Melanonychia describes a brown or black pigmentation of the nail plate caused by the presence of melanin. In this article, we review possible causes of melanonychia and discuss the main problems of management of patients with this condition. The goal in the management of melanonychia is early diagnosis of melanoma of the nail matrix and bed. Melanoma of the nail bed is also known as subungual melanoma. We discuss clinical, dermoscopic features that may help the clinician in selecting lesions that should have excisional biopsy and evaluate different options for the excision. Addressing melanonychia is still a difficult task, and the correct management of pigmented bands in children is far from established. Dermoscopy is possibly a useful tool but the real benefit of this technique, screening lesions to determine which ones need to be removed, remains to be proven.
malities (Fig. 2). Nail melanocyte activation occurs with trauma of the proximal nail fold overlying the matrix and not with trauma affecting the tip of the toes, as occurs in athletes.

Onychomycosis
In onychomycosis, melanonychia may be caused by melanocyte activation but also by direct melanin production by the fungi. Some nondermatophytic molds (Scytalidium dimidiatum and Alternaria alternata) and Trichophyton rubrum (var nigricans) produce pigmented hyphae (dematiaceous) that can cause diffuse or banded nail pigmentation.5,6

Systemic Causes
Although frequently mentioned in textbooks, melanonychia caused by systemic diseases is rare, the most common association being with endocrine disorders, such as Addison’s disease.

Benign Conditions
Nonmelanocytic benign and malignant tumors and benign conditions often cause melanonychia by stimulation of melanocytes; these include onychomatricoma, Bowen’s disease, squamous cell carcinoma, myxoid cysts, and warts.

Melanonychia Caused by Melanocyte Hyperplasia
The pathology is characterized by a proliferation of melanocytes within the nail matrix and/or the nail bed epithelium. This type of melanonychia is a challenge both for the clinicians and the pathologists to rule out melanoma of the matrix.

Lentigo/Benign Melanocytic Hyperplasia
The prevalence of melanonychia caused by lentigo is not known. The pathological criteria for this diagnosis are not well established, and some authors consider melanocytic hyperplasia to be a potentially malignant lesion.7 Lentigo is characterized by an increased number of melanocytes arranged as single cells within the epithelium of the nail matrix.5,8 Amin et al10 found that the number of melanocytes in lentigo ranged from 5 to 31 (with a median 14) per millimeter of basal membrane length.

Benign melanocytic hyperplasia was diagnosed in 12% of the biopsies of melanonychia in adults3 and 30% in children.8 There are no clinical or dermoscopic parameters that allow specific differentiation of melanonychia due to lentigo. The dermoscopic literature does not address this variety of melanonychia but rather places lentigo among the causes of melanocyte activation.12

Nevus of the Nail Matrix
Nail matrix nevi typically are seen in young people and may be congenital or acquired. They have been reported to represent approximately 12% of longitudinal melanonychia in adults11 and 48% in children.8 The nail has one or more longitudinal pigmented bands varying in size from a few millimeters to the whole nail width and in color from light brown to black.

Dark bands often are associated with pseudo-Hutchinson’s sign, because the dark nail plate pigmentation is visible through the transparent nail fold (Fig. 3a, b). The pigmentation may be homogenously distributed or darker bands may appear over a diffuse pale pigmentation. The fingernails are more often involved than the toenails, with no predilection for a particular digit.

Some clinical features of nail matrix nevi in children can be alarming, including the following:

- Hutchinson’s sign, periungual pigmentation: Congenital nevi often involve the nail folds and the hyponychium.
- Variation in the width of the band: In children, it is not uncommon to notice a gradual enlargement of the band that may have a broader proximal part than the distal part, resulting in a triangular shape.
- Variation in the color of the band: Darkening and spreading of the pigmentation is not unusual.

Thinning and fissuring of the pigmented nail plate may also occur.

In children, it is also quite common to observe gradual fading of the band. Fading of the pigmentation is not an indication of regression of the nevus but just of reduced melanin production from nevus cells.8 Pathologically, most of the nevi in children are junctional nevi.8 The rate of progression of nail matrix nevi to melanoma is not known but is probably rare.

Melanoma
Melanoma of the nail unit is a rare entity, comprising only 0.7% to 3.5% of all forms of melanoma and typically presenting in a more advanced...
The overall prognosis is poor, with the 5-year survival rate ranging from 16% to 87% depending on the case series. Delay in the diagnosis is common and is associated with poor prognosis.

Melanoma of the nail bed and matrix is more frequent in ethnic populations. Because most articles on the epidemiology and treatment of melanoma of the nail unit do not distinguish nail matrix melanoma from subungual melanoma but just use the term subungual melanomas to refer to all lesions, we don't really know if most melanomas arise from the matrix or from the nail bed; therefore, we use the term melanoma of the nail unit in the following discussion.

Although melanoma of the nail unit represents 1% to 2% of all melanomas in Caucasian populations, it has a prevalence of 10% to 23% in Asian populations and 25% in African-American populations.3,17 Melanoma of the nail unit may occur at any age, but it is a rare occurrence in children.18 There is no demonstrable association between the development of subungual melanoma and melanoma of the nail matrix with excessive exposure to ultraviolet light. The nail plate acts as a barrier to UVB radiation. Although trauma is often implicated as the putative factor, its role as the causative factor is unknown.19

The thumb and the big toe are the most frequently affected locations. In a study performed by Tan and colleagues14 in which they examined 124 cases of melanoma of the nail unit (the largest series reported until now), the most common site was the great toe (24%) followed by the thumb (18%). A study performed by Cohen and colleagues16 of 49 patients showed that most patients were female with a median age of 66 years; the most common site again was the big toe (53%), followed by the thumb (31%) and the fifth finger (10%).

The clinical presentation of melanoma of the nail unit depends on the site of origin which is either the nail matrix or the nail bed. The appearance is summarized as follows:

**Nail Matrix Melanoma**
Lesions that originate in the nail matrix usually cause a banded pigmentation of the nail plate (longitudinal melanonychia). This is the first symptom in up to 70% of cases. The color of the band is usually light brown to black and of variable width (Fig. 4a, b). The nail plate may present as fissure or a split corresponding to the band, indicating compression or destruction of the nail matrix epithelium by the melanoma.

**Nail Bed Melanoma (Subungual Melanoma)**
This causes a pigmented or nonpigmented (25% to 30% of cases) subungual nodule. Nail bed ulceration and bleeding occur when the tumor grows. Clinical differential diagnosis with ulcerated nail bed tumors and with nail bed pyogenic granuloma is often impossible.

An ABCDEF rule for the early detection of melanoma of the nail unit has been proposed (Table 1).20 Hutchinson’s sign describes the presence of pigmentation in the periungual skin and represents the radial growth phase of melanoma of the nail unit. Although Hutchinson’s sign is not exclusive to melanoma, its presence requires a biopsy (Table 2).

**Dermoscopy**
When performed by trained examiners, dermoscopy has already proven its efficiency in the differential diagnosis of cutaneous pigmented tumors. The main difficulty in the evaluation of nail pigmentation is that the lesions that are examined with dermoscopy correspond to melanin deposition in the nail plate and not to the site of melanin production, which is in the nail matrix or in the nail bed. Intraoperative nail matrix dermoscopy permits more accurate diagnosis but it is an invasive procedure that cannot be used routinely.

Dermoscopy also has been proposed before surgery to select the anatomic site to be explored. Examination of the distal edge of the nail establishes the localization of pigment within the nail plate and then which part of the matrix is involved. Pigmentation of the lower nail plate corresponds to a distal nail matrix origin, and pigmentation of the higher nail plate corresponds to a proximal localization of responsible melanocytes. In most cases of melanonychia, the pig-

![Figure 3](image-url) **Figure 3** (a) Three-years old boy with a dark band of melanonychia of the right thumbnail. (b) Dermoscopy shows a brown band with longitudinal parallel lines; note the pseudo Hutchinson’s sign. The band was excised and pathology revealed a nevus (Fotofinder, ×20 magnification).
ment is in the lower (ventral) part of the nail plate as most bands originate from the distal matrix.21

Nail Plate Dermoscopy

Dermoscopy of the nail plate always requires oil or gel immersion because of the convex shape of the nail. Dermoscopic patterns for evaluation of nail pigmentation have been described but their accuracy in the diagnosis of subungual melanoma has not been established.12,22-24 Similarly, there are no evidence-based studies to inform the clinician as to the frequency of dermoscopic follow-up in patients with melanonychia. There are no precise dermoscopic criteria that can be used to decide when to biopsy the lesion. Dermoscopy of the nail plate permits differential diagnosis between non-melanocytic and melanocytic pigmentation and may permit differential diagnosis between nail matrix melanocyte activation and hyperplasia but should not be considered a substitute for pathology in the differential diagnosis of longitudinal melanonychia.

Dermoscopic Patterns That Suggest Subungual Hematoma

Irregularly-shaped purple to brown-black areas with round, dark red spots at the periphery and a “filamentous” distal end are patterns that have been associated with subungual hematomas. However, we should remember that the presence of blood extravasation does not exclude an associated melanoma.

Dermoscopic Patterns That Suggest a Diagnosis of Melanocyte Activation

A gray background with thin gray regular parallel lines suggests melanonychia due to nail matrix melanocyte activation. In traumatic melanocyte activation, tiny dark red to brown spots corresponding to blood extravasation may also be seen.

Dermoscopic Patterns That Suggest a Diagnosis of Nevus

The presence of a brown background with longitudinal brown to black regular parallel lines often suggests a nevus. In children, black dots (less than 0.1 mm) similar to those described in skin melanocytic lesions are frequently observed and correspond to pigment accumulation in the nail plate.

Dermoscopic Patterns That Suggest a Diagnosis of Melanoma

Brown background with longitudinal, brown to black lines with irregular coloration, spacing or thickness and parallelism disruption suggest melanoma. Dermoscopy also can be used to detect Hutchinson’s sign before clinical detection by unaided visual inspection. Dermoscopy of eroded nodules of the nail bed often permits detection of peripheral pigmentation in amelanotic melanoma, allowing differential diagnosis from pyogenic granuloma and non-melanocytic nail tumors.

Nail Matrix Dermoscopy

Intraoperative dermoscopy permits direct visualization of the site of melanin production in the nail bed or matrix with patterns that are

Table 1 ABCDEF Mnemonic for Subungual Melanoma

<table>
<thead>
<tr>
<th>A</th>
<th>B</th>
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<th>D</th>
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<tbody>
<tr>
<td>= age (peak in 5th-7th decades), Asian, African-Americans, Native Americans</td>
<td>brown to black band width breadth of 3 mm or more with variegated borders</td>
<td>change in nail band despite treatment</td>
<td>digit</td>
<td>extension of pigment onto the proximal and/or lateral nailfold (Hutchinson’s sign)</td>
<td>family or personal history of dysplastic nevus or melanoma</td>
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Table 2 Nail Pigmentation: Clinical Signs that Suggest Immediate Excisional Biopsy of the Pigmentation to Exclude Nail Melanoma

- Lack of homogeneity of the pigmentation, with bands or lines of different color
- Presence of nail plate fissuring or splitting
- Proximal part of the band broader than the distal (triangular shape)
- Blurred lateral borders of the band
- Pigmentation of the periungual skin (Hutchinson’s sign)
similar to those found in skin melanocytic lesions. Dermoscopy of
the nail bed and matrix is also very useful to select the surgical
margins, and may avoid omission of small pigment foci.23,26

Management

The main challenges in the management of a patient with mela-
nonychia are to distinguish melanoma from benign conditions
(avoiding delayed diagnosis), to define proper guidelines for follow-
up, to establish the best modality for obtaining a pathological sam-
ple from a suspicious lesion, and to establish the pathological diag-
nosis of in situ melanoma.

Early Diagnosis of Subungual
Melanoma and Melanoma of the Nail Matrix

Early diagnosis and treatment of melanoma offer the only possibility
of curative treatment. When considering a pigment band, we should
first establish whether the pigmentation is caused by melanin or by
another pigment. If the pigmentation is melanin, then it is important
to differentiate bands caused by melanocyte activation from bands
caused by melanocyte hyperplasia. This can usually be done by
history, clinical examination, and dermoscopy.

Nonmelanin nail pigmentation and melanonychia caused by mel-
anocyte activation do not need invasive investigations and close follow-
up. Management depends on the patient’s age. We believe that it is
probably advisable to obtain a biopsy for bands caused by melanocytic
hyperplasia in adults but not in children, in whom a “wait-and-see”
attitude can be adopted until puberty. However, the decision must also
take into account the size of the lesion, because small (3 mm) bands
can be excised without residual scarring. Excision avoids anxiety of patients
and parents who may be reluctant to accept “simple” clinical follow-up.
It is always important to ask for personal or familiar history of mela-
nona or atypical nevi syndrome.

In adults, clinical features that suggest melanoma and require
immediate excision and pathological study of the pigmented band
include a single affected digit, lack of homogeneity with bands or
lines of different color, presence of nail plate fissuring or splitting,
rapid enlargement of the band, a proximal part of the band that is
blurred lateral bor-
nique to evaluate the whole lesion and provide accurate diagnosis without leaving a definitive nail dystrophy. This approach is still new and requires confirmation.

We have no data at all about progression of nevi that have not been excised. There is evidence that some nevi may become “amelanotic,” therefore, they are clinically invisible and possibly persist undetected in adult life. The significance of benign melanocytic hyperplasia is unknown as is the possible risk of evolution to malignant melanoma.

Our attitude is to excise, with a nail matrix shave biopsy, lesions with alarming clinical and/or dermoscopic features, particularly bands that enlarge or darken, and to follow up every 6 months all the other lesions that we usually excise after puberty.

**Second Problem**

Is Nail Plate Dermoscopy Reliable in the Evaluation of Nail Pigmentation?

The experience in this field is still limited, and there are no data showing that dermoscopy is superior to clinical evaluation in early detection of melanoma of the nail unit. In 2007, Professor Nilton Di Chiacchio from San Paulo and one of the authors (AT) founded an International Study Group on Melanonychia, which now has 30 members from 12 countries around the world with the goal of defining the fields that need investigation and of providing evidence-based information to clinicians dealing with nail pigmentation. These include creation of a melanonychia registry on a dedicated Web site, pathological consensus on diagnosis of pigmented nail lesions, consensus on guidelines for management of melanonychia in children, follow-up of patients undergoing excisional biopsies with different techniques to establish recurrence rate, long-term follow-up of nevi with nail plate dermoscopy. This will possibly permit collection of evidence-based data to diagnose and manage patients with melanonychia better and then improve the prognosis of patients with subungual melanoma.

**References**

12. Thomas L, Dalle S: Dermoscopy provides useful information for the management of melanonychia striata. Dermatol Ther 20:3-10, 2007