined patients aged up to 19 years, no LM cases have been found. In our material, among 348 patients consulted due to various nail apparatus pathologies, 72 cases of LM have been identified, including 8 cases of childhood LM (2.0%).

The relationship between LM and NAM is currently undeniable. Even though the optimal patient management has not been established yet, most authors recommend histopathological evaluation of the disorder [8, 9]. Unfortunately, it is a painful procedure associated with the risk of permanent nail deformation. Therefore, in childhood LM the biopsy is performed in cases suspected of melanoma or on the parents' request.

The prediction of the course of LM in children is challenging. Although melanoma represents up to 2% of all diagnosed cancers in children, NAM is very rare. Unfortunately the disease, if occurs, is characterized by high mortality [10].

Goettmann-Bonvallot *et al.* [11] analysed 40 cases of LM in patients aged less than 16 years. The study identified 19 nevi (17 junctional and 2 compound), 12 lentigines and 9 cases of melanocytes activation as the background of LM. Similarly, Tosti *et al.* [12] and Léauté-Labrèze *et al.* [13] did not observe melanoma in 22 and 8 children with LM, respectively. Ohn *et al.* [14] analysed 58 nail matrix nevi in 56 children and concluded that nail matrix nevi in children significantly more often reveal melanoma-associated features like: multicolour pattern, darker discolouration, pseudo-Hutchinson sign, triangular sign, and the presence of dots/globules.

There are no definite recommendations regarding the management of childhood LM, therefore in our material, dermoscopic indications to biopsy were the same as in adults: the presence of Hutchinson sign, width of the pigmented band > 1/3 of the nail plate width, dark-brown colour of the background and irregular dermoscopic pattern (various colours and thicknesses of the longitudinal lines with uneven intervals) [15]. Another important factor was the change in clinical and dermoscopic presentation, which implicated prompt histopathological evaluation in patient 7.

In the literature some cases of spontaneous regression of childhood LM have been described [16–18]. Murata and Kumano [17] showed that randomly distributed dots and lines that follow the melanocytic lines may be indicators of spontaneous fading of LM in children. In our material, we did not observe these structures in a patient with spontaneous regression.

Our observations and literature data strongly support the rarity of childhood NAM in Caucasian population. To the best of our knowledge, only 15 cases (including 3 that occurred in fair-skinned Caucasians) of NAM in children have been reported so far. Most childhood NAMs described so far presented clinically as LM; the histopathological examination confirmed the diagnosis of melanoma "in situ" (Table 2) [19–29]. Nonetheless, in 2 cases, melanoma had an aggressive course [26]. Interestingly, these both cases did not present as LM, what indicates indirectly that nail matrix melanocytes were not the origin of melanoma.

The difficulties in interpretation of the histopathological picture of nail apparatus pigmentation disorders are being emphasized. Some aspects, considered in adults as evidence of malignancy (e.g. nucleus atypia, moderate migration of melanocytes), may be present in benign pigmented lesions in children, what we present in patient 7 [19, 20, 26, 30, 31]. In Goettmann-Bonvallot *et al.* report [11], nuclear atypia and moderate migration of melanocytes were observed in 15% and 20% of cases, respectively. The fact brings into question 4 cases of NAM diagnosed by Kato *et al.* [22] and Kiryu [23]. Recently, Bonamonte *et al.* [28] mentioned a possible utility of a novel immunohistochemical marker, anti-p16, in such cases.

## Conclusions

In the light of presented facts, the risk of childhood NAM in Caucasian population appears to be low. The presented cases seem to support the thesis of benign background of LM in children. Histopathological evaluation or removal of the lesion does not seem to be reasonable in every case. Nevertheless, regarding the rarity of this entity (especially in Caucasian population), difficulty in predicting the evolution of the lesion, and the possibility of development of NAM in adults with LM observed since the childhood, regular long-term follow-up should be recommended. Dermoscopy is useful in initial and subsequent patient assessment.

## Conflict of interest

The authors declare no conflict of interest.

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